Rare disease

A rare presentation of spinal epidural abscess

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A 77-year-old retired engineer presented to accident and emergency with deteriorating shortness of breath that had been troubling him for several months. At that time, he was being investigated by a chest physician who had identified bilateral diaphragmatic paralysis on ultrasound and was awaiting further imaging. Clinical assessment and nerve conduction studies on this admission were compatible with a diagnosis of motor neuron disease but specialist neurology input recommended an MRI to rule out cord pathology. This proved problematic as the patient was non-invasive ventilation dependent and unable to lay supine as this further compromised his respiratory function. To ensure that a potentially reversible cause for his symptoms was identified, the patient was intubated for an MRI which subsequently demonstrated multi level spinal epidural empyema. The benefits of neurosurgical intervention were judged to be uncertain at best, and following discussion with the family, active care was withdrawn. The patient passed away shortly thereafter.

BACKGROUND

This case demonstrated a rare pathology, important learning points and some demanding clinical problems.

First, remembering mechanical causes of breathlessness. Second, considering the differential diagnoses when electromyogram (EMG) studies are consistent with motor neuron disease (MND). Third, the importance of appropriate imaging in reaching a diagnosis. Finally, from a clinical standpoint, the difficulty of imaging a patient who was unable to lay supine, and the complex process behind deciding on the benefits of surgery in this situation.

CASE PRESENTATION

The patient presented to the emergency department with worsening shortness of breath which had already been troubling him for around 8 months. During this period, he had been assessed by a chest physician who had identified bilateral diaphragmatic paralysis on ultrasound and was arranging further investigations. His dyspnoea had gradually deteriorated over this time but accelerated in progression over the last month such that he was breathless at rest. In addition, he complained of some upper limb weakness over the previous month, and some pain in his neck and upper chest for the last few days. There had been no pain or focal neurological disturbance during prior consultations with the chest physician.

His medical history included benign prostatic hyper trophy, hypertension, basal cell carcinoma and a transient ischaemic attack 5 years earlier. Socially, he was a retired engineer who was usually fully independent with an excellent exercise tolerance. He had no smoking history and minimal alcohol intake, with no recent long haul travel.

Clinical examination elicited decreased breath sounds bilaterally, and decreased power at the shoulder and elbow bilaterally. The left arm reflexes were depressed with altered perception of pinprick in a C5 distribution.

INVESTIGATIONS

Blood gases on admission demonstrated type II respiratory failure with compensated respiratory acidosis. Routine bloods demonstrated consistently elevated white blood cells and C-reactive proteins (CRPs) throughout his admission. The creatine kinase was raised which was thought to be statin induced as this had recently been stopped by his general practitioner because of myalgia. The cerebrospinal fluid (CSF) demonstrated a raised number of white cells, $40 \times 10^6/l$, with 60% lymphocytes and 40% polymorphonuclear leucocytes. Total CSF protein was 1045 mg/l and plasma/CSF glucose was 7.1 and 3.5 mmol/l, respectively. No organisms were identified. Blood cultures yielded no organism but the sputum culture grew *Candida albicans*. EMG studies demonstrated widespread chronic denervation and fasciculations suggesting the possibility of anterior horn cell disease. The MRI report detailed a cervical epidural collection suggestive of an empyema with severe cord compression and oedema (figure 1).

DIFFERENTIAL DIAGNOSIS

Before the MRI was conducted the following conditions were considered:

- MND
- Focal cord pathology.

Before the EMG studies, primary myopathy was also considered in the differential diagnosis.

TREATMENT

The patient was treated with non-invasive ventilation which provided some symptomatic relief of his breathlessness and reversed his type II respiratory failure. Antibiotics were instigated for the persistently raised CRP although this was on an empirical basis. On determining the diagnosis, the definitive treatment in this case would have been drainage of the collection thereby decompressing the cord,

Figure 1 (A, B) Sagittal T1 MRI images demonstrating high signal areas representing a multi level cervical epidural abscess with cord oedema. The thoracic collections were thought to be more in keeping with haematomas.

but due to the chronicity of the insult, a good outcome was unlikely.

