CASE REPORT

Unicentric Castleman's disease: an uncommon cause of posterior mediastinal mass

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SUMMARY

Castleman's disease is a rare lymphoproliferative disease that may be unicentric or multicentric in presentation. It may develop anywhere along with the lymphatic system such as the abdomen, neck and thoracic cavity. However, mediastinum is the most common location for unicentric disease. Here, we discuss a unicentric Castleman's disease in a 28-year-old woman who presented with cough, mild dysphagia and a large posterior mediastinal mass.

BACKGROUND

Castleman's disease (CD) is an uncommon and non-well-recognised lymphoproliferative disease also called angiofollicular lymph node hyperplasia. It has two clinical variants, unicentric and multicentric (MC). On histopathology, it has three subtypes, hyaline vascular CD (HV-CD), plasma cell CD (PC-CD) and HH8/HIV-associated variants with HV-CD found to be the most common subtype. Most cases of localised disease have been reported to be in the mediastinum; especially in the anterior mediastinum. The majority of patients are young adults with no typical symptoms. Surgical resection remains the main treatment for localised HV-CD, while unresectable, partially resected and MC tumours are treated by systemic chemotherapy or radiation.¹⁻⁴ Here, we describe a case of posterior mediastinal CD adjacent to the oesophagus in a 28-year-old woman who presented with cough and dysphagia; the tumour was resected completely with thoracotomy.

CASE PRESENTATION

She was a 28-year-old Asian woman presented with a several-month history of dry cough and mild dysphagia to solid food. She remembered no aggravating or attenuating factor for her symptoms. She did not complain of asthenia, anorexia, and fever or weight loss. There was no significant illness in her medical, family or social history. She never smoked and was not an illicit drug user.

INVESTIGATIONS

The whole physical examination was normal. Tests of the blood, electrolytes, renal function, liver function and erythrocyte sedimentation rate were all normal.

A chest radiograph showed a large mediastinal mass (figure 1). The most probable clinical diagnosis was oesophageal leiomyoma, so she underwent a barium swallow study that revealed extrinsic

Figure 1 Chest radiography showing a mediastinal mass.

compression of the oesophagus by a large mediastinal lesion (figure 2).

CT of the thorax showed the lesion to be a large posterior mediastinal mass compressing both the oesophagus and the left atrium (figure 3). The endoscopic ultrasound-guided biopsy of the mass was not helpful and was reported to be non-specific lymphoid hyperplasia. Therefore, she underwent thoracotomy.

Figure 2 Barium study of the oesophagus showing extrinsic compression of the oesophagus by a large mediastinal mass.

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Figure 3 A contrast-enhanced CT scan of the chest showing a large mediastinal mass compressing mediastinal structures.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes tumours of the oesophagus, hiatal hernia, enteric cysts and neurogenic tumours such as schwannoma, neurofibroma and peripheral nerve sheath tumours.

TREATMENT

She underwent total surgical resection of the tumour by right-sided thoracotomy. An encapsulated tumour measuring 7.5×5×4 cm was totally resected. No excessive bleeding occurred during surgery. The histopathological examination of the tissue revealed CD, HV type. Her postoperative period was uneventful without any major complication and she was discharged on the fifth postoperative day.

OUTCOME AND FOLLOW-UP

Four months after surgery, she did well and there was no evidence of tumour recurrence on thoracic imaging (figure 4).

DISCUSSION

CD is a rare lymphoproliferative disease that may develop as a lymphoid mass along the lymphatic chain anywhere in the body. In a majority of patients, CD is localised (70%). However, MC and disseminated variants of CD are not uncommon. Unicentric or a localised form of CD arises more commonly in the mediastinum (60–75%), mimicking more common mediastinal lesions

Figure 4 A CT scan of the chest 4 months after surgery showing no residue of the primary tumour in the mediastinum.

such as thymoma, neurogenic tumours and lymphoma.^{1–5} In addition to the mediastinum, other intrathoracic locations such as the pleural, intrapulmonary regions and the chest wall may be involved. Anterior or middle mediastinum localisation is more common than the posterior compartment. Shah and Shah⁶ reported that there are less than 10 cases of posterior mediastinal CD.^{7–8} There are three histological subtypes of the tumour: HV-CD, which is the most common (90% of cases), PC-CD and mixed cell or HH8/HIV-associated subtypes. Most cases of localised intrathoracic tumours are of the HV subtype. This variant carries the most favourable prognosis. On the basis of a systemic analysis of 406 patients with CD, the 3-year disease-free survival (DFS) rate for patients with the unicentric HV variant was 92.5%, while this rate was 45.7% for patients with the MC-PC variant. Patients with HIV who had the MC-PC variant exclusively had the poorest prognosis with a DFS rate of only 27.8%.⁹ Patients with CD, especially unicentric HV-CD, usually do not have systemic symptoms. Therefore, the tumours are often diagnosed incidentally or on evaluation of symptoms such as cough or dysphagia due to the compressive effect of a large mediastinal mass. The MC, disseminated and PC-subtypes of diseases are more commonly associated with systemic symptoms. Increased sedimentation rate, hypergammaglobulinaemia, peripheral neuropathies, nephropathies and other autoimmune diseases have been reported in such patients.¹⁰

The tumour may develop in any age group. There are controversies in disease predilection for female patients. It seems that unicentric HV disease develops more commonly in young female patients while MC CD develops in older male patients.⁹

CD is