

Disorders of the thoracic spine: pathology and treatment

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Disorders and their treatment

Tumours of the thoracic spine

Spinal neoplasms, both primary and secondary, are unusual causes of thoracolumbar pain. However, because these lesions are associated with high mortality, examiners must always be aware of the possibility of neoplastic diseases and must include them in their differential diagnosis.

Malignant spinal tumours occur mainly in the middle aged and are characterized by a more or less sudden onset of the symptoms; often a minor trauma is thought to be the inciting cause and progression is swift.

According to their localization (in or outside the spinal canal) tumours can be classified as intra- or extraspinal:

• *Intraspinal tumours* are sited in the spinal canal. They are further divided into intra- and extradural tumours. The former may be intra- or extramedullary.

• *Extraspinal tumours* involve the bony parts of the vertebrae. Benign tumours are usually located in the posterior parts (spinous and transverse processes), malignant tumours in the vertebral body.

Intraspinal tumours

Thoracic neurofibroma

Pathology

This benign tumour usually originates from the dorsal root and arises from proliferating nerve fibres, fibroblasts and Schwann cells. Sensory or motor fibres may be involved.¹ Some confusion exists about the terminology: various names, such as neurofibroma, neurinoma, neurilemmoma or schwannoma have been used. Some believe that all these terms cover the same type of tumour; others distinguish some slight histological differences between them. Multiple tumours in nerve fibres and the subcutaneous tissues, often accompanied by patchy café au lait pigmentation, constitute the syndrome known as von Recklinghausen's disease.

Neuromas are the most common primary tumours of the spine accounting for approximately one-third of the cases. They are most often seen at the lower thoracic region and the thoracolumbar junction.² More than half of all these lesions are intradural extramedullary (Fig 1), 25% are purely extradural, 15% are both intradural and extradural, and very rarely they are seen intramedullary.^{3,4} They may become symptomatic in patients at any age, but the peak incidence is around the fourth and fifth decades.⁵ The tumours are benign, slow-growing and remain solitary,⁵ well circumscribed and encapsulated. Often there is a cystic degeneration within the tumour. A tumour that is within the intervertebral foramen is often shaped like an hourglass or dumb-bell, one limb of which can give rise to a paravertebral extraspinal extension, which is sometimes palpable.

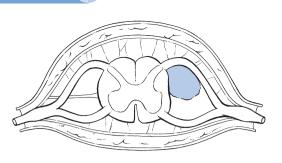


Fig 1 • Intradural extramedullary neurofibroma.

History

Symptoms are usually due to the compression of dura, spinal cord and roots. Few symptoms are present until the tumour reaches a large mass. Cystic tumours have a high risk of causing progressive symptomatic worsening as a result of cyst expansion.⁶

In the early stage, the diagnosis is often difficult because neurofibromas usually give rise to symptoms almost identical to those of a disc lesion.^{7,8} Thoracic neuromas may even simulate a disc problem at the lumbar level.^{8–11}

The main symptom is pain, most frequently present as root pain, exceptionally as a dural pain. It is increased by a deep breath or by a cough and may be present at night. Sometimes the patient prefers to sleep sitting up in a chair, a situation that is most unusual for a disc lesion. Activity or movement influences the pain only exceptionally. Pain increases slowly over months and years, a characteristic that indicates an unusual, slowly progressive disorder.

Involvement of the sensory fibres may result in a segmental band-shaped area of numbness. The tumour may also compress the spinal cord, affecting both motor and sensory elements (see p. 390). As a consequence, the patient may complain of stiffness of the legs, muscle spasms, extrasegmentally referred pins and needles, and disturbed sphincter function with loss of bowel or bladder control.¹⁰ Exceptionally Brown–Séquard syndrome occurs.⁵

Clinical examination

Interference with the dura usually produces a clinical picture almost identical to that of an ordinary disc lesion.

Neck flexion and scapular approximation are often painful. These signs, together with pain on deep inspiration, are typical of a lesion that is interfering with the dura mater or with the dural nerve sleeve. They are not specific to a disc lesion and may occur in any space-occupying lesion.

Because the tumour lies outside the intervertebral joint, it does not interfere with articular movements and therefore daily activities and movements are hardly affected. Side flexion away from the painful side is sometimes the only painful and restricted movement and this pattern is unlikely to result from a disc lesion.

All signs slowly and progressively increase. Finally, neurological signs may develop but they come on much later than in malignant tumours. They may consist of a band-shaped numbness related to one dermatome. The tumour may also affect motor fibres but segmental motor deficit is very difficult to detect. Once the tumour compresses the spinal cord, any of the signs of cord compression may be encountered: depression of abdominal reflexes, hyperactive patellar and Achilles tendon reflexes and sensory loss.⁴

When a neurofibroma is suspected, the paravertebral area should always be palpated because a neurofibroma may expand extraspinally via the intervertebral foramen to give rise to a paravertebral mass.

Warning

Warning signs in neurofibroma:

- Pain: slowly increasing pain over months, mainly felt at night, uninfluenced by activities
- Patient prefers to sleep sitting up
- Side flexion away from painful side is the only painful and limited movement
- Band-shaped area of numbness related to one dermatome
- Presence of pins and needles in one or both feet or any other sign of cord compression
- Disorder mistaken as a disc lesion, which does not respond to manipulations.

Special investigations

On plain radiography there may be an enlarged intervertebral foramen and erosion of the vertebral body or the pedicles. X-ray of the thorax may show an abnormal soft tissue mass, extending outside the spinal canal (hourglass tumour).

MRI appears to be the most sensitive investigation for identifying these lesions. Nerve sheath tumours have equal or less signal intensity on T1-weighted images and mild to marked hyperintensity on T2-weighted images as compared to the spinal cord.¹² Focal areas of even greater hyperintensity on T2-weighted images often correspond to cystic portions.

Treatment and prognosis

Neurofibromas may undergo malignant change or produce cord compression. Therefore, they should be surgically removed. Small posteriorly or laterally sited tumours can usually be dealt with easily. Anterior neurofibromas present more problems and usually can be only partly excised.¹³

Other intraspinal masses

These include all other infra- and extradural tumours of the spinal canal. Despite the advantage of specialized technical investigations (radiography, myelography, CT, MRI), patients with intraspinal tumours are often misdiagnosed.

Clinical presentation

The primary and most universal symptom is pain, which is usually felt centrally in the back and may spread bilaterally as girdle pain.¹⁴ The pain increases progressively and is relentless, despite the patient's attempts to limit activities. In anterior compression of the dural sac, L'hermitte's sign is sometimes present.¹⁵

Occasionally the pain is worst at night. Although this is classically regarded as being suggestive of a tumour, it is rather rare.^{8,9}

Straining and coughing may increase the pain as may active movement, but to a lesser degree than in mechanical disorders. $^{9,16}\,$

The clinical pattern depends on the extent of the tumour: all tests can be completely normal or movement may be considerably limited. If the latter, anteflexion is usually involved and there is often associated muscle spasm. As a rule, in intraspinal soft tissue masses, not much is learnt from articular movements. Besides the positive articular signs, all intraspinal masses give rise sooner or later to neurological signs caused either by involvement of one or more nerve roots or by compression of the spinal cord (see p. 390).

