SELF ASSESSMENT ANSWERS

A woman with painless burns

Q1: What is the likely diagnosis?
Syringomyelia.

Q2: What is the differential diagnosis?
Cervical spondylosis, multiple sclerosis, and motor neurone disease (see discussion below).

Q3: What relevant investigations would you request?
Routine blood analysis including erythrocyte sedimentation rate, thyroid function tests, and B12; lateral cervical spine radiography and magnetic resonance imaging (MRI) of cervical spine.

This woman's MRI scans (see figs 1 and 2) revealed a dilated syrinx extending from C2–T3 with characteristic septation in the lower cervical region. There was no evidence of the Arnold-Chiari malformation or a cord neoplasm (see discussion below).

Discussion

Syringomyelia is a rare, chronic, progressive disorder in which tubular cavities form at or close to the central canal of the cervical spinal cord. It typically presents in young adults with a mean age of onset of 30 years.

The condition rarely occurs in isolation. Over two thirds of cases are associated with the Arnold-Chiari malformation in which the cerebellar tonsils herniate through the foramen magnum of the skull. In addition, a syrinx may develop within or in close proximity to an intramedullary tumour, generally a glioma[1]. Also, cavities in the cord may also develop as a late consequence of cord trauma.

Symptoms depend primarily on the location of the syrinx, but the latter is most commonly located in the lower cervical region. As the syrinx enlarges, it compresses the following structures in the cord:

- Decussating spinothalamic fibres—results in loss of pain and temperature in a "cape" distribution (with preservation of light touch/vibration sense, that is, dissociated sensory loss) as well as pain radiating from the shoulders down dermatomes which is typically exacerbated by coughing/sneezing; the subsequent impairment of pain sense results in Charot (neuropathic) joints evolving typically at the elbow or wrist.

- Anterior horn cells—results in wasting and weakness of the small muscles in the hand, spreading proximally.

- Decussating corticospinal tract fibres—results in a spastic paraparesis in the lower limbs with extensor plantar responses.

- Cervical sympathetics—results in a Horner's syndrome and/or disturbances in sweating (hyper or hypohydrosis).

As the syrinx is usually asymmetrically placed, manifestations in the arms and hands tend to be similarly asymmetric. It is worth noting that sphincter function is typically preserved until very late on in the disease process.

The syrinx may ascend into the medulla. So called syringobulbia is evidenced by dysphagia, palatal weakness, nystagmus, asymmetric weakness of the tongue with atrophy, and dissociated sensory loss in the distribution of the trigeminal nerve.

Important differential diagnoses to be excluded include cervical spondylosis, multiple sclerosis, and motor neurone disease; they may all cause a spastic paraparesis with upper limb neurological signs, but there are important differences. Cervical spondylosis is common, but not within young adult populations. It may be impossible to exclude on neurological examination and of course may actually coexist with a syrinx! Important clues are reduced range of neck movements and neck pain. Multiple sclerosis does present in this age group, but there will be no dissemination in time and location of the central nervous system symptoms/signs and early wasting of the hand muscles is not a feature of multiple sclerosis. Motor neurone disease may cause weakness, wasting, and loss of reflexes in the upper limb but patients tend to present in middle age, there are no sensory signs, and early fasciculations of the upper limb muscles is an early sign.

Another differential to exclude is that of an intrinsic spinal cord neoplasm. As mentioned above, this may coexist with a syrinx. From the discussion below it is noted that the tumour may actually be involved in the pathogenesis of syringomyelia. Thus gadolinium enhanced MRI is an essential investigation in the work-up of such patients in order to exclude an intrinsic cord neoplasm.

The pathogenesis of syringomyelia is uncertain, but various theories exist.[2 3] The hydrodynamic theory depends on the obstruction of the normal outlets of the fourth ventricle by developmental or other anatomical anomalies. By this theory, it is the abnormal cerebrospinal fluid (CSF) pulsations that lead to the enlargement of the cavity. Alternatively, it has been proposed that the Arnold-Chiari malformation is an acquired anomaly secondary to excessive moulding of the head during difficult, usually high forceps, deliveries. The subsequent deformity could dissociate pressures between cranial and CSF spinal pressures, leading to syrinx formation. Numerous other theories include cystic degeneration of an intramedullary glioma or after resorption of an intramedullary haematoma after cord trauma.

