A pain in the neck

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A previously healthy and independent 84-year-old woman was referred to the neurology department with leg weakness. Nine days earlier she had experienced an acute onset of severe neck pain whilst sitting at rest, which radiated down both arms. Although initially feeling unable to move, she attended a casualty department where analgesics and diazepam were prescribed and she was sent home. However, on leaving the hospital her legs felt heavy and she required the assistance of two people.

At home the next day she was unsteady on her feet and fell. Over the ensuing 4 days she developed leg cramps, urinary retention and constipation, necessitating her admission to hospital. On neurological examination, cranial nerves were intact but there was a quadriparesis mainly affecting the distal arm muscles and leg flexors with grade 3–4/5 power in the wrist extensors and finger abductors, 4/5 power in quadriceps and 3/5 strength in the hamstrings. Plantar responses were bilaterally extensor. Neck movements were markedly restricted by pain. Because of the nature of the history and the distribution of clinical signs, magnetic resonance imaging (MRI) of the spine was undertaken (figure).

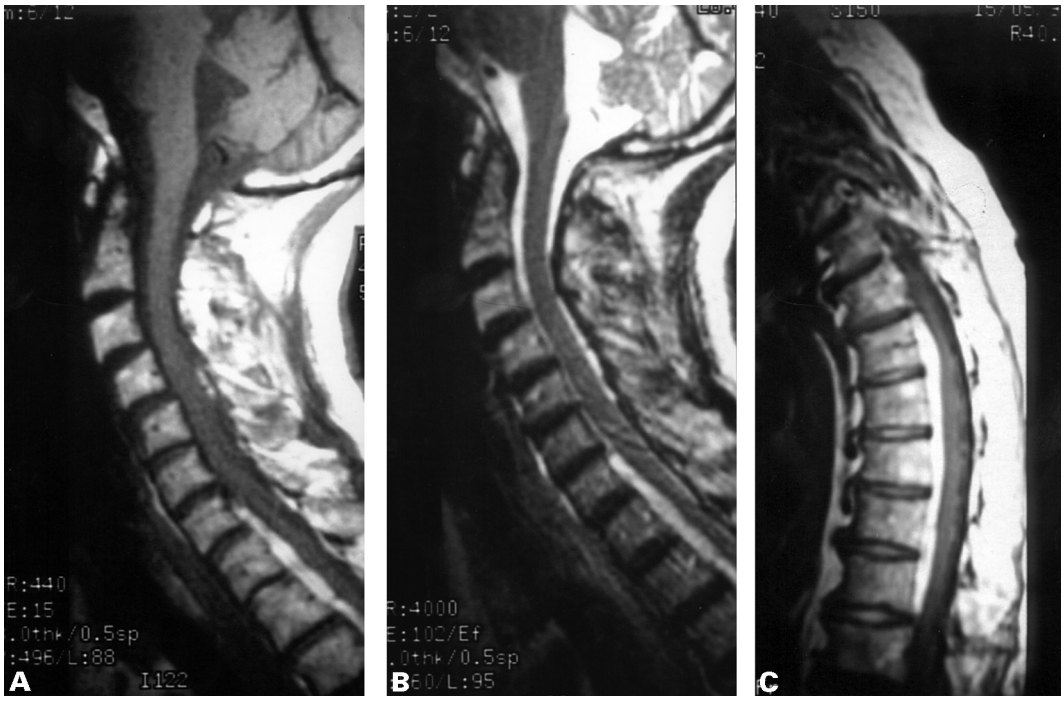

Figure MRI images of the spine. (A) Sagittal T1-weighted image of the cervical spine (TR 440 ms, TE 15 ms), (B) sagittal T2-weighted image of the cervical spine (TR 4000 ms, TE 102 ms); (C) sagittal T1-weighted image of the dorsal spine (TR 440 ms, TE 15 ms)

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Questions

1 What is the differential diagnosis of the spastic quadriparesis in this patient?
2 What do the MRI scans show?
3 What is the diagnosis?
4 What further elements of the history are relevant and what other investigations are indicated?
5 How should the patient be managed?

Answers

QUESTION 1

The differential diagnosis of spastic quadriparesis is obviously wide, as indicated by the non-exhaustive list in box 1. However, the acute nature of the onset in this patient narrows the list considerably, with a vascular aetiology and an acute central cervical disc herniation being most likely, given the lack of any obvious spinal cord trauma. The absence of a prodromal illness or systemic upset weighs heavily against an inflammatory aetiology whilst demyelinating disease and hereditary spastic paraplegia would be unlikely to present for the first time in this late age group.

