CASE REPORT

Mediastinal mass: to resect or not to resect?

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SUMMARY

We describe a case of a 63-year-old man presenting in an acute confusional state with a markedly abnormal chest radiograph. During investigation, he developed aspiration pneumonia and was ventilated on the intensive care unit before confirming that he had autoimmune encephalopathy secondary to benign thymoma. After treatment with intravenous immunoglobulins he made an impressive recovery allowing him to be transferred to a tertiary unit for resection of the underlying mass, thus resulting in an excellent neurological recovery.

BACKGROUND

Acute confusion is a common presenting complaint on the acute medical take; this case demonstrates a step by step approach to this clinical problem that results in a rare diagnosis. This is pertinent because as we demonstrate, autoimmune encephalopathy can result in rapid deterioration but is potentially completely reversible.

CASE PRESENTATION

A 63-year-old man presented to the emergency department with a 1 week history of confusion, personality change and short-term memory loss. He had been seen by the general practitioner, and started on antidepressants and benzodiazepines, but he became progressively more agitated. He had recently retired from being a motor technician and had no medical history other than an episode of supraventricular tachycardia. He had never been a smoker and drank 10 units of alcohol per week. Examination revealed no focal neurology but he was disorientated and his abbreviated mental test score was 4 of 10.

INVESTIGATIONS

Initially, a septic screen was performed to exclude infection. Blood investigations confirmed normal inflammatory markers, bone profile and renal and liver enzymes. Urine dipstick was negative for nitrites and leucocytes. CT and MRI of the brain were unremarkable. Cerebrospinal fluid (CSF) was clear and colourless with raised protein at 1.03 g/L, white cells were 12×10^6 /L and there was no growth on culture.

Chest radiograph revealed a large mass in the right lower zone possibly arising from the mediastinum. CT of the thorax demonstrated a large well-defined heterogeneous mass, approximately 13 cm, arising from the anterior mediastinum. The patient was reviewed by a neurologist (MW) who thought his condition was likely to be an autoimmune encephalopathy secondary to either a thymoma or

teratoma. As a result, N-methyl D-aspartate and potassium channel antibodies were checked as advised, and subsequently both found to be negative.

The case was discussed in the lung multidisciplinary team meeting and a plan for biopsy of the mass was made.

While awaiting percutaneous biopsy of the mediastinal mass, the patient became more confused, agitated and drowsy. On day 5 of the admission, his oxygen saturations dropped and a repeat chest radiograph revealed consolidation around the mediastinal mass and in the left lower zone. A diagnosis of aspiration pneumonia was made, and he was started on intravenous antibiotics. He developed severe type one respiratory failure with a pO₂ of 6.02 kPa on FI02 of 0.85, and was transferred to intensive care where he was intubated and ventilated. During his stay on ICU, he had a biopsy of the mediastinal mass and a normal bronchoscopy. A trial of extubation failed due to increased oxygen requirements and a weak cough. Following a further neurology consult, he was given intravenous immunoglobulins in a dose of 30 g daily for 5 days and a continued course of corticosteroids. Shortly after this he became much more alert, better orientated and was successfully extubated on day 17.

Histology from CT-guided biopsy of the mediastinal mass was consistent with thymoma. The patient was transferred to a tertiary cardiothoracic centre for surgical resection of the mass. A 20×20 cm mass arising from the inferior pole of the right lobe of thymus was identified and resected: histology showed a completely resected benign thymoma (type A).

DIFFERENTIAL DIAGNOSIS

In view of the acute history, infection was initially at the forefront of the differential diagnosis; normal inflammatory markers and absence of temperature excluded this in the main. However, the elevated CSF protein did raise the possibility of viral encephalitis. The key in this case was the chest radiograph, which was carried out after the CT of the brain. The appearance of a smooth edged mass, probably arising from the mediastinum, narrowed the unifying diagnosis to teratoma or thymoma. Thus the proof was in the biopsy. Despite the negative antibodies; the response to intravenous immunoglobulin not only confirmed the diagnosis, but also facilitated extubation and therefore transfer of the patient for surgical resection.

TREATMENT

Initial treatment was supportive. After the lumbar puncture, intravenous acyclovir was started. Early involvement of the neurologist (MW) guided

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further treatment, which initially consisted of methylprednisolone at a dose of 500 mg a day for 3 days. Subsequently, the patient was given intravenous immunoglobulins and a continued course of corticosteroids were administered before the patient was accepted for resection of the thymoma.

OUTCOME AND FOLLOW-UP

On clinical review 3 months postsurgery, the patient's behaviour and personality had returned to normal and all that remained was impaired memory of his time on intensive care.

DISCUSSION

Thymoma is the commonest anterior mediastinal mass. Its association with antiacetylcholine receptor antibodies and myasthenia gravis is well described. However, thymoma has been linked to other autoimmune-mediated neurological phenomena including encephalitis, neuromuscular hyper-excitability, stiff-man syndrome and progressive encephalomyelitis with rigidity and myoclonus (PERM).^{1 2}

Our patient presented with an encephalopathy that is described in the context of thymoma in just under 40 cases²: presenting symptoms described include seizures, personality change and agitation. Vernino and Lennon³ analysed stored serum of 201 patients with a histological diagnosis of thymoma (195) or thymic carcinoma (6) with and without neurological symptoms to assess the frequency of neuronal and muscle autoantibodies. Voltage-gated potassium channels were found in those with neuromuscular hyper-excitability and encephalitis. Forty per cent of patient with thymoma but without neurological features had one or more positive autoantibodies. Rizzardi *et al*² describe a case of thymoma with biopsy proven encephalitis where antibodies were negative. Paterson *et al*⁴ looked specifically at voltage-gated channel complex antibodies and found that highly positive levels were only 'definitely' clinically relevant in 38% of cases. In our case, the voltage-gated potassium channel antibody was negative on two occasions.

Treatment of autoimmune encephalopathy is aimed at managing the underlying mass (if present), usually by resection, with or without immunosuppression or immunomodulation.⁵ Suh *et al*⁶ describe a similar case to the current one, with a good response post-thymectomy for a grade B thymoma with myasthenia gravis and encephalopathy. The difficulty in our case was confirming the diagnosis and timing of surgery in a

tertiary centre in a ventilated patient being managed in another centre.

Encephalopathy in association with thymoma is a rare condition. Confirmation of diagnosis may be sought serologically or histologically. However, as in our case, it may be a clinical diagnosis confirmed by response to treatment. Management in this case included a good response to parenteral immunoglobulins following a poor response to high-dose steroids and prompt resection of the underlying mass.

Learning points

- ▶ Chest radiograph should always be part of initial investigation into acute confusion.
- ▶ Early involvement of the neurology team may help guide ongoing management.
- ▶ Autoimmune encephalopathy may completely resolve if a mass associated with it is removed.

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