SELF ASSESSMENT ANSWERS

Back pain and systemic compromise

Q1: What is the most likely clinical diagnosis?

The triad of back pain/tenderness, neurological deficits, and systemic illness are highly suggestive of a spinal epidural abscess.

Q2: What does the MRI scan show?

The MRI scan (see p 373) shows a large central disc prolapse at the L4/L5 level. There is also, however, loss of cerebrospinal fluid signal behind the dura from the first lumbar vertebral level caudally, suggestive of a compressive lesion.

Q3: Discuss the management of this condition

Urgent laminectomy was performed. At operation free pus was found in the muscular and fascial layers. There was a large epidural abscess which had caused severe compression of the lumbar thecal sac posteriorly. The epidural space was debrided and irrigated. Cultures of the specimens as well as blood cultures revealed a staphylococcal infection. The patient was started on high dose intravenous flucloxacillin, metronidazole, and gentamicin.

Postoperatively, he was improved neurologically with near normal power in the lower limbs bilaterally and normal sensation including an improvement in his perianal sensation. His subsequent recovery, however, was complicated by septicaemia, acute respiratory distress syndrome, and disseminated intravascular coagulation, which were successfully treated.

Discussion

The risks of developing an abscess in the spine are greater in those with diabetes mellitus, intravenous drug abuse, tuberculosis, malnutrition, chronic renal failure, and cancer.[1]

Making the diagnosis of an epidural abscess can be difficult. Reliance on imaging alone may be misleading since the radiological changes, as in this case, may be subtle. Furthermore, the condition may be masked by other more common pathologies. In this case, there was a large L4/L5 central disc prolapse, although the neurological deficits were far more extensive than that expected from such a disc prolapse. It is therefore important to pay careful attention to the clinical findings.

Cardinal features of spinal epidural abscess are fever, spinal tenderness, and neurological deficit. Pain is the most consistent symptom and together with fever often precedes the development of hard neurological signs.[2] This natural history contrasts markedly to that of the acute or chronic degenerative pathologies of the spine which tend not to exhibit local tenderness or systemic compromise. Most patients are thought to have major neurological signs prior to surgery.[2] When septicaemia dominates the picture as in this case, the neurological symptoms may be missed. This is especially true for those patients who may have been confined to bed for some time and therefore not regularly assessed.[2]

Management includes surgical decompression, debridement, and broad spectrum antimicrobials. Parenteral treatment should be continued for at least four weeks and may be needed for eight weeks if osteomyelitis is suspected.[3 4] Prognosis is related to the delay in presentation.[2 4] Patients who present with frank septicaemia or those with gross neurological signs do poorly.[4]

Conclusion

Spinal epidural abscess remains a clinical diagnosis. A high index of suspicion and rapid neurosurgical attention are essential to minimise mortality and long term morbidity.

Final diagnosis

Lumbar spinal epidural abscess.

References

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A bad dose of 'flu

Q1: What is the most likely diagnosis and how would you confirm it?

The most likely diagnosis is one of acute rhabdomyolysis together with myoglobinuria, as demonstrated by the generalised oedema, muscle tenderness and weakness, and the brown discoloration of the urine. To confirm the diagnosis, the urine should be tested for myoglobin (radioimmunoassay is the best technique) and muscle enzymes should be measured.

In this patient, the urine was positive for myoglobin and had an acidic pH of 6.0. Muscle enzymes were grossly raised, with creatine kinase 30 000 U/l, lactate dehydrogenase 2900 U/l, and aspartate aminotransferase 710 U/l.

The differential diagnosis of a patient becoming weak on a background of such a prodromal illness as that described here would normally include an acute inflammatory demyelinating polyneuropathy (or Guillain-Barré syndrome), but the presence of such oedema and muscle tenderness makes this an unlikely diagnosis even prior to the biochemical results.

A patient presenting in such a manner in the postoperative period would lead to suspicion of a diagnosis of malignant hyperthermia. This is a rare autosomal dominant condition, linked to a mutation in the ryanodine receptor gene on chromosome 19,[1] in which susceptible individuals are endangered by exposure to certain anaesthetic triggering agents (see box 1).

Q2: What is the possible aetiology of this condition?

The potential aetiology of rhabdomyolysis is wide (see box 1). In this particular patient there would appear to be three main possibilities. Firstly given the nature of the prodromal symptoms, any one of a range of infectious agents could be implicated (see box 2). Secondly, rhabdomyolysis is well recognised as a complication of certain types of drug therapy and this patient had been exposed to several agents over the 48 hours immediately before admission, although not before the time that he first became unwell. Thirdly, the possibility of a primary muscle disease should always be considered (many would not necessarily be relevant in this particular age group), although such an acute, de novo presentation would be unusual.