It should be noted that a tumour is not always found where it would be expected on a clinical basis. Cases have been reported where upper thoracic tumours gave rise to pain and neurological signs in the lower limb or in the lumbar area.^{8,9}

Further investigations

In all instances, further investigation is called for. Examination of the cerebrospinal fluid may show elevated proteins, a finding strongly suggesting a neoplasm.⁹

The imaging techniques used to define intraspinal tumours are MRI and CT-myelography. The advantages of MRI are: ease of obtaining sagittal sections; rapid assessment of the entire thoracic spine; patient acceptability because of the absence of radiation and low invasiveness; and lack of morbidity and complications.

Neoplasms that are frequently associated with skeletal metastases include tumours of the breast, prostate, lung, kidney, thyroid and colon.^{17–20} Data from autopsy material suggest that up to 70% of patients with a primary neoplasm from one of these sources will develop pathological evidence of metastases to vertebral bodies in the thoracolumbar spine.²¹ Because the majority of metastases occur in the vertebral body, they may cause anterior compression of the spinal cord, either directly by tumour growth or by a pathological fracture with retropulsion of bone and disc fragments into the spinal canal.²² Finally compression of the cord can result from an intradural metastasis.²³

Extraspinal tumours

Neoplasms located outside the spinal canal are called extraspinal tumours.

In general, benign tumours are located in the posterior elements of the vertebrae and are found in patients under 30 years of age, whereas malignant tumours (both primary and metastatic) are located in the anterior components of the vertebrae and are more common after the age of 50. Myelomas and metastases are the most frequent malignancies.¹⁷

Multiple myeloma is a malignant tumour of plasma cells. Plasma cells produce immunoglobulins and antibodies and are located throughout the bone marrow. Multiple myeloma is the most common adult primary malignancy of bone. The patients are usually in an older age group, ranging between 50 and 70. The disease leads to widespread bone destruction, abnormal immunoglobulin production and infiltration of bone marrow. Therefore a patient with multiple myeloma develops a broad range of clinical symptoms that extend beyond the orthopaedic field. Symptoms are also related to hypercalcaemia (anorexia, nausea, vomiting, kidney stones and changes in mental state), abnormal immunoglobulin concentrations (renal insufficiency and amyloidosis), haematological changes (anaemia and bleeding disorders) and generalized weakness.

Diagnosis is made on laboratory examination and radiographic evaluation.

Spinal metastases are much more common than primary tumours. The prevalence increases with increasing age. The lumbar and thoracic spines are affected in approximately 46– 49% of cases, with a predilection for the thoracolumbar junction (the so-called forbidden area). Consequently signs of cord compression are often present.

In other circumstances, radicular pain occurs. This is most misleading in the thoracic area, where it often mimics visceral problems.

Clinical presentation

In 95% of cases, the first symptom is local neckache or local thoracic backache, which goes on to radiate. Suspicion should arise when this occurs in patients over 50 years of age complaining for the first time of backache not preceded by trauma. The pain tends to increase in intensity progressively and to involve a larger area: expanding pain. If radicular pain is present, it is usually worse at night (Borenstein and Wiesel:¹ p. 309).

In vertebral metastases a full articular pattern is usually present, characterized by marked painful limitation on extension and symmetrical limitation of both side flexions and rotations. Muscle spasm may also be present. Local tenderness over the spinous process is found on palpation.

Radicular signs or those of cord compression may occur abruptly, with full neurological features developing in 48 hours or more commonly subacutely over a week or more. Some patients have a much slower course – neurological symptoms and signs progressing over 4–6 months.

Tumours of T12 or L1 may compress the conus medullaris, containing the S3–S5 nerve roots. This may lead to dysfunction of the urinary or anal sphincter, which is sometimes associated with saddle anaesthesia.

Differential diagnosis (see Tables 1, 2) must be careful,^{8,9,16, 24–29} because the majority of the signs and symptoms also occur in ordinary thoracic disc lesions. It is based mainly on clinical examination, because up to 30% of the bone mass has to be lost before metastases may become visible on radiography.³⁰ When there is the slightest possibility that there is a tumour, manipulation should never be done and further investigations such as a bone scan must be carried out.

Extradural haematoma

Extradural spinal haematoma at the thoracic level can be either traumatic or nontraumatic. Spinal epidural haematoma (SEH) is an uncommon complication of severe spine injury with or without fracture of the thoracic vertebral bodies.

Table 1 Differential diagnosis of thoracic disc protrusion and tumours

	Thoracic disc protrusion	Benign tumour	Malignant tumour	
			Intraspinal	Extraspinal
Age (years)	20–50	Any age	>50	>50
Evolution	Swift or chronic (ups and downs)	Very slow	Swift	Swift
Pain At night	Shifting Better	Expanding Worse	Expanding Worse	Expanding Worse
Dural symptoms On inspiration or cough	±	+	±	±
Articular signs	Partial articular pattern	Full range, or side flexion away from painful side is the only painful and limited test	All tests normal, or limitation of anteflexion Muscle spasm	Full articular pattern Muscle spasm
Dural signs				
Neck flexion Scapular approximation	+ +	+ +	+ +	+ +
Neurological signs	Unusual	+	+	±
Palpable mass	-	±	-	-

Table 2 Differential diagnosis of thoracic disc protrusion and neurofibroma

	Neurofibroma	Thoracic disc protrusion
Age	Young	20-50 years
Pain	Slowly increasing	Swift onset; if chronic, no increasing pain but ups and downs
On inspiration or cough	+	+
Preferred sleeping position	Sitting up	Lying down
Articular movements	Usually negative Side flexion away from the painful side: painful and limited?	Partial articular pattern
Dural signs		
Neck flexion	+	+
Scapular approximation	+	+
Neurological signs		
Band-shaped area of numbness in one dermatome	+	Unusual
Segmental motor deficit	+	Unusual
Signs of cord compression	+	Unusual
Palpable mass	±	_

Compression of the cord by a haematoma is also a wellknown, although rare, complication of spinal surgery.³¹ The reported frequency is between 1 and 6 per 1000 operations. Traumatic bleeding in the epidural space has also been reported after epidural injections and chiropractic manipulations.³²