MRI is the diagnostic test of choice. Cystic enlargement of the cord generally extends over several segments with signal intensity similar to that of CSF. Cyst margins are typically irregular with folds and septations. If a syrinx is identified, one must specifically look for craniovertebral junction anomalies and intramedullary tumours (with gadolinium enhancement).

Management of these patients is controversial and varies from a "wait and see" policy to one of aggressive neurosurgery.[4] On a background of slowly progressive minimal disability, the associated morbidity of surgery and a lack of unequivocal benefit from most surgical series, many neurologists advocate a conservative approach. On the other hand, many recommend neurosurgical treatment, especially in the context of severe/rapidly progressive symptoms, cord neoplasms, or an Arnold-Chiari malformation. In tumour associated syringomyelia, excision of the mass nearly always results in resolution of the syrinx. In post-cord trauma patients, simple drainage by percutaneous needle aspiration or open syringotomy provide temporary relief at best, as the

Figure 1

Figure 2

cavity will re-expand on spontaneous closure of the syringotomy. Prolonged successful drainage necessitates placement of a silastic tube connecting the syrinx to either the pleural or pericardial space. In Arnold-Chiari associated syringomyelia, ventriculoperitoneal shunting is normally performed if there is significant hydrocephalus present. If no hydrocephalus is present, or if shunting fails to relieve symptoms, posterior fossa decompression usually shrinks or even completely resolves the syrinx.

Final diagnosis
Syringomyelia.

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An uncommon cause of lumbar radiculopathy

Q1: What are the features seen on the MRI scan (see p XXX)?
- The sagittal T2-weighted image shows low signal (suggestive of disc degeneration) at L4/5 and L5–S1 levels and a high intensity cystic lesion at L4/5 level with a low signal intensity rim due to the cyst wall.
- The axial T2-weighted image shows that the cystic lesion arising from the right L4/5 facet joint and indenting the theca posterolaterally.
- Facet joint hypertrophy/arthritis.

Q2: What is the likely diagnosis?
Facet joint cyst is the diagnosis. Facet joint cysts can be either synovial cysts or ganglia. Synovial cysts have a synovial lining and communicate with the joint whereas ganglia, in a similar site, lack a synovial lining but otherwise have similar components. Only histopathological examination can aid in differentiating between the two. Both types of the cysts arise in association with degenerative disease of spine and can cause similar symptomatology.

Q3: What is the usual line of management?
Surgical decompression and resection of the cyst has been the most effective and widely used treatment modality in symptomatic patients. Conservative management including bed rest, anti-inflammatory medications, immobilisation with a brace, and observation for spontaneous resolution can be used if symptoms are non-intrusive and acceptable to the patient. Facet joint aspiration and injection with steroids and long acting local anaesthetics may provide temporary or prolonged pain relief in patients unfit for surgery.

Discussion
Radiculopathy presenting as sciatica is a common clinical scenario. Facet joint cysts can

present as radiculopathy mimicking the more common prolapsed intervertebral discs. As radiculopathy is very common in clinical practice, surgeons and physicians alike should be aware of this entity and its clinical presentation.[1] The incidence of lumbar facet synovial cysts as a cause of back pain and sciatica based on MRI has been suggested to be 0.65%.[2]

Facet joint cysts occur adjacent to a facet joint in the extradural space. These are most common between the fourth and fifth lumbar vertebrae (68%). Fifteen per cent occur at L5–S1, 12% at L3/4, and 5% at more proximal levels.[2] The majority occur on the medial aspect of facet joints within the posterolateral aspect of the spinal canal at 2–5 o'clock (on the left) or 7–10 o'clock (on the right) positions.

The aetiology of facet joint cysts is poorly understood. The favoured theory is excess stress (motion or direct trauma) or osteoarthritic changes causing herniation of synovial tissue through a capsular defect. Other theories are (1) mucous degeneration of connective tissues, (2) proliferation of developmental synovial rests, and (3) metaplasia of pluripotential mesenchymal cells.[1]

These cysts present with chronic low back pain with 84% exhibiting radicular symptoms. Rarely they can be asymptomatic.[2] Depending on size and location, neurogenic claudication may be the presenting symptom. Cauda equina syndrome can be caused by spontaneous or post-traumatic haemorrhage into the cyst.