In this particular patient, the aetiological agent was a virus, influenza type B, serological testing indicating a rising antibody titre to 1 in 320.

Box 1: Aetiology of rhabdomyolysis

- Trauma.
- Seizures.
- Ischaemia.
- Metabolic defects: glycogenoses, carnitine palmitoyltransferase deficiency.
- Drugs: clofibrate, gemfibrozil, epsilon-aminocaproic acid, statins, etretinate, high dose steroids.
- Alcohol.
- Infectious diseases.
- Malignant hyperpyrexia: halothane, enflurane, isoflurane, succinylcholine, calcium channel blockers.
- Malignant neuroleptic syndrome.
- Electrolyte imbalance: hypokalaemia, hypomagnesaemia, hypophosphataemia.

Q3: What potential complications may occur?

There are several potential complications. Massive rhabdomyolysis may result in electrolyte imbalance, particularly hyperkalaemia, hyperphosphataemia, and hypercalcaemia. Disseminated intravascular coagulation and venous thromboses may also occur. Most seriously, severe myoglobinuria may cause renal damage and anuria. The exact mechanism by which this occurs is uncertain but hypotheses include renal tubular obstruction by precipitated myoglobin, reduction in renal blood flow, and direct toxic injury to the tubular epithelium.

Q4: How would you manage this patient?

In general, good nursing and medical care with close monitoring of electrolyte balance, renal function, and urinary output are essential. More specifically, a high fluid intake and alkalinisation of the urine by infusion or ingestion of sodium bicarbonate helps to protect the kidney by preventing the formation of myoglobin casts. This patient received 3 g of sodium bicarbonate, every two hours, by intravenous infusion, until the urine pH was >7.0. He was also treated with 2 mg/kg of dantrolene by intravenous infusion, daily for five days.

Box 2: Infectious agents implicated in rhabdomyolysis

- Influenza A and B.
- Coxsackie virus.
- Epstein-Barr virus.
- Cytomegalovirus.
- Echovirus.
- Adenovirus.
- *Legionella pneumophila*.
- *Streptococcus pneumoniae*.
- HIV.

Box 3: Learning points

- Acute rhabdomyolysis represents a medical and neurological emergency.
- Rhabdomyolysis may complicate even usually innocent infections.
- Maintenance of high fluid intake and alkalinisation of urine are fundamental to preserving renal function.
- Intravenous dantrolene is a useful therapeutic option.
- Patients should be monitored for the possible development of a compartment syndrome and these should be managed appropriately.

Discussion

In the period immediately after admission, the patient continued to deteriorate. There was progression in muscle weakness with lower limb muscle strength ranging between 0/5 at knee flexion and 3/5 at knee extension. In the upper limb, weakness was less marked, muscle strength generally 3–4/5. He also developed decreased sensation on the dorsum of the right foot and increased tension in the anterior tibial compartments of both legs. Consideration at this stage was given to surgical release.

The creatine kinase level continued to rise, peaking at 140 000 U/l 48 hours after admission, before declining rapidly thereafter. Electromyography and nerve conduction studies showed clear myopathic changes in all sampled muscles with evidence of moderate bilateral carpal tunnel compression and moderate-severe right peroneal and tibial nerve lesions. The neuropathy was attributed to oedema and compression.

In the days after the decline in creatine kinase, the patient's condition stabilised and then gradually improved. Four weeks after the onset of illness he was discharged home, mobilising with the aid of a Zimmer frame. At the most recent follow up several months later, he was mobilising independently, had full return of upper limb strength, and had full power in the lower limb, apart from the right ankle where dorsiflexion and plantar flexion were 4/5.

Infectious agents are believed to be implicated in only 5% of cases of rhabdomyolysis.[2] The association with influenza infection, although uncommon, is well described in the literature. A recent review listed 22 cases of rhabdomyolysis associated with generalised, non-pneumonic influenza A infection.[3] Of these, 12 developed acute renal failure (55%) and four of these died. Influenza type B is much less commonly implicated in rhabdomyolysis. There has been a report of two cases of influenza B in elderly men being associated with rhabdomyolysis, but each of these occurred in the context of hyperpyrexia and the clinical course was much milder with smaller enzyme rises than that documented in our patient.[4]

The mechanism by which viruses can cause rhabdomyolysis is unclear, although at least two theories have been advanced.[5] One theory is that there is direct viral invasion into muscle fibres causing myonecrosis. This is supported by muscle biopsy findings of lymphocytic infiltrates, viral inclusions, and viral DNA identified by polymerase chain reaction. Conversely, where muscle biopsy is either normal or shows hyaline degeneration and myonecrosis but no viral particles, it is postulated that the virus releases a circulating toxin or stimulates cytokine release. To date however, no putative toxins have been identified.