Non-traumatic extradural spinal haematoma is an uncommon condition often associated with a poor outcome. There seems to be an increase of the incidence, probably from the increased use of thrombolytic and anticoagulant therapy.³³⁻³⁵ Other causes of non-traumatic extradural spinal haematoma include vasculitis such as systemic lupus erythematosus (SLE), spinal arteriovenous malformations and haemophilia.^{36,37}

A spinal epidural haematoma may present acutely or subacutely over a number of days or weeks and with fluctuating symptoms. The patient usually suffers from increasing and expanding thoracic back pain, followed by progressive signs and symptoms of major neurological dysfunction secondary to cord compression.³⁸ Conditions that may mimic an acute spinal haematoma include extradural abscess and extradural metastatic infiltration. It is important to make an early diagnosis because surgery may offer the best hope of restoring neurological function. MRI is the examination of choice and provides characteristic findings that allow a prompt diagnosis.³⁹ The technique can also provide useful information about the age of the haematoma.⁴⁰

Spinal epidural haematoma has always been considered a neurosurgical emergency. The treatment of choice is decompressive surgery as soon as possible because permanent neurological disability or death may follow if neurosurgical intervention is delayed.^{41,42} However, during the last decade several reports have been published showing that non-operative treatment may be successful in cases with minimal neurological deficits, despite cord compression revealed by MRI.^{43–45}

Spinal cord herniation

Spinal cord herniation is a rare, although increasingly recognized, cause of spinal cord dysfunction. It has been ascribed to a dural defect, either congenital or acquired, in the anterior surface of the dural sac through which the spinal cord herniates.^{46,47} The number of published cases in the English language literature markedly increased after 2000. Awareness of the clinical setting and the wider use of MRI in myelopathy are considered the pertinent factors in this increase.⁴⁸

The main clinical features are thoracic pain and a Brown–Séquard syndrome. Although the dura is sensitive to pain, review of the literature shows that only 48% of the patients had thoracic pain.^{49,50} About 73% present with Brown–Séquard syndrome^{51,52} (spastic paralysis on the ipsilateral side together with numbness on the contralateral side). This is probably caused by tethering of the spinal cord at the side of the herniation which results in unilateral damage of the lateral funiculus.⁵³ Some patients have signs of only spasticity or numbness in one leg.⁵⁴

MRI is the gold standard technique for diagnosis of spinal cord herniation. On sagittal MRI, typical features are ventral displacement, sharp ventral angulation of thoracic spinal cord, and enlargement of dorsal subarachnoid space.^{55,56}

Surgery is crucial in the management of this rare entity, and duraplasty the more widely performed method.⁵⁷

Thoracic spinal canal stenosis

This may be the outcome of either congenital deformation or hypertrophy of the posterior spinal elements. Most often it occurs in association with generalized rheumatological, metabolic or orthopaedic disorders, such as achondroplasia, osteofluorosis, Scheuermann's disease, Paget's disease or acromegaly. It is rare in the absence of a generalized disorder.^{58,59} Degenerative changes in the facet joints and the intervertebral disc can diminish the volume of the spinal canal and cause cord compression.^{60,61} The latter is most frequently found at T11 and T12 in middle-aged people.^{62,63}

Clinical presentation

Patients usually complain of pseudoclaudication, characterized by pain in the lower back, buttock, thigh and in the legs, coming on during walking; sometimes standing may provoke symptoms.

In addition to pain, paraesthesia may be present in one or both lower limbs. The sensation is worse on walking and improves on sitting. Numbness in the lower limbs may follow and may be associated with muscular weakness, difficulty on walking and easy fatigue. Spastic paraparesis with diminished or lost reflexes in the lower limbs, together with urinary disturbance, can be present. In rare cases, hyperreflexia is noted.

Extension of the thoracic spine while upright may further compromise the cord within the narrowed canal and increase the symptoms, while improvement occurs on anteflexion.

On clinical examination there may be some degree of limitation of movement in the spine, because of osteoarthritis leading to stiffness. In such a case, a capsular pattern is found.

Arterial pulses in the lower limb are normal, which largely excludes vascular problems.

Further investigations

The radiograph is often unremarkable and myelography can be misleading. CT scan and MRI are usually required.^{58,62}

Differential diagnosis

Two differential diagnoses should be considered:

- *Intermittent claudication:* when pseudoclaudication is present, differential diagnosis must be made from intermittent claudication caused by vascular abnormalities. In spinal stenosis, some patients have symptoms only on standing. In intermittent claudication the pain is brought on by walking and relieved on standing still; arterial pulses are usually diminished or absent. Doppler probe and arteriography may confirm the diagnosis.
- Disc protrusion: in spinal stenosis all postures or movements that bring the spine into anteflexion usually relieve the pain, whereas in a disc protrusion the opposite is more usual. Moreover, in a simple disc protrusion, activity of the lower limbs has no influence on the symptoms.

Treatment

Surgical intervention is necessary but established neurological features seldom regress.

Chest deformities

Hyperkyphosis

The normal thoracic spine is kyphotic but if the kyphosis is beyond 40°, hyperkyphosis is present.⁶⁴ The condition may occur at one or more levels and may be the result of several disorders.⁶⁵

Juvenile kyphosis

Juvenile kyphosis has its clinical onset in adolescence between the ages of 14 and 18 years; for this reason the condition is sometimes known as adolescent osteochondritis. There is a slight preponderance in females.⁶⁶

The Thoracic Spine

It develops because of a disturbance in growth in the vertebral rim epiphysis akin to osteochondritis dissecans.⁶⁷ Although the exact aetiology is unknown, it is generally believed to result from an anterior endplate lesion, through which herniations of the intervertebral disc protrude into the adjacent bone (Schmorl's nodes).⁶⁸ The intervertebral disc itself becomes narrowed, mainly anteriorly. The protrusion interferes with the growth of the vertebral ring epiphysis, which finally leads to about 5° of anterior wedging of the vertebral body. Should this occur over several levels, thoracic hyperkyphosis (with the apex normally around T7–T9) results. In this event, the condition is named Scheuermann's disease.

Clinical presentation

In adolescents the disorder usually remains painless. It usually gives rise to progressive silent hyperkyphosis, with backache present only in a minority of cases.^{69,70} At a later age, however, there is a predisposition to thoracic pain⁷¹ (see p. e175 of this chapter).

On clinical examination, all movements are free except active extension, which may be limited. If hyperkyphosis is present, it does not disappear in prone position, and when a hyperextension thrust is given to the thoracic spine, stiffness is felt and slight discomfort provoked.

Radiography

The diagnosis is mainly based on radiographs, which are often taken for some other reason. They show narrowing of the intervertebral joint space and multiple Schmorl's nodes, giving an irregular aspect to the surface of the vertebral body. In the later stage, focal anterior wedging of the latter is seen.