OUTCOME AND FOLLOW-UP

The benefits of surgery were discussed at length with the family who decided that the chance of regaining an acceptable quality of life was very low. With the family in full agreement, the patient was made comfortable and active management was withdrawn. He passed away shortly afterwards with his family present.

DISCUSSION

Spinal epidural abscess is uncommon, with Rea and Rigamonti reporting a frequency of 0.2–1.2 cases per 10 000 hospital admissions.^{1 2} The non-specific presentations of this challenging condition often lead to a delay in diagnosis and subsequent treatment, which inevitably results in poorer outcomes.^{3–5} The mean age affected is 50–60 years, and there appears to be a slight male preponderance, with ratios reported up to 2:1.⁶ Seeding of the infection from a distant source is almost always the cause, and back trauma, diabetes, intravenous drug abuse, degenerative joint disease, renal and hepatic failure are thought to be predisposing factors.⁶ *Staphylococcus aureus* is widely reported as the most frequently isolated organism^{2–9} accounting for between 60%–90% of cases,⁶ although many other organisms including fungi have also been implicated.^{5 9} The abscess damages the spinal cord via direct compression and also thrombosis of spinal blood vessels.⁶ Abscesses have been noted at all levels of the spinal cord but thoracic and lumbar involvement is most common, with cervical involvement in 10%–25% of cases.⁶

Clinically there are typically four stages in the evolution of the condition, initially with focal vertebral pain at the affected level of the spine, progressing through radiculopathy with paraesthesia, followed by weakness and finally paralysis at stage 4.⁶ Cases reported include this sequential picture together with selected components of the classical manifestation,^{3–5 7–9} although neck/back pain was clearly shown to be the most common symptom.^{3–9} It is therefore not surprising that with the rarity of this condition

juxtaposed with the ubiquity of back pain, making a swift diagnosis can prove difficult.⁶ A meta analysis of 915 cases reported that paralysis only affected 34% of patients, and diaphragmatic paralysis was not reported as a distinct presenting feature in any case.⁸

Dyspnoea secondary to bilateral diaphragmatic paralysis is in itself a rare cause of respiratory failure.¹⁰ The differential diagnosis of bilateral diaphragmatic paralysis is wide ranging including such entities as acid maltase deficiency¹¹ and spinal cord infarction.¹² Classically it presents with severe orthopnoea, as when standing patients are able to expand the rib cage and relax abdominal muscles, facilitating the downward movement of the diaphragm.¹¹ This was the case with our patient and delayed his MRI as careful consideration was given to the benefits against risks of having to intubate him to allow the imaging to take place.

Diagnosis of spinal epidural abscess is best made with enhanced MRI imaging^{5 6 9} accompanied by haematological investigation including blood cultures. A raised CRP is frequently present,³ and raised white cells with an increased erythrocyte sedimentation rate are common, although non-specific.⁶ Blood cultures are valuable although negative in 30% of cases and an lumbar puncture should not be undertaken routinely due to the risk of spreading the infection subdurally.^{6 9} Should cerebrospinal fluid be obtained, raised white cells and protein are often found, but a causative organism is not detected in up to 80% of samples.⁶ Treatment is primarily surgical drainage to relieve cord compression and long-term targeted antibiotics; unfortunately neither of these measures was considered either effective or viable in this case. It is reported that delayed intervention, more than 48 h following the onset of paralysis, results in irreversible cord injury,⁶ hence the poor prognosis for this patient. Early diagnoses increases chances of survival but overall mortality is reported between 5% and 13% even if surgery is undertaken.⁶

This case demonstrates the importance of considering central mechanical causes of breathlessness and the non-specific presentation but potentially devastating nature of spinal epidural abscess. One must also be mindful of the value of imaging in such patients and the practical challenges of imaging a breathless patient.

Learning points

- ▶ Remember to consider mechanical causes when assessing the breathless patient.
- ▶ Be mindful of the differential diagnoses of MND.
- ▶ Appreciate the value of imaging in such patients.
- ▶ Acknowledge the difficulties of imaging the breathless patient.
- ▶ Carefully consider the lengths to go to when investigating a reversible cause.

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Competing interests None.

Patient consent Obtained.

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