Although they would in any case have a more chronic course, syringomyelia would most commonly manifest with lower motor neurone signs in the arms and a dissociated sensory loss whereas motor neurone disease does not in general present with a 'pure' spastic quadriparesis. HTLV-1 mediated tropical spastic paraparesis usually spares the upper limbs and is rare in Caucasian populations.

Differential diagnosis of spastic quadriparesis

- vascular: anterior spinal artery occlusion, aortic dissection, arteriovenous malformation, epidural or subdural haematoma
- degenerative: central spondylosis, central cervical disc herniation, syringomyelia, Paget's disease
- neoplastic: intramedullary (glioma, ependymoma, lymphoma); extramedullary (neurofibroma, meningioma, ependymoma, bronchial and breast metastases, myeloma)
- inflammatory: viral, bacterial, fungal, parasitic, granulomatous, post-infectious, post-vaccine
- demyelinating: multiple sclerosis
- metabolic: subacute combined degeneration of the cord
- hereditary: hereditary spastic paraplegia
- other: motor neurone disease, HTLV-1 mediated tropical spastic paraparesis, parasagittal meningioma

Box 1

QUESTION 2

Figure (A) demonstrates high signal present anterior to the cord on T1-weighted images of the cervical spine from the level of C7. The T2-weighted image in figure (B) shows a similar area of high signal intensity arising from the same area. Figure (C) indicates that this area of high signal intensity also extends down the anterior surface of the dorsal cord and indeed reaches as far as the conus (not visible).

QUESTION 3

The MRI appearances are typical of a subacute haematoma in the epidural space. Within 24 hours of onset a haematoma is usually isointense with the cord on T1-weighted images but with the development of methaemoglobin in the haematoma it becomes of high signal intensity on both T1- and T2-weighted images. The thin rim of low signal intensity just visible in the figures separating the haematoma from the cord is dura mater.

QUESTION 4

Although spinal epidural haematoma is 'spontaneous' with no obvious precipitating factors in over 50% of cases, certain conditions and triggers are believed to be associated. These include: bleeding diatheses associated with haematological disorders and with the use of anticoagulants and thrombolytics; spinal trauma including surgery and spinal puncture; tumours; hypertension, and arteriovenous malformations. Rarer associations have been reported with rheumatological conditions, alcohol, pertussis and pregnancy. Further questioning and investigations should therefore be directed appropriately. In this instance there had been no history of anticoagulant therapy, trauma or hypertension and there was no evidence of a tumour or vascular malformation on MRI. Clotting profile and platelet aggregation studies were normal.

QUESTION 5

The options in the management of this condition include surgical intervention, involving laminectomy and removal of the clot, or a conservative approach. In view of the extensive nature of the lesion and the advanced age of the patient, the latter approach was adopted. There was no further clinical deterioration and within 72 hours a slow clinical improvement was becoming apparent. Within 8 weeks there had been a full neurological recovery with return of sphincter function and the patient was living independently once more.

Discussion

Spinal epidural haematoma is an uncommon and rarely reported clinical entity which does, however, represent a neurological emergency. Although a painless onset has been described, this is exceptional and, as in this case, the disease is typically characterised by sudden onset of neck or back pain followed by sensory and motor dysfunction. The full symptom complex may evolve rapidly within one hour or may take weeks or even months to develop fully (box 2).

Early diagnosis may be difficult since a combination of interscapular and radicular pain can mimic an aortic dissection and radicular pain may be difficult to differentiate from pain due to a cardiac or pulmonary event. Acute

Typical presentation of spinal epidural haematoma

- neck/back pain
- radicular pain
- sensory disturbance
- motor weakness
- sphincter disturbance

Box 2

onset of pain in the upper neck may also raise suspicion of subarachnoid haemorrhage. However, bilateral symptoms of myelopathy in connection with neck or interscapular pain radiating into the upper extremities should lead to suspicion of cervical pathology including disc prolapse, spinal tumour or metastasis and epidural haematoma.