Treatment

This should aim to prevent further deformation during the growth period. If the kyphosis is still reversible, active exercises for the extensor muscles of the back should suffice. In more progressive cases, in which hyperkyphosis increases or progression of anterior wedging of the vertebrae is found, a Milwaukee brace until growth has ceased is the best advice.

Hyperkyphotic posture in the elderly

Age-related hyperkyphosis is an exaggerated anterior curvature in the thoracic spine that occurs commonly with advanced age. Epidemiologic studies have demonstrated that age-related hyperkyphosis commonly affects the elderly population with estimates ranging from 20% to 40%.⁷² The 'dowager hump' is well recognized, and most clinicians and patients equate this with spinal osteoporosis and vertebral compression deformity or angulation.⁷³ Increasing thoracic kyphosis, in particular when linked to back pain, is considered a signature of possible vertebral body compression fracture. However, approximately two-thirds of those who are most hyperkyphotic don't have vertebral fractures.⁷⁴ In the absence of vertebral compression fracture, changes in the spinal support tissues (i.e. ligaments, tendons, disk annulus and nucleus) or supporting musculature could also lead to a progressive increase in curvature.⁷⁵

Apart from being a cosmetic deformity, most cases of hyperkyphosis do not cause much pain or suffering. However, in some instances they may lead to an increased risk of vertebral fractures⁷⁶ or may be complicated by a thoracic postural pain syndrome (see following page).

Therapy includes the treatment of the acute exacerbations (see fractures), and of a thoracic postural pain syndrome. Also pharmacological treatment of the underlying osteoporosis with calcium and vitamin D supplementation together with bisphosphonates may be indicated.⁷⁷

Vertebral body fractures

The thoracolumbar spine is the most common site for vertebral fractures.⁷⁸ In younger patients, thoracolumbar vertebral fractures are usually caused by high-energy accidents such as falls, or motor vehicle accidents; whereas in elderly patients, osteoporosis is the dominant aetiology.⁷⁹ Vertebral body fractures may also occur spontaneously as the result of an underlying disorder, such as a vertebral tumour, infection, or ankylosing spondylitis. All are classified as pathological fractures.⁸⁰

High-energy fractures usually arise from an axial load in combination with flexion or lateral bending. If a compression injury with a significant flexion component is the cause, a wedge-shaped deformity with considerable loss of anterior vertebral height is commonly present (Fig. 2), resulting in angular kyphosis. Lateral bending leads to lateral wedging. In some cases the whole vertebral body is shattered (burst fracture) which can lead to serious neurological complications (Fig 3). A uniformly flattened vertebral body is more indicative of a pure axial component. Vertebral compression fractures due to osteoporosis are usually wedge fractures and have a milder clinical appearance.

Osteoporotic compression fractures

A fracture occurs when the weight of the upper body exceeds the ability of the bone within the vertebral body to support the load. In cases of severe osteoporosis the trauma may be subtle, such as stepping out of a bathtub, vigorous sneezing, or

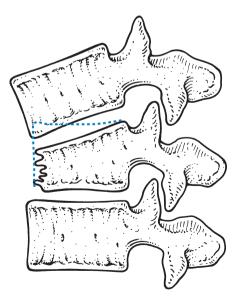


Fig 2 • Wedge compression fracture: the middle column (posterior part of the vertebral body) remains intact.

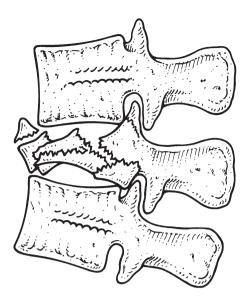


Fig 3 • Burst fracture: the posterior half of the vertebral column is disturbed and retropulsion into the neural canal may take place.

lifting a trivial object, or the trauma may result from the load caused by muscle contraction. In cases of moderate osteoporosis, more force or trauma is required to create a fracture, such as falling off a chair, tripping or attempting to lift a heavy object.

Symptoms

Central thoracic pain and bilateral girdle pain referred to the corresponding dermatomes. Standing or walking exacerbates the pain. Unsupported sitting is also uncomfortable. There may be twinges. Lying in the supine position generally relieves some of the discomfort.⁸¹ Spinal compression fractures can also be insidious and may then produce only modest back pain early in the course of the progressive disease. This may explain the fact that only one-third of vertebral fractures are actually diagnosed, as the patient regards his back pain as a normal part of aging.⁸²

Signs

Active and passive movements show a symmetrical pattern with pain at the end of both rotations and sideflexions. Extension is painful and limited and flexion is performed with support of the arms. Taking a deep breath may influence the pain. In cases of uncomplicated compression fractures, neck flexion and SLR are negative and neurologic examination is normal. Palpation shows tenderness directly over the area of the fracture, and an increased kyphosis may be noted.

Radiography

Plain frontal and lateral radiographs are the initial imaging study obtained for a suspected compression fracture. Compression of the anterior aspect of the vertebrae results in the classic wedge-shaped vertebral body with narrowing of the anterior portion. A decrease in vertebral height of 20% or more is considered positive for compression fracture.⁸³ Computed tomography can be used for evaluating the posterior vertebral wall integrity and for distinguishing a wedge fracture from a burst fracture. In the latter, the middle column, consisting of the posterior half of the vertebral body, the posterior

longitudinal ligament and the posterior fibres of the annulus fibrosus, is disturbed, and varying degrees of retropulsion into the neutral canal takes place, provoking neurological signs (Fig. 3).⁸⁴

Natural history

After about 2 weeks the pain diminishes progressively and it takes about 3 months for an uncomplicated wedge fracture to heal, leaving the vertebral body with a permanent wedge shape. Pain that is still present after the fracture has healed is explained by collateral damage that has been done to the disc or the surrounding ligaments. It should be noted that the kyphosis deformity may also cause later development of spinal stenosis or thoracic postural pain syndrome.

Treatment

Uncomplicated wedge fractures do not need specific treatment apart from relative rest during the period of spontaneous healing. If there is still pain after 3 months, a complete clinical re-assessment should be performed to reveal the source.

During the last decades vertebroplasty and kyphoplasty have been promoted for the management of severe recalcitrant pain. Initially reported by Galibert *et al* in 1987, vertebroplasty involved the destruction of an angioma through consolidation of the vertebral column by percutaneous injection of acrylic cement;⁸⁵ however, vertebroplasty is now commonly used in treatment of painful osteoporotic vertebral compression fractures.⁸⁶ Kyphoplasty involves the use of an inflatable bone tamp that when introduced into the vertebral body, restores vertebral height and forms a space for injection of acrylic cement.⁸⁷

Pathological fractures

These are mainly the result of vertebral infections, ankylosing spondylitis and primary tumours or metastases.⁸⁰ If the patient complains of severe pain after trivial injury, a pathological fracture should be suspected. Sometimes the event may have been so minimal that the patient does not always mention it. The clinical presentation is the same as for other fractures of abrupt onset except that the pain may not fully disappear on recumbency.