With facet joint cysts, sciatica can be due to direct pressure on the nerve root or to irritation associated with inflammatory response around the cyst and often does not respond to conservative measures. Inflammatory response or ischaemia of the nerve root itself can be the causative mechanism. Spontaneous resolution also occurs and cyst rupture has been cited as an explanation for this.[3] Lumbar facet synovial cysts are sometimes an incidental finding and careful consideration of clinical findings is essential when assessing the pain.

MRI provides an accurate method of diagnosis in many cases. Facet joint degeneration is an almost universal finding. Degenerative spondylolisthesis is seen in 42% to 65% of cases.[2] The cyst size varies from 5–25 mm with varying degrees of nerve root and thecal compression. The wall of the cyst is best identified in T2-weighted images. Rim enhancement is a frequent finding after intravenous injection

with gadolinium chelate, which is associated with histological evidence of subacute inflammatory change. Previously myelography, computed tomography, computed tomographic myelography, facet arthrography, and computed tomographic facet arthrography were used.

The differential diagnosis on MRI and computed tomograms includes sequestered disc prolapse, conjoint nerve root, an intraspinal cyst and a cystic neurofibroma.

Surgical decompression and resection of the cyst has been the most effective and widely used treatment modality in symptomatic patients with a success rate of 80%.[4] Conservative management including bed rest, anti-inflammatory medications, and immobilisation with a brace can be used if symptoms are non-intrusive and acceptable to the patient. Symptomatic improvement in about 55% of patients has been reported.[5] Facet joint aspiration and injection with steroids and long acting local anaesthetics may provide temporary or prolonged pain relief in patients unfit for surgery.[2 6]

Final diagnosis
Right sided lumbar facet joint cyst.

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A painful and deformed wrist

Q1: What does the radiograph show (see p XXX)?
The radiograph is an anterior posterior view of the left wrist, showing fracture of the radial styloid, transverse middle third fracture of the scaphoid, and radiocarpal dislocation with ulna displacement.

Q2: What are the possible complications associated with this injury?
Complications could be early or late. Early complications are neurological injuries, with symptom and signs of median nerve compression, and injury to the ulna nerve. Vascular injury is another early complication. Compartment syndrome of the hand could occur with the hallmark of diagnosis being pain on

Learning points

- Radiocarpal dislocation is a rare injury of the wrist.
- The patient should be examined for neurovascular deficit.
- This injury requires urgent reduction and stabilisation.
- In post-traumatic carpal instability, a contralateral radiograph is useful in excluding ligaments laxity.

passive stretching of the involving intrinsic muscles and intrinsic paralysis. Late complications are non-union of the scaphoid fracture, post-traumatic osteoarthritis of the radiocarpal joint, and post-traumatic carpal instability.

Q3: How will you treat this injury?

Initial assessment of the patient should be carried out, and the hand should be examined for neurovascular deficit, and other associated injuries, followed by splinting before radiography.

Manipulation should be done under general anaesthesia as an emergency to reduce the dislocation and the radial styloid fracture and to fix it with Kirschner wires. Considering that this is a high energy injury the carpal tunnel will be decompressed.

It may be necessary to fix the scaphoid fracture internally if there is instability or displacement after reducing and fixing the radial styloid fracture.

Below elbow plaster should be applied for six weeks, then a futura splint for another two weeks. This should be followed by physiotherapy, weekly radiographs for the first three weeks, and another at six weeks after the injury. Kirschner wire should be removed after three weeks.

Discussion

Radiocarpal dislocation is a rare injury to the wrist and the incidence with or without fracture of the styloid process or intercarpal subluxation is about 0.2% of all dislocations.[1]

The mechanism of this injury is not really known but Rosado has tried to explain the mechanism as injury resulting from increasing violence.[2]

1. Contusion and sprain with no bone or ligament damage.

2. Radiocarpal dislocation with torn radiocarpal ligament but intact intercarpal ligaments, the hamate impinges on the anterior radial lip and prevents spontaneous reduction of the carpus.