Dantrolene has been used to beneficial effect in rhabdomyolysis arising from malignant hyperpyrexia, malignant neuroleptic syndrome, exertion, acute alcoholic intoxication, and both cocaine and ecstasy overdose. It is known to decrease the release of calcium from the sarcoplasmic reticulum.[6] Calcium may play a part in muscle pain as has been suggested in McArdle's disease and in the development of malignant hyperthermia. Muscle cell necrosis is also explained by an increased calcium influx into the cell, triggering a vicious cycle of mitochondrial overloading and energy depletion, which leads to hypercontraction and finally, to cell death.

Experiments with dantrolene in cases of exertional rhabdomyolysis have shown an 83% reduction in intracellular calcium levels, which has been associated with marked improvements in both clinical symptoms (muscle stiffness, rigidity and pain) and laboratory values (83% reduction in creatine kinase by day 4).[7]

This case demonstrates the potentially acute course of rhabdomyolysis in even healthy individuals, highlights the potential complications arising from this condition and emphasises the routine but effective methods of avoiding these.

Final diagnosis

Acute rhabdomyolysis with myoglobinuria.

References

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A case of acute swelling of the left shoulder

Q1: What disease process are the *x* ray and MRI findings diagnostic of?

The differential diagnosis of an acute monoarthritis includes a pyogenic infection of the joint. Tuberculosis should also be considered in an individual who is PPD positive even with a normal chest radiograph. The resorption of the humeral head with a sharp line of demarcation, almost as if surgically excised, is diagnostic of this disease. Shards of bone can sometimes be identified in the soft tissue and was noted in a computed tomogram and MRI of the shoulder (not shown). This picture is very characteristic of the atrophic form of a neuropathic arthropathy of the shoulder[1] with the most common cause being syringomyelia. The MRI (fig 2; see p 374) confirms the diagnosis of syringomyelia with a large syrinx in the spinal cord and all cultures were negative.

Box 1: Causes of osteolysis

Trauma
- Reflex sympathetic dystrophy.

Skin
- Psoriasis.

Infection
- Direct.
- Indirect—for example, leprosy.

Tumour
- Plasmacytoma.
- Lytic metastases.

Collagen vascular diseases
- Rheumatoid arthritis, usually the lateral end of the clavicle.
- Scleroderma.

Endocrine
- Primary hyperparathyroidism.
- Secondary hyperparathyroidism.

Chronic renal failure

Neurology
- Neuroarthropathy.

Q2: What are the other causes of osteolysis?

The causes of osteolysis are shown in box 1.

Q3: What are the other clinical features of this disease?

Syringomyelia can be associated with Chiari I and II malformations, Dandy-Walker malformation, and basilar invagination. Frequently, the signs and symptoms in children and adolescents may include scoliosis, dissociated sensory loss, neck pain, vomiting, and lower motor neuron signs in the upper limbs. In adults, the clinical presentation is more variable, but usually point towards dysfunction of the cervical cord. It is worth noting that the classic "dissociated sensory loss" is present in only 49% of patients with a syrinx.[2] More commonly, lower motor neuron signs are seen in the upper limbs and long tract signs are seen in the lower limbs. Other findings include dizziness, nystagmus, scoliosis, and even brain stem symptoms like dysphagia, facial numbness, and vertigo when there is extension of the syrinx into the medulla.

Discussion

Syringomyelia is a cavitation within the spinal cord appearing in the third to fourth decade. Familial cases have also been described.[3] The Arnold-Chiari malformation with herniation of the cerebellar tonsil is found in more than two thirds of patients. It may also be a late consequence of spinal cord trauma, with delayed onset observed in 5% leading to an ascending spinal syndrome. As a result of arachnoiditis, cerebrospinal fluid circulation is impaired. The syrinx mainly in the lower cervical region interrupts decussating spinothalamic fibres leading to a loss of pain and temperature in a shawl-like distribution with preservation of light touch, vibration, and proprioception. Painless ulcers of the fingers may be a presenting feature. Extension of the syrinx into the anterior horn results in a loss of motor neurons and an amyotrophy. It begins in the small muscles of the hand with asymmetric weakness and early loss of muscle stretch reflexes in arms. Extension into the lateral columns results in lower extremity

Key learning points

- Recognise the radiographic appearance of osteolysis and know its causes.
- The most common cause of a neuropathic joint of the shoulder is syringomyelia.
- Resorption of the head of the humerus is a characteristic radiographic finding of syringomyelia.

spasticity, paraparesis, brisk tendon reflexes, and a positive Babinski sign.