Diagnosis is made with imaging techniques.

Warning

The following may indicate a fracture:

- Severe bilateral girdle pain after a minimal injury in the absence of pain on flexion of the neck: pathological fracture?
- Very severe pain on extension pressure.
- Capsular pattern following an injury.

Thoracic postural pain syndrome

Cyriax regarded this pain syndrome, which affects mainly young adults and the middle aged, as being the result of a posterior displacement of the whole intervertebral disc content, occurring mainly in patients with a marked thoracic kyphosis (Cyriax⁷). Although dural compression by the entire disc is very likely, ligamentous overstretching of the supra- and interspinal ligaments may also be a contributing factor. Prolonged and repeated anteflexion stretches the posterior longitudinal ligament, which may become too elastic. At the same time, the kyphotic posture causes a posterior directed force on the disc. After some years, the whole disc content bulges posteriorly through the longitudinal ligament. At first this occurs only during prolonged anteflexion and ceases slowly on lying down – the phenomenon is self-reducing. Later the displacement may become permanent. Once the disc comes in contact with the dura mater, or when the posterior ligaments become severely overstretched, the patient complains of pain, which is initially temporary but later becomes permanent.

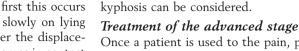
The whole process depends on three elements, all of which aggravate the disorder:

- The duration of forward bending
- The degree of physiological kyphosis
- The load put on the disc.

Clinical presentations

The history is highly characteristic. Initially, the patients are well on first waking up in the morning, but pain starts after some hours of forward bending or after prolonged sitting in a kyphotic position. It is usually felt in the centre of the upper or mid-thoracic area but later may spread over the whole posterior thorax. The pain increases on carrying heavy objects or when the period of anteflexion is of longer duration. Initially, on lying down the pain disappears after some minutes, but in advanced cases it may take hours to ease off. As the condition develops, interscapular pain begins and increases slowly and steadily during the day, even if the patient does not bend forward. Finally pain becomes continuous day and night – the stage of anterior erosion has been reached.

On clinical examination, only a few articular signs are present and they are not very pronounced. In this disorder, a deep breath usually hurts at the moment the pain is maximally present. This element draws attention to a dural involvement (Fig. 4).



Once a patient is used to the pain, prediction of exactly after how many hours the pain will take to come on is possible. At this stage, the advice is to lie down before severe pain starts.

Early treatment is in fact prevention: all kyphotic postures

over several adjacent levels around the maximum point of the

If this is not sufficient, ligamentous sclerosant infiltrations

Besides preventive rest, daily traction can be given over 2 weeks, followed by a traction-free interval of 1 week, and with repetition for a further 2 weeks. The period without traction is progressively increased to a month, alternated with 2 weeks of traction. If this is successful, it should be continued indefinitely.

Anterior erosion

Treatment of the initial stage

should be avoided.

This is the final stage of the postural pain syndrome and is encountered only in the elderly. Progressive increase in hyperkyphosis finally erodes the anterior portion of the disc and the anterior bony parts of the vertebrae are in contact with each other. Bone sclerosis develops and anterior osteophytes form (Fig. 5). Pain that previously ceased at night is now constantly present and is probably due to the osseous contact between the vertebrae, with subsequent local inflammation. For this reason the pain remains local.

On clinical examination there is a marked thoracic kyphosis with very rigid joints. Therefore, the active articular movements no longer have much influence on the pain, but passive

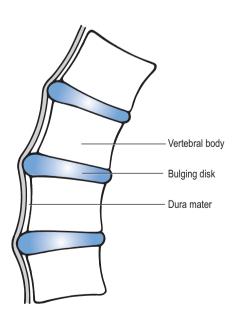


Fig 4 • Thoracic postural pain syndrome.

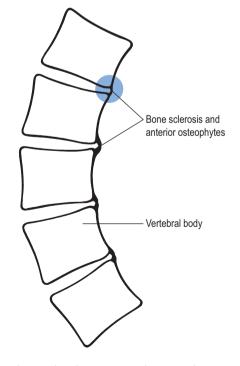


Fig 5 • Anterior erosion: bony contact between the two vertebral bodies at the anterior side may result in bone sclerosis and pain.

articular movements may slightly increase the pain. Anterior osteophytes are visible on the radiograph.

Treatment consists of repeated traction at intervals as the complaints demand, as for the thoracic postural pain syndrome (see earlier). Recent studies demonstrated a significant improvement in pain and posture after rehabilitation of the kyphotic spine (combination of manual mobilization, taping and exercises).^{88,89}

Scoliosis

Scoliosis⁹⁰ is defined as a lateral curvature of the spine of greater than 10° . The angle is determined by drawing lines across the upper surface of the vertebral bodies at which the curve changes direction – i.e. the vertebrae that tilt maximally into the concavity of the curve. Lines perpendicular to these are then drawn. The angle between the two perpendicular lines is Cobb's angle (Fig. 6). If it is greater than 10° , clinical scoliosis is present.

Sometimes scoliosis is associated with an increased thoracic kyphosis (posterior convex curvature) or a diminished lumbar lordosis (anterior convex curvature).

There are two types of scoliosis.⁶⁴ In structural scoliosis, the deformity cannot be voluntarily corrected by the patient. It is characterized by a fixed rotation on forward bending. Some cases are congenital, others acquired. Non-structural scoliosis is frequently postural and remains under voluntary control. No fixed rotation is present on anteflexion. It is not progressive.

Structural scoliosis

Congenital scoliosis

This is the result of congenital malformation such as hemivertebra. It is frequently severe and may produce grotesque

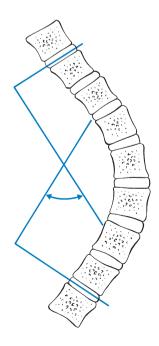


Fig 6 • Cobb's angle.

cosmetic deformity, reduced pulmonary function and ultimately paraplegia.

Acquired scoliosis

This is either secondary or idiopathic.

Secondary scoliosis

This has a local cause, often in association with developmental disturbances. Examples are intraspinal tumours (neurofibromatosis), neuromuscular dysfunction as in myopathies (muscular dystrophy), mesenchymal disorders (Marfan's syndrome), vertebral tumours, rheumatoid diseases, fractures and nerve root irritation. The pathophysiological mechanism of the thoracic malformation remains unknown in many of these.

