Rare disease

Sorry... It was not a sore throat!! Castleman's disease in a 19-year-old, previously normal, young patient

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Summary

A 19-year-old male patient presented to the emergency department with a small amount of haemoptysis preceded by coughing and sneezing. He came back from Portugal a week ago. His physical examination was unremarkable and his routine blood tests were normal apart from a slightly raised D dimer. His chest x-ray revealed a mediastinal mass, which appeared on CT as a right-sided mediastinal mass with low attenuation compressing the superior vena cava. The CT angiogram showed multiple arterial supply from the brachiocephalic, vertebral and bronchial arteries; they drained into internal jugular, subclavian and azygous system. CT-guided biopsy was performed followed by operative excision of the mass 5 weeks later. Both histology results confirmed a hyaline vascular variant of Castleman's disease with no evidence of any malignancy.

BACKGROUND

Castleman's disease is a rare disease of autoimmune lympho-vascular origin; it is also referred to as angiofollicular hyperplasia. It affects men and women of all ages. Many of these cases presented with mild symptoms or as incidental findings; the treatment may involve surgical excision and patients may need major operations.

CASE PRESENTATION

A 19-year-old male patient presented with a 2-day history of dry cough and sneezing. Four hours prior to his presentation to the emergency department, he coughed up small amounts of dark red blood. The patient denied any fever, loss of weight, poor appetite or any contact with a patient with chronic cough. He had no bowel or urinary symptoms.

He is a non-smoker and works as a conditioner engineer. The patient had just returned from a 2-week holiday in Portugal with his girlfriend who appears to have no symptoms.

He had an unremarkable physical examination. His initial laboratory investigations were normal apart from a slightly raised D dimer of 128. His chest x-ray showed a right-sided mediastinal mass.

INVESTIGATIONS

Blood investigations

Full blood count: white cell count 6.7, haemoglobin 15.4, platelet 233 and D dimer 128.

Coagulation, urea and electrolytes, and liver function test within normal range.

Chest x-ray

Right-sided mediastinal mass displacing trachea slightly to the left (figure 1).

CT chest/abdo/pelvis with contrast

There is a 9.2×8.1 cm² right middle mediastinal mass (figure 2). The mass is rather vascular with some feeding vessels related to it. The mass abuts the ascending aorta as well as the right subclavian artery medially and inferiorly it is related to the right pulmonary artery. This shows peripheral enhancement but central area of low attenuation in keeping with central necrosis. The mass is causing some displacement of the mediastinum and compresses the superior vena cava and the confluence of the brachiocephalic veins but these appear patent.

There is a little indentation of the trachea and the right main bronchus by the mass.

No mediastinal lymphadenopathy is seen. No focal lung abnormality is identified. Normal appearances of the liver, spleen, kidneys, adrenals and the pancreas. No intra-abdominal lymphadenopathy.

No destructive bony lesions seen.

Right middle mediastinal mass as described. The differential for this includes teratoma and nodal mass.

CT thorax with biopsy

An intensely enhancing vascular tumour is noted in the right side of the mediastinum (figure 3). There are large arterial supply vessels noted arising from the right brachiocephalic trunk, the right vertebral artery and tortuous vessels noted in its mid and lower part arising from the bronchial arteries

Figure 1 First chest x-ray.

Figure 2 (A–D) CT thorax pre.

Figure 3 (A, B) CT-guided biopsy.

Figure 4 CXR post.

that arise from the descending thoracic aorta. Multiple draining veins are seen emptying into the azygous system as well as the right subclavian vein. Venous drainage also noted into the right internal jugular vein. Numerous smaller vessels noted in the periphery of the tumour with branches arising from the right internal mammary artery.

Histology

Mediastinal mass – Castleman's disease hyaline vascular type.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis:

1. Hodgkin's lymphoma
2. Castleman's disease
3. Thymoma.

TREATMENT

Patient had surgical excision of the tumour 5 weeks after his presentation; he recovered well from the operation (figure 4).

OUTCOME AND FOLLOW-UP

Patient to have 6-month follow-up and a baseline CT scan on his first visit to compare in-case of recurrence.

DISCUSSION

'In the course of studying tumors of the thymus gland we came through a small group of cases in which enlarged mediastinal lymph nodes resembled thymic tumours grossly, radiologically, and even microscopically and they had been so classified'. By these words Benjamin Castleman *et al* started reporting a series of 13 cases diagnosed with

a lymphoproliferative disorder, which was later on called after Dr Castleman.^{1 2}

Castleman's disease is a rare type of lymphoproliferative disorder with lymph node hyperplasia and vascular proliferation.^{3–5} It is thought to be due to excessive proliferation of B lymphocytes and plasma cells in lymphoid organs associated with increased IL-6 production by these organs; the trigger of this pathology is unknown.⁶ Bilal *et al* reported a case of Castleman's disease with cutaneous involvement, which responded to anti-IL-6 treatment.⁷

Clinically, it can be subdivided into two types: a localised type in which the lymphoid hyperplasia is – by definition – restricted to one site and is associated with excessive angiogenesis.² The patients are usually asymptomatic and most of the lesions present themselves as an incidental finding or by the mass effect – usually as trachea-bronchial compression; their symptoms include cough, shortness of breath and haemoptysis. It is a benign with good postexcision prognosis.⁸ The other type is a multicentric type, which is associated with symptoms such as asthenia 65%, weight loss 67%, fever 69% and 24% of the patients may develop POEMS (peripheral neuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin signs).⁶ HIV patients with multicentric Castleman's disease have more tendencies to develop pulmonary signs and Kaposi's sarcoma.^{9 10}

It can also be classified into thoracic and a rare extra-thoracic type with abdominal lesions in most of the cases.

Furthermore, it can be subdivided into two histological variants: hyaline vascular type and a plasma cell variant; the former is characterised by hyaline follicles and vascular proliferations, it is more common, more benign and has a good prognosis postexcision. Most of the patients diagnosed with the hyaline vascular type are asymptomatic. The plasma cell variant is less common but more aggressive. It can involve any organ and usually associated with multiple lymphadenopathies. Patients with the plasma cell variant are usually symptomatic and may need chemo or radiotherapy. Patients with localised

disease show symptomatic relief after surgical excision. Lesions with the plasma cell variant are less vascular; hence, they demonstrate mild to moderate enhancement on CT.

Learning points

- Castleman's disease is a differential that need to be kept in mind in fit patients with superior mediastinal mass on x-ray.
- Despite been of benign pathology, its close relation to neurovascular structures may cause compression symptoms or make the operation difficult.

Competing interests None.

Patient consent Obtained.

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