Patients may present with an acute enlargement of the shoulder with destruction of the head of the humerus. Remarkable subcutaneous oedema of the hands referred to as "main succulente" has been described. Interruption of the central autonomic pathways results in hyperhydrosis. Resorption of the head of the humerus with a well defined margin is a characteristic radiographic finding of syringomyelia.

Final diagnosis

Syringomyelia with resorption of the humeral head representing a neuroarthropathy

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Abdominal pain in a diabetic myeloma patient with cirrhosis

Q1: What is the differential diagnosis?

Multiple myeloma (with low concentrations of uninvolved immunoglobulin concentrations), diabetes, cirrhosis, and hospitalisation constitute major risk factors for severe infections in this patient. Spontaneous bacterial peritonitis must always be considered for this kind of cirrhotic patient as well as the possibility of a urinary tract infection, because of the marked pyuria and multiple risk factors. Although these two conditions are the most probable, all other causes of lower abdominal pain—for example, intra-abdominal abscess, perforated viscus, mesenteric vascular accident, and diverticulitis—must be excluded, especially in this uncooperative patient.

Q2: What abnormalities are seen on the radiographs?

The *x* ray film obtained after contrast enhanced computed tomography, while the patient was excreting contrast material, shows the borders of the urinary bladder and gas bubbles within its wall clearly as a radiolucent line (fig 1; see p 375). The extent of gas collection can be better appreciated in the computed tomogram (fig 2; see p 375). The source of this gas within the urinary tract may arise from infection, penetrating trauma, gastrointestinal fistulas or iatrogenic causes, such as diagnostic or surgical instrumentation.

Our patient gave no history of trauma, and no urinary instrumentation had been performed previously. Fistulous tracts, abscess, and mesenteric occlusion can be excluded on computed tomography. The patient's risk factors, marked pyuria, foul smelling urine, and predominant gas collection in the bladder wall make a urinary tract infection most likely.

The final diagnosis was emphysematous cystitis. This is a rare form of urinary tract infection in which fermentation of glucose by bacteria causes carbon dioxide production in the bladder wall, which is seen on a plain film as a radiolucency confined to the bladder wall. Gas bubbles collect in the submucosa and eventually rupture, resulting in gas within the bladder lumen.[1] Computed tomography is a very sensitive tool for demonstrating the gas within the bladder wall and the extent and location of the gas collection.

Patients may complain of lower abdominal pain, dysuria, and pneumaturia or may have no symptoms. Likewise, severity of the illness ranges from an asymptomatic condition to life threatening cystitis.

Q3: What are the predisposing factors?

More than 50% of patients with emphysematous cystitis have diabetes mellitus. Patients with bladder outlet obstruction, neurogenic bladder, and recurrent urinary tract infections are at increased risk. Immunocompromised and debilitated patients are especially susceptible. Females are two times more likely to be affected than males.[2] The condition most commonly results from infection with *Escherichia coli* but proteus, klebsiella, staphylococcus, streptococcus, nocardia, and clostridium have also been described in the literature.[3] Long term broad spectrum antibiotic therapy and indwelling Foley catheters constitute another risk group for candida related emphysematous cystitis.[4]

Q4: How would you treat this condition?

Early diagnosis, strict control of the blood glucose level in diabetic patients, immediate antibiotic therapy, and adequate continuous bladder drainage are the general treatment principles. Patients must be admitted to hospital for observation and proper intravenous antibiotic therapy. After successful elimination of infection, radiographic and clinical resolution usually occurs within a few days, because carbon dioxide is readily absorbed in human tissue.[3 5]

It is important to differentiate emphysematous cystitis from emphysematous pyelonephritis, in which gas involves the renal parenchyma, since the latter has a mortality rate of about 40% and generally requires nephrectomy. In contrast surgical intervention is rarely needed in emphysematous cystitis except when an anatomical abnormality like an obstruction or stone is present.[3]

Final diagnosis

Emphysematous cystitis.

References

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An eponymous reaction to a knife wound

Q1: Where is the anatomical site of injury and which spinal tracts have been damaged?