Idiopathic scoliosis

In this, an underlying disorder is not present. Ninety per cent of cases are in this group, and some authors estimate the incidence as approximately 4.5% of all schoolchildren.^{1,91}

Depending on the age of onset it is known as infantile scoliosis (0–4 years), juvenile scoliosis (4–10) and adolescent scoliosis (over 10 to skeletal maturity). Adolescent scoliosis sometimes has adult onset when it affects females between 20 and 40 years of age. Severe adolescent scoliosis affects girls four times more often than boys, whereas less pronounced deviations are seen equally in both sexes.⁹¹

One curious feature of the disorder is that in childhood, 90% of the curvatures are convex to the left, whereas in adolescence this is reversed.⁹² The condition worsens as bone growth increases and rarely changes after bone growth has stopped. However, if the angle of deformity between the lumbar and thoracic spines remains less than 60°, an effect on vital functions is not to be expected.

Clinical presentation

Controversy surrounds the incidence of thoracolumbar pain in scoliosis. Some authors contend that in the main the condition is asymptomatic and that patients with scoliosis do not suffer more from back pain than those with a normal back.⁹³ Others maintain that the larger the scoliosis the more likely it is that pain will occur.⁹⁴ However, some patients initially experience pain at the end of the day, which stops rapidly on lying down.⁹⁴ The cause may be overstretching of ligaments, overloading of the facet joints or temporal displacement of the disc. All these possibilities are compatible with a non-articular pattern on clinical examination. The pain is usually located at the apex of the curvature. Progression of the deformity is usually associated with increased pain.

On inspection, attention must be paid to scapular asymmetry, prominence at one side of the thorax, the distance between the arms and trunk and the level of the pelvis. Because a contralateral rotation occurs on side flexion, scoliosis is associated with a unilateral thoracic prominence at the convex side of the scoliosis.^{64,95} Prominence of the thorax is most easily noted when the patient bends over.

During functional examination, one side flexion is sometimes limited. Pain, if present, is of the mechanical type and gives rise to a partial articular pattern on clinical examination. Advanced scoliosis may lead to a decreased vital capacity

Further investigations

and to cardiorespiratory dysfunction.

Radiography establishes the exact location of the curvature, its degree of severity (Cobb's angle) and the stage of skeletal maturity, all of which are important for diagnosis, follow-up and treatment. Skeletal maturation is judged from the ossification and closure of the vertebral ring apophyses.⁶⁴

Progression is defined as an increase in Cobb's angle of at least 5° between two consecutive radiographs.

Prognosis

The higher the curvature is situated in the spine, the less favourable the outcome. Progression is possible for as long as vertebral growth continues.

Treatment

Different types of treatment may be indicated: observation, drugs, exercises, braces and surgery (see Box 1). It should be noted that there is an important distinction to be made between the treatment for children and that for adults.

Observation

This is undertaken in the following:

- All patients who have an immature skeleton, and with curve of less than 20°
- Curves of less than 40° in skeletally mature patients
- Curves that are already thought to be non-progressive.

In all such cases, observation should be made at regular intervals to detect whether the curve progresses.⁹⁶ Usually it is sufficient to see the patient every 6 months. In adolescents

Box 1

Treatment of scoliosis

Observation	Immature skeleton, <20°
	Mature skeleton, <40°
	Non-progressive curves
Active treatment	1 0
Brace	Indications:
	Immature skeleton, >30°
	Progressive curves of >20°
	Mature skeleton <40°, no severe
	pain in young adult
	Contraindications:
	Mature skeleton
	Curve >40°
	Thoracic lordosis
Spinal fusion	Indications:
	Immature skeleton, >40°
	Mature skeleton, >50°
	Thoracic lordosis
	Failure of orthotic treatment
	Uncontrollable pain
	Progressive curve in adults
	-

nearing the growth spurt, a check-up every 3 months is more appropriate. Exercises are of no use in this stage.

Active treatment

Patients with a progressive curvature of 20° or more, or with an established curve greater than 30° , and who are skeletally immature require active treatment by either orthosis or operation:

- Brace: a young adult having a Cobb's angle of less than 40° , in the absence of severe pain, is best treated by conservative means: non-specific anti-inflammatory agents, facet infiltrations and physical monitoring. A Milwaukee brace can also be considered. The main goal of the use of a brace is to prevent further progression, rather than to correct the curvature. A Milwaukee brace or plastic jacket can be used, which extends the spine by pushing it up cranially from the hips. However, a recent study of late-onset idiopathic scoliosis raises questions about the efficacy of spinal orthoses.⁹⁷ Braces are contraindicated in skeletally mature patients or if the curvature is over 40° - the latter usually do not respond well. A brace cannot be used in thoracic lordosis. When brace treatment is given, associated exercises become important in order to keep the muscles of the trunk and the abdomen in good condition.
- Operation: if the curve is over 40° in a skeletally immature patient, or when it is progressive or over 50° in a mature skeleton, spinal fusion is indicated to straighten and stabilize the spine.⁹⁶ Other indications for operative intervention are when orthotic treatment has failed, if pain becomes uncontrollable or when a thoracic lordosis is found.

Older adults are more likely to suffer from neural compression. The major aim of treatment is to maintain function. With a curve under 40° this is usually possible without surgery. If there is uncontrolled pain or a progressive curve or structural disabilities, neurological complications or cardiorespiratory problems, surgical internal fixation is essential.^{5,98}

Non-structural scoliosis

The deformity is mainly found in adolescents and can be corrected by a voluntary effort. It is most frequently postural, occasionally the result of mental disturbance. On forward bending, the patient aligns the head and spine over the pelvis so that the scoliosis disappears. The term 'compensatory scoliosis' is sometimes used. The condition is not of clinical importance.

Scoliosis associated with inequality of limb length is usually confined to the lumbar spine. $^{99}\,$

Lateral erosion

Just as hyperkyphosis may lead to thoracic postural pain syndrome, and finally to anterior erosion, moderate and severe scoliosis may result in lateral erosion. The process is the same for both, but in scoliosis the entire disc of several consecutive levels shifts laterally to one side. After some years, this may

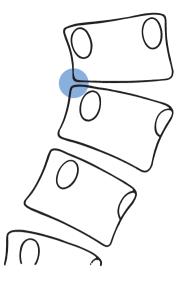


Fig 7 • Lateral erosion: bony contact between the two vertebral bodies at the concave side may result in bone sclerosis and pain.

lead to bony contact between the vertebral bodies at the concave side of the scoliosis (Fig. 7).

Because the disc is displaced directly laterally, pressure on the dura mater or nerve root is not to be expected. Consequently, pain is felt only late in the development of the condition when bone sclerosis starts. The pain remains unilateral and is not increased by a deep breath. Initially, it is felt only at the end of the day and disappears at night or after a period of recumbency. Later, the pain comes on earlier in the day and disappears only after lying down for longer and longer periods. Finally, it becomes constant.

Treatment

Traction in the same way as for anterior erosion can be given but is often not successful. Arthrodesis is the alternative.

Fracture of a transverse process

At the thoracic level this is usually the result of a direct blow or of an extension manipulation. After the injury the patient suffers immediately from unilateral segmental pain, increased on active and resisted side flexion towards the painless side. Resisted extension is also painful. Spontaneous resolution of the pain occurs in about 2 weeks.