3. Anterior lunate dislocation with torn radiocarpal and intercarpal ligament.

4. Displaced fracture dislocation with multiple fractures and ligament damage.

Reduction is usually easy but stability has to be maintained either by external or internal skeletal fixation.

If stability without fixation is attempted in dorsal dislocation then it should be immobilised in extension,[3] and palmar dislocation should be immobilised with above elbow cast in slight flexed position for four weeks then gentle mobilisation.[2] This injury is quite unstable and regular review with radiographs is necessary to detect early loss of reduction.

Dislocation without fracture could be stabilised with Kirschner wire passed from the radial styloid into the carpus and plaster.[4] If

the radial styloid is fractured it should be fixed with Kirschner wire and plaster applied for six weeks.

If there is an associated scaphoid fracture, internal fixation may be necessary if it is displaced or unstable.

Post-traumatic carpal instability could be a complication of radiocarpal dislocation. While this instability pattern may occasionally be seen with traumatic laxity of the palmar radiocarpal ligament, it is frequently seen bilaterally as a congenital condition, possibly a sequel of ligament laxity.[5][6]

Radiography of the contralateral wrist will often be helpful in differentiating those patients with post-traumatic instability from ligament laxity.

Final diagnosis

Radiocarpal dislocation of the wrist.

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Massive haemoptysis in a young woman

Q1: What investigation would you perform next?

Massive haemoptysis is a life threatening event since flooding of the airways and alveoli may lead to respiratory failure. It requires rapid evaluation, therefore, bronchoscopy under general anaesthesia should be performed first for the visualisation of the bronchial tree in this patient.

Q2: What possible diagnoses are compatible with the patient's clinical presentation?

Other possible diagnoses include neoplasm (primary or metastatic carcinoma of the lung), a fungus ball, hydatid cyst, granulamatous diseases (nodular sarcoidosis), connective tissue diseases (rheumatoid arthritis), and vasculitis (Wegener's granulamatosis).

Q3: What is the most likely diagnosis?

The patient's history of recurrent orogenital ulcers and erythema nodosum, and radiological features of two bilateral and rounded opacities with an average size of 3 × 5 cm, suggested Behçet's disease and lung involvement with pulmonary artery aneurysms in this case.

Q4: What investigations would you perform to confirm your diagnosis?

Demonstration of the pulmonary artery aneurysms can be made by non-invasive tech-
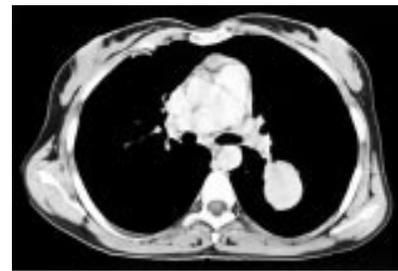

Figure 1 Dynamic computed tomogram revealing simultaneous contrasting opacities with pulmonary arteries.

niques such as dynamic computed tomography, helical computed tomography, or magnetic resonance imaging (MRI) of the thorax, as well as by invasive techniques such as pulmonary angiography.

Q5: How would you manage this patient?

Resection of the aneurysms is indicated and lifesaving in such cases with massive haemoptysis. Medical treatment for Behçet's diseases includes glucocorticoids, and a cytotoxic agent such as chlorambucil, azathioprine, or cyclophosphamide.

Outcome

The patient refused to have surgery, and even though a combined regimen including prednisone (1 mg/kg/day) and cyclophosphamide (2 mg/kg/day) was given she died from massive haemoptysis.