A left sided hemisection of the spinal cord at T8 plus bilateral posterior column loss. This is due to the knife track coming obliquely from the right, across both posterior columns of the spinal cord before hemisecting the left side of the cord (see figs 1 and 2). The left sided tracts transected include the corticospinal tract, dorsal column, and spinothalamic tract.

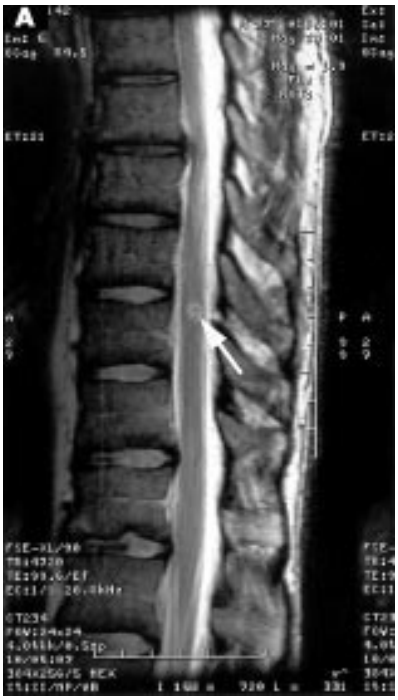

Figure 1 T2 weighted sagittal MRI image of the thoracic cord showing a mixed signal abnormality (arrowed) slightly to the left of the cord at D10, representing the knife track within the cord.

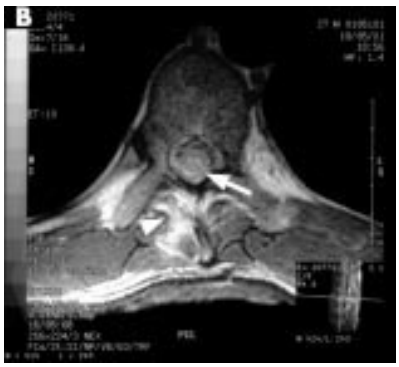

Figure 2 T2 weighted axial MRI image of the thoracic cord showing an area of high signal posteriorly and to the right of the cord, consistent with the track of the knife through the soft tissues (arrowhead). The lesion within the cord is seen again slightly to the left of the midline (arrowed).

Q2: What is the name of this syndrome?

Brown-Séquard syndrome.

Q3: Give three other causes of this syndrome

Other causes of Brown-Séquard syndrome include multiple sclerosis, unilateral disc herniation, extrinsic cord lesions—for example, metastases, epidural haematomata, and unilateral ischaemic lesions of the cord.

Q4: What is the cause of his headache?

The headache is a classical presentation of low pressure headache due to leakage of cerebrospinal fluid from a dural tear.

Q5: How would you treat his headache if the symptoms persisted?

The recommended management for persistent low pressure headache in the context of a stab wound of this nature would be surgical exploration and repair of the dural defect with increased fluid intake. A blood patch or oral caffeine treatment could also be considered.

Discussion

The neurological injury is consistent with left sided Brown-Séquard syndrome.[1] One of the points of interest in this case is the bilateral posterior column loss and the left sided Brown-Séquard syndrome, although the point of knife entry was right sided. The explanation is due to the oblique knife injury with the track of the blade crossing the posterior columns from the right and impinging in the left side of the cord (figs 1 and 2). A classical presentation for Brown-Séquard is ipsilateral loss of the corticospinal, posterior column, sympathetic, and spinothalamic tracts.[1] Clinically this results in ipsilateral pyramidal deficit with ipsilateral loss of joint position, vibration, and soft touch at the level of the lesion. There is contralateral loss of pain and temperature sensation that manifests itself a few segments below the level of the lesion because the decussating fibres enter the spinothalamic tract a few segments rostrally to the level of entry of the nerve root.

Stab wounds are a common cause of Brown-Séquard syndrome[2] with rarer causes including primary or secondary cord tumours, degenerative disc disease, cord ischaemia, inflammatory or infectious conditions—for example, herpetic infections or multiple sclerosis and subdural/epidural haemorrhage (reviewed in Peacock *et al*[3]).

The low pressure headache was as a result of continuing cerebrospinal fluid leakage from a dural tear. Low pressure headaches of this nature are also reported after some lumbar punctures and can also occur spontaneously. The classical features are a positional headache that is worse on sitting up; nausea and photophobia may also occur.[4] Headaches of this nature may respond symptomatically to increased fluid intake and the use of caffeine compounds. Occasionally epidural blood patches may be useful in plugging the cerebrospinal fluid leak.[4]

In this case, the patient's low pressure headache resolved spontaneously without the need for surgical exploration of his wound.

Final diagnosis

Brown-Séquard syndrome.

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