Spinal infections

Vertebral osteomyelitis develops when an infectious organism enters the bone. A number of bacteria cause pyogenic osteomyelitis. Tuberculosis produces Pott's disease. An infection limited to the intervertebral disc is known as adult intervertebral disc space infection. In a later stage, when the infection progresses, it may also affect the vertebral body.

Both osteomyelitis and disc space infections must be differentiated from discitis or intervertebral disc inflammation, which only affects children.

Vertebral osteomyelitis

Pyogenic osteomyelitis

Since the introduction of antibiotics this disorder has changed from a disease of childhood and adolescence to one affecting mainly adults. The lumbar spine is most commonly involved.¹⁰⁰ The most frequent organism is a *Staphylococcus* sp.¹⁰¹ In about 40% of patients, an extraspinal primary source of infection is found, very often sited in the pelvis (urinary tract, postpartum, bowel).¹ The presence of diabetes mellitus, intravenous drug abuse, underlying chronic debilitating diseases or immunosuppression, previous infections, preceding bacteraemia and recent vertebral surgery are significantly associated with pyogenic osteomyelitis.^{102,103}

Clinical presentation

Although the onset may be abrupt, pyogenic osteomyelitis is most commonly insidious and progresses slowly. The pain is primarily felt locally at the site of the infection and is constantly present but often worse at night. At onset, the pain may be provoked or increased by activity and relieved in recumbency.

As the condition progresses, pain usually becomes more dramatic, leading to a full articular pattern with gross limitation mainly of extension, of both side flexions and rotations. Rigidity and muscle spasm are also present. Gross tenderness on local pressure over the spinous processes is found. A soft tissue mass may be palpable. Chills and weight loss complete the clinical presentation. Thecal sac neurocompression by the formation of an epidural abscess may cause severe neurological disturbances (paraplegia or paraparesis).¹⁰⁴

Further investigations

The erythrocyte sedimentation rate (ESR) is constantly raised and is a good indicator of the activity of the disease. 105,106 An increased white cell count is found in only half of the cases, and it seldom rises above $15\,000.^{102,107}$

Spinal radiographs become positive at the earliest after 2 or 3 weeks of infection,¹⁰⁷ when a narrowed disc space is the first sign found. This narrowing cannot be distinguished from narrowing that is the result of degenerative changes. Only later, after 10–12 weeks, do the adjacent endplates become more dense and eventually blurred, once destructive erosion has set in. Healing is associated with osteosclerosis and leads to bony fusion across the disc space.

Spongy bone erosion and rarefaction is apparent on CT scan before it is on plain radiography. However, gallium bone scan proves to be the most valuable technique to detect early inflammatory changes and it becomes positive earlier than does CT scan. In a recent study, gallium scanning proved to be 100% sensitive, specific and accurate.¹⁰⁸

Treatment

The administration of antibiotics and bed rest until the ESR has returned to normal. Treatment of neurological deficit caused by epidural abscess is prompt surgical decompression, with or without fusion.¹⁰⁹

Tuberculosis of the spine

Vertebral tuberculosis most frequently involves the lower thoracic and upper lumbar spines. It is always the consequence of haematogenous spread from a primary visceral focus (pulmonary or urinary tract) and it is the vertebral body that is involved, because the disc is avascular. However, spread to adjacent structures – discs, muscles and ligaments – may occur and a paravertebral abscess may form.

Clinical presentation

This may be very different from a pyogenic vertebral infection. The onset is very slow and it is not unusual that many months will have elapsed before the diagnosis becomes obvious. Backache of lesser severity than in pyogenic infection is usual. Other signs and symptoms – intermittent fever, night sweating, weight loss and fatigue – may draw attention to tuberculosis.

A localized angular bony deformity from vertebral collapse may be visible. On functional examination a full articular pattern is found, together with severe local tenderness on extension pressure. Dural symptoms and signs are not present.

Further investigations

The ESR is elevated but the white cell count may vary. Radiography of the chest and bacteriological examination of urine must always be done.

On the radiograph, involvement of one vertebra is typical.¹¹⁰ The most common early findings are a diminished disc space and osteolysis of the vertebral body. Later, paravertebral granulation tissue is formed, resulting in paravertebral mass.¹¹¹ This is usually followed by collapse of the vertebral body with formation of an angular kyphosis.

A positive Mantoux test is not diagnostic in itself but does arouse suspicion of tuberculosis.

Treatment

This comprises bed rest and administration of antituberculous drugs. Early surgical intervention with anterior debridement and grafting has been advocated to shorten the course of the disease, and it decreases the risk of neurological complications of deformity.^{102,110}

Intervertebral disc inflammation: discitis

This disorder of unknown cause affects only children, most commonly between 2 and 6 years of age. Whether infection is involved remains uncertain. The lumbar spine is most usually affected.

The first complaint is of back or hip pain; smaller children may just refuse to walk.

On clinical examination, limited spinal movement with muscle spasm and local tenderness is found. Fever is present.

Some weeks after onset, some narrowing of the intervertebral disc space and erosion of the endplates can be seen. Although these radiographic findings are usually sufficient to establish the diagnosis of discitis, MRI is the diagnostic study of choice especially in a differential diagnosis with suspected vertebral osteomyelitis.¹¹² Immobilization of the spine and antibiotics are the main treatment required.

Lateral recess stenosis

In some older patients, the intervertebral disc becomes completely worn out and its fragments displace posteriorly because of physiological kyphosis. As a consequence, the intervertebral space decreases progressively and the posterior longitudinal ligament becomes relatively long. Under the influence of the body weight it bulges posteriorly and may come in contact with the dura mater, causing central backache radiating bilaterally around the trunk, or may compress the nerve root, causing unilateral root pain (Cyriax:⁷ p. 211). This condition is characterized by pain that comes on immediately on standing and, unlike lumbar lateral recess stenosis, is not altered by sitting or stooping. Only when the patient lies down for 1 or 2 minutes does the pain cease, when it stops almost immediately.

Because the condition occurs mainly in an elderly patient with a very stiff vertebral column, no appreciable discomfort can be provoked by articular movements. In root compression, side flexion towards the painful side is occasionally painful.

Treatment

This depends on the site of the pain.

Central pain

Daily traction can be tried. The aim is to bring as much of the disc material back into the intervertebral space as possible, so that on load the intervertebral height diminishes to a lesser degree and compression of the dura does not occur. If successful, it should be continued indefinitely on every other day. If traction fails arthrodesis is called for.

Unilateral root pain

If instead of central pain unilateral root pain is present, an infiltration around the nerve root should be given and can make the inflammation disappear. If necessary, it can be repeated every 6-12 months (for technique, see Ch. 27).