Discussion

Behçet's disease is a chronic multisystem vasculitis of unknown aetiology affecting all sizes of arteries and veins. The *sine qua non* is aphthous ulcers in addition to two or more of the following: genital ulceration; eye lesions such as anterior/posterior uveitis, hypopyon, or retinal vasculitis; and skin lesions such as pustules, nodules, erythema nodosum, or dermatographism. The pathergic skin test is present in 85% of patients. Lungs, kidneys, joints, central nervous system, gastrointestinal tract, cardiovascular system, the epididymis, and muscles can be involved.[1]

Although worldwide in distribution, most large series have been reported from the shores of the Mediterranean, particularly Turkey and Greece, as well as the Middle East and Japan.[1][2]

Lung involvement in Behçet's disease is rare, occurring in only 1%–5% of patients. Aneurysm formation of pulmonary arteries is one of the most significant lesions with potential erosion into the bronchial tree with consequent exsanguination. These aneurysms may be either bilateral or unilateral and tend to affect predominantly young males. Hughes-Stovin syndrome (deep venous thrombosis associated with multiple pulmonary artery aneurysms) may be a form of Behçet's disease.[1]

Haemoptysis, sometimes massive, is the most common clinical manifestation of pulmonary involvement, and it is a poor prognostic sign, leading to death in 30% of patients, usually within two years. Haemoptysis may be caused by the rupture of a pulmonary artery aneurysm with erosion into a bronchus (pulmonary artery bronchial fistula) or it may be the result of the development of in situ thrombosis related to the active vasculitis.[3]

Learning points

- Lung involvement in Behçet's disease is rare, occurring in only 1%–5% of patients. Pulmonary artery aneurysm is the most important type of lung involvement and is responsible for the increased risk for fatal massive haemoptysis.
- Although pulmonary artery aneurysms almost always occur in male patients with Behçet's disease, it should also be considered in females, especially when they present with massive haemoptysis and have either single or multiple sharply demarcated round opacities on the chest radiograph.
- Demonstration of pulmonary artery aneurysms can be done safely by dynamic computed tomography of the thorax instead of invasive procedures.
- When haemoptysis occurs, it is a sign for a poor prognosis; however, urgent surgical resection may be lifesaving in cases with massive haemoptysis.

Pulmonary angiography is regarded as the gold standard for the detection of pulmonary artery aneurysms in general, but this procedure may carry some risks for patients with Behçet's disease. Venous puncture or rapid injection of a large quantity of contrast medium may initiate a thrombus or aggravate an existing one in a patient with Behçet's disease. Therefore non-invasive procedures such as helical computed tomography, dynamic computed tomography, or MRI angiography are suggested as the more appropriate techniques in such patients.[4] Demonstration of the pulmonary artery aneurysms in the presented case was made by dynamic computed tomography of the thorax (fig 1).

Numerous therapeutic regimens including prednisone, cyclophosphamide, chlorambucil, and colchicine have been suggested for Behçet's disease. Although a radiographic regression in response to medical treatment has been demonstrated, the development of vascular aneurysms is considered a poor prognostic sign. In cases of massive haemoptysis related to ruptured aneurysm, urgent surgical resection may be necessary.[1]

This case illustrates that a pulmonary artery aneurysm can develop in association with Behçet's disease even in young females and it is a fatal complication of the disease and does not respond to any treatment. Therefore, Behçet's disease should be considered in the differential diagnosis of such cases with massive haemoptysis and chest radiography showing multiple round consolidations, especially in regions where Behçet's disease is common.

Final diagnosis

Lung involvement of Behçet's disease with multiple pulmonary artery aneurysms.

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An unusual presentation of calvarial tuberculosis

Q1: What is the differential diagnosis and what other investigations are indicated?

The differential diagnosis of multiple osteolytic lesions of the skull would include metastases, multiple myeloma, histiocytosis and pyogenic, fungal, or tuberculous osteomyelitis.

Investigations for this case should include the work-up for primary malignancy, myeloma, tuberculosis, or other systemic infections and chronic inflammatory disease. Apart from a raised erythrocyte sedimentation rate (75 mm at one hour), routine haematology was normal. Chest radiography showed mildly prominent hilar opacities. Ultrasound of the abdomen revealed mild enlargement of the para-aortic lymph nodes. The bone scan showed hot spots in the corresponding regions in the skull and also in D4, D12, and L4 vertebrae. Urinary Bence-Jones proteins were negative.

