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. 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 Charcot (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 asymptomatically 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 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. 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. 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 or 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
cavity will re-expand on spontaneous closure of the syringomyelia. 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.

**References**


**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/arthritus.

**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. 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%. 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 pluripotent mesenchymal cells. These cysts are found with chronic low back pain with 84% exhibiting radicular symptoms. Rarely they can be asymptomatic.3 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.4 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 spondylothesis is seen in 42% to 65% of cases.5 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%.6 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.7 Facet joint aspiration and injection with steroids and long acting local anaesthetics may provide temporary or prolonged pain relief in patients unfit for surgery.

**Final diagnosis**
Right sided lumbar facet joint cyst.

**References**


**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
passive stretching of the involving intrinsic muscles and intrinsic paralysis. Late complications are non-union of the scaphoid fracture, post-traumatic osteoarthrits 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 this 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 future 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. The mechanism of this injury is not really known but Rosado has tried to explain the mechanism as injury resulting from increasing violence.

1. Contusion and sprain with no bone or ligament damage.
2. Radiocarpal dislocation with torn radio-
carpal ligament but intact intercarpal liga-
ments, the hamate impinges on the anterior
carpal ligament but intact intercarpal liga-
ment damage.
3. Anterior lunate dislocation with torn
di-carpal and intercarpal ligament.
4. Displaced fracture dislocation with mut-
iple 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 immobi-
1ised in extension, and palmar dislocation
should be immobilised with above elbow cast
in slight flexed position for four weeks then
gentle mobilisation. This injury is quite un-
stable and regular review with radiographs
is necessary to detect early loss of reduction.

Dislocation without fracture could be stabi-
1ised with Kirschner wire passed from the
radial styloid into the carpus and plaster. 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 dis-
placed 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.

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.

References


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
may lead to respiratory failure. It requires
massive haemoptysis in a young woman.

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, granuloma-
tous diseases (nodular sarcoidosis), connec-
tive tissue diseases (rheumatoid granulomatosis), and vasculitis (Wegener granulomatosis).

Q3: What is the most likely diagnosis?
The patient’s history of recurrent orogenital
ulcers and erythema nodosum, and radiologi-

cal features of two bilateral and rounded
opacities with an average size of 3 × 5 cm,
suggested Behcet’s disease and lung involve-

ment with pulmonary artery aneurysms in
this case.

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

Demonstration of the pulmonary artery aneu-
rysms can be made by non-invasive tech-
niques such as dynamic computed tomogra-

hy, helical computed tomography, or mag-

netic 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 haemo-
ptyysis. Medical treatment for Behçet’s dis-
cases 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 pred-

nison (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 vas-
culitis 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 der-

matographism. The pathergic skin test is
present in 85% of patients. Lungs, kidneys,
joints, central nervous system, gastro-

intestinal tract, cardiovascular system, the
epididymis, and muscles can be involved.

Although worldwide in distribution, most
large series have been reported from the
shores of the Mediterranean, particularly Tur-
key and Greece, as well as the Middle East and
Japan.

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 exanguination. 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.

Haemoptysis, sometimes massive, is the
most common clinical manifestation of pul-
monary involvement, and it is a poor prognos-

tic 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 (pul-
monary artery bronchial fistula) or it may be
the result of the development of in situ
thrombosis related to the active vasculitis.
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. 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 hae-moptysis, pulmonary artery aneurysm may not respond to any treatment.

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 extra-dural granulation tissue. The histopathological examination was consistent with tuberculosis showing non-caseating granulomas composed of epithelioid 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:

- Circumscribed lesions of the sclerotic and lytic type.
- Diffuse tuberculosis of the cranium.
- Through bone involvement maybe variably calvarial tuberculosis often presents with painful scalp swellings, subgaleal collections, draining 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 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. 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. 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.

Skull tuberculosis is very rare and is reported to occur in only 0.01% of patients with mycobacterial infections. Most cases occur in the first two decades; however infants are rarely affected, probably because of the paucity of cancellous bone in the skull. The frontal and the parietal bones are usually involved. The type of clinical presentation depends perhaps on the immunity of the individual. 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. If the process is rapid, reabsorption may occur; this is known 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 intra-dural 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 bone is the commonest presentation of skull tuberculosis. "When multiple they are often with serpiginous and geographical defects of extensive bone loss." 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 extracranial 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.

**References**