Arthritis of the costovertebral and costotransverse joints

Disorders of these joints that give rise to clinical symptoms are rare. They may be affected by osteoarthritis, ankylosing spondylitis and by rheumatoid arthritis.^{113,114} Degeneration is identified mainly in the costovertebral joints of the first, eleventh and twelfth ribs, all of which have only one articular facet. It is possible that these are more vulnerable to mechanical irritation by continuous rib motion, sometimes as the outcome of a mechanical injury.¹¹⁵ Less frequently the sixth, seventh and eighth costovertebral joints are affected, and in these it is most frequently the inferior hemifacet.¹¹⁶ It has been suggested that patients often have an increased thoracic kyphosis with associated scoliosis.¹¹⁵

Involvement of these joints gives rise to unilateral paravertebral pain radiating along the ribs. The pain is described as aching or burning, is usually worse in the morning and is aggravated by deep inspiration and a cough.^{115–117} Exceptionally, on breathing, a sudden twinge that arrests further inspiration is felt. On clinical examination an asymmetrical pattern is encountered. Neck flexion and scapular movements are negative. Pain is elicited, on performing the springing test of the rib.

When the first costotransverse joint is affected, the patient complains of pain unilaterally at the base of the neck. The pain is provoked by the following movements of the neck: active and passive flexion, contralateral side flexion and resisted side flexion towards the painful side. Pain may also be present on active and passive elevation of the arm and the scapula.

Standard radiography, CT scan and technetium bone scans may show abnormalities of these joints.

Treatment

If only one intercostal joint is affected, an intra-articular injection should be given. Should this be unsuccessful, surgical excision of the joint may be considered.

Warning

There is a considerable risk of pneumothorax from these injections. Extreme care must be taken and should only be performed under radiological control.

Arthritis of the thoracic facet joints

Degeneration of thoracic facet joints most frequently affects levels C7–T1, T3–T5 and T11–L1.^{118,119} The diagnosis should be considered in those patients whose complaint is a unilateral and local paravertebral pain. There are no dural symptoms or signs. The sole clinical finding is pain on one of the rotations, actively and/or passively performed. This can be either the rotation towards or away from the painful side, a situation resembling facet disorders in the cervical area. Other passive movements are far less painful or not painful at all and resisted rotations are painless. Extension pressure on the spinous processes elicits pain on two consecutive processes, indicating the level of the lesion.

These patients are often first mistakenly regarded as suffering from a disc lesion, because they show a partial articular pattern. After manipulation they may obtain some relief but it usually does not last longer than a few hours to a day. Therefore, if there is doubt about the diagnosis, the capsule of the affected facet joint should be infiltrated with 2 ml of lidocaine (lignocaine) 2%, which provides immediate relief in facet joint disorder.

Treatment

Cure is obtained with one or two infiltrations of 1 ml of triamcinolone over 1-2 weeks.

Technique: injection of a thoracic facet joint (Fig. 8)

The patient lies prone with the arms along the body. The two most tender spinous processes are marked and the facet joint between the corresponding vertebrae is identified. Depending on the level, it lies about 1 (upper and lower thorax) to $1\frac{1}{2}$ (mid-thorax) interspinal levels higher than the lower tender spinous process.



Fig 8 • Injection of a thoracic facet joint.

A syringe is filled with 1 ml of triamcinolone and a 4 cm needle is fitted. The needle is inserted vertically downwards about 1 cm from the midline. The needle is usually stopped by the lamina. It is now slightly withdrawn and moved upwards or downwards until the typical capsular resistance is felt before ending at the bone. The steroid is then injected with the needle in bony contact.

The patient must return after 2 weeks. If there is still some pain, the injection is repeated.

If the treatment fails, a blockade with phenol solution of the medial branch of the dorsal ramus of the thoracic spinal nerve can be performed. Good results have also been reported with percutaneous radiofrequency denervation of the thoracic facet.¹²⁰

Paget's disease

This focal, non-generalized osseous disorder is characterized by thickening and deformation of the affected bone and is caused by hyperactive osteoclasts and osteoblasts, which give rise to increased bone resorption together with formation of disorganized new bone. It occurs more commonly than is generally believed and affects about 3% of the population above 40 years of age. This figure rises to about 5–10% in people aged over 80.¹²¹ There is a slight preponderance of males and the condition is most frequently encountered in Western Europe, Australia and New Zealand.

Flat bones and the ends of the long bones are mainly affected. Most frequently involved are the spine, skull, sacrum, pelvis and femurs. About 60% of patients with Paget's disease have involvement of the lumbar spine, and 45% of the thoracic spine and sacrum. In only 15% is the cervical spine involved.¹²²

Clinical presentation

Only about 12% of the patients affected by Paget's disease suffer from backache from the disease as such.¹²³ It should be recognized that other disorders, such as disc protrusions and facet joint problems, may be the basis of the pain in patients who are identified as having Paget's disease. In two-thirds the disease remains clinically asymptomatic.¹²⁴ These cases are

usually detected incidentally on a radiograph taken for some other reason or by an increased serum alkaline phosphatase level.

Paget's disease usually starts in the middle aged or elderly and may give rise to local pain, local heat and deformation. The complaint is usually of a dull pain that does not increase at night. Characteristic deformities – lateral bowing of the femur, anterior bowing of the tibia, increased size of the skull and decreased length of the spine – may occur. Osteoporosis and fractures from minimal trauma are often encountered and usually present in the femur and tibia, less often in the spine. The typical Paget's stature with dorsal kyphosis may be seen. Involvement of the spine occasionally results in spinal stenosis.

Complications

Because of multiple arteriovenous fistulae in bone, there is lowered peripheral resistance with a rise in cardiac output, which may lead to cardiac decompensation. Enlargement of the skull, with secondary compression of peripheral nerves, may result in hearing loss, tinnitus and vertigo.¹

Pagetoid bone may convert to a highly malignant sarcoma with very poor prognosis. The probability is higher when there is polyostotic involvement.

Radiology

Radiological findings depend on the stage. The initial phase is characterized by lytic bone lesions: osteoporosis circumscripta. These are rarely seen in the spine. Later, a mixed stage develops in which both demineralization and sclerosis of bone exist. The final sclerotic stage is characterized by a homogeneous increase of bone density.

In the vertebrae, coarse trabeculae and a marked edge of the vertebral body ('picture frame' appearance) are present. The central portion appears decalcified. Ultimately, the bone becomes dense. When diagnostic doubt exists, bone scans must be taken. They are more sensitive than plain radiographs.

Laboratory

Serum calcium and phosphorus concentrations are within normal limits, but the serum alkaline phosphatase is usually very high.

Treatment

Most patients are asymptomatic and do not need any treatment. More active disease may require calcitonin or bisphosphonate to reduce the symptoms but none of them is curative.

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