Since the laboratory and radiological investigations were inconclusive, the patient underwent an open biopsy of the right frontal bone lesion for a definitive diagnosis. At surgery the scalp was normal. Soft granulation tissue was encountered where the cortical bone was destroyed. There was no extradural granulation tissue. The histopathological examination was consistent with tuberculosis showing non-caseating granulomata composed of epitheloid histiocytes and multinucleated giant cells of Langhan's and foreign body type.

Q2: What are the different forms of calvarial tuberculosis and what is the common presentation?

The different radiological forms of calvarial tuberculosis described are[1]:

- Circumscribed lesions of the sclerotic and lytic type.
- Diffuse tuberculosis of the cranium.

Though the bony involvement maybe variable calvarial tuberculosis often presents with painful scalp swellings, subgaleal collections, discharging sinuses, and variable amount of extradural granulation tissue. Isolated skull bone involvement is rare.

Q3: What is the management of this condition and indication for surgery?

Management of calvarial tuberculosis includes establishment of the diagnosis and adequate antituberculous therapy, along with appropriate surgical intervention. An urgent surgical intervention is indicated in the presence of large extradural collections causing mass effect and neurological deficits. The presence of large collections of pus in the subgaleal plane and sinus formation necessitates thorough debridement and excision of the sequestrum along with the infected granulation tissue. Associated secondary infection needs treatment with appropriate antibiotics. Finally an excision biopsy of the lesion would

Learning point

Tuberculous osteomyelitis should be considered in the differential diagnoses of multiple lytic lesions of the skull especially in endemic areas. High index of suspicion and excision biopsy can lead to early diagnosis and good outcome.

be indicated, to establish the diagnosis, where there is high index of suspicion and other results are inconclusive. Surgical treatment should always be followed by adequate antituberculous therapy, considering the indolent nature of infection.

Discussion

Calvarial tuberculosis was first reported by Reid in 1842.[1] It usually occurs secondary to haematogenous spread from a primary focus elsewhere in the body that may not always be evident. Trauma and surgery can result in direct inoculation of the organism.[2][3] It is also proposed that the increased vascularity and transient decreased resistance at surgery may result in specific homing of bacilli, as the inflammatory cells are attracted to the site of trauma and act as vectors for the bacilli.[1]

Skull tuberculosis is very rare and is reported to occur in only 0.01% of patients with mycobacterial infections.[4] Most cases occur in the first two decades; however infants are rarely affected, probably because of the paucity of cancellous bone in the skull.[5] The frontal and the parietal bones are usually involved.[6] The type of clinical presentation depends perhaps on the immunity of the individual.[7] Once the marrow of the diploe is seeded with the inoculum, the infection spreads towards the inner and outer table, causing bone destruction and formation of granulation tissue. The extension of the infection through the diploe is resisted by proliferation of an encircling layer of concentrically placed fibroblasts and if the process is not arrested, extension then takes place through either tables.[5] If the process is rapid, sequestration may occur; this appears as "bone sand" on radiography. Involvement of the outer table is usually associated with scalp swelling or a discharging sinus while involvement of the inner table results in extradural granulation tissue. The dura acts as a barrier to further spread, however intradural involvement is occasionally seen. A good immunity will cause slow and restricted evolution of the lesion, while decreased resistance will rapidly lead to subgaleal or extradural collections. Rare forms of presentation include seizures, motor deficits, meningitis, and non-specific headache as in our case.

A solitary discrete round or oval punched out osteolytic defect with minimal surrounding sclerosis in the frontoparietal bones is the commonest presentation of skull tuberculosis. When multiple they are of diffuse type with serpiginous and geographical defects of extensive bone loss.[8] In our case, though an individual lytic lesion would have suggested a tuberculous aetiology, their multiplicity was unusual. There was no clinical or radiological evidence of involvement of surrounding soft tissue. Other investigations revealed no definitive evidence of an extracalvarial primary tuberculous focus. Thus a possibility of malignancy could not be ruled out without a

histopathological diagnosis. A high index of suspicion and biopsy resulted in early diagnosis of this condition, before the adjacent soft tissues and the central nervous system were involved. Prompt treatment with antituberculous therapy resulted in cure and total regression of these lesions with no morbidity.

Final diagnosis

Calvarial tuberculosis presenting as multiple osteolytic lesions.

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