Spinal epidural haematomas occur in all age groups but are more common in middle or later life and are more frequent in males than females. Usually the haematoma lies in the dorsal epidural space but the anterior location demonstrated in this patient has been previously described.[1] The most frequent levels of involvement are the cervical and thoracic segments with thoracolumbar and lumbar regions being less commonly affected.[2] The extent of the lesion in this patient is unusual since most epidural lesions involve just 2–4 segments although a haematoma extending over 11 segments has been reported.[3]

Although no underlying cause for spinal epidural haematoma can be determined in at least 50% of cases,[4] it is well recognised that predisposing factors include trauma, bleeding diatheses and anticoagulant therapy (box 3). Cases have also been reported as occurring in connection with apparently routine procedures such as epidural anaesthesia and diagnostic lumbar puncture.[5] The significance of factors such as hypertension and underlying vascular malformations remains more controversial since it has been noted that hypertension is no more prevalent in patients with spinal epidural haematoma than in age-matched controls,[4] and vascular malformations have been associated in less than 5% of cases at most.[6]

The MRI characteristics of spinal epidural haematoma are quite specific.[7] On sagittal sections it is clearly outlined with tapering superior and inferior margins. Dura mater is seen as a curvilinear low signal separating the haematoma from the cord. Within 24 hours of onset the haematoma is isointense with the cord on T1-weighted images and heterogenous on T2-weighted images. However, methaemoglobin accumulates after 24 hours causing T1 shortening and an increase in the signal on T1-weighted images. The differential diagnosis of such appearances should include spinal subdural haematoma, epidural neoplasm and abscess.

Conditions associated with spinal epidural haematoma

- bleeding diatheses
- drugs: aspirin; anticoagulants; thrombolytics
- spinal surgery, trauma, puncture
- spinal tumours
- arteriovenous malformations
- rheumatological disorders
- pregnancy
- alcohol

Box 3

The spinal subdural haematoma is far less frequent than the epidural haematoma and is rarely spontaneous, usually occurring in relation to anticoagulant therapy, blood dyscrasias or spinal trauma. MRI does not show a low signal dural rim. Epidural metastases give low signal or are isointense with the cord, with irregular margins and without tapering ends. These are generally associated with vertebral infiltration, which appears as diffuse or focal areas of homogenous low signal on T1-weighted images. Epidural abscesses are fusiform and contiguous with an infected disc and vertebral bodies. They are centred on the disc and in most cases give lower signal than the spinal cord on MRI. Both metastases and abscesses usually also show enhancement on T1-weighted images post gadolinium.

In general, surgical intervention in cases of spinal epidural haematoma has been considered mandatory. Indeed it has been found that delay of more than 36 hours before decompressive laminectomy and evacuation of clot is associated with a poor prognosis.[8] However in recent years there have been several reported instances of spontaneous recovery following conservative management in patients with mild or non-progressive neurological deficits.[9] The outcome in this case lends further support to these observations.

In conclusion, although the differential diagnosis of spastic quadriparesis is wide, this case emphasises the importance of suspecting a diagnosis of spinal epidural haematoma in any patient presenting with acute onset of severe neck or back pain when this is associated with radicular symptoms or neurological signs consistent with cord compression. Predisposing factors should be sought and urgent surgical intervention should be considered although the potential for spontaneous recovery does exist in certain cases.

Summary points

- spinal epidural haematoma is a neurological emergency which occurs most commonly in middle life or later and is more common in males
- MRI is the diagnostic tool of choice
- cervical and upper thoracic spinal segments are most commonly involved
- about 50% of cases occur spontaneously
- bleeding/clotting disorders should be excluded
- urgent decompressive laminectomy and evacuation of clot is indicated in patients with significant or progressive deficits
- conservative management may be appropriate in mild and non-progressive cases and culminate in spontaneous recovery

Box 4

Final diagnosis

Spontaneous spinal epidural haematoma.

Keywords: epidural haematoma; MRI; spine

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Images in medicine

Epidural lipomatosis

A 51-year-old man presented with radicular-like pain in the left lower limb and intermittent claudication lasting for 1 year. Pain in the legs was evoked by standing or walking more than 300 meters and rapidly relieved by sitting. Clinical examination only disclosed significant overweight (weight 100 kg; height 163 cm; BMI: 37.6 kg/m^2). Electromyography showed chronic neurogenic abnormalities in the muscles supplied by the left L5 nerve root. Magnetic resonance imaging (MRI) of the lumbar spine disclosed cauda equina compression by epidural lipomatosis (see figure on opposite page, A and B).

After 3 months on a hypocaloric diet leading to 20 kg weight loss, pain and claudication had completely resolved. MRI showed a dramatic regression of the epidural lipomatosis (figure, C and D). In the differential diagnosis of lumbar stenosis, the clinician must keep in mind the possibility of epidural lipomatosis, even in patients without systemic glucocorticoid therapy or Cushing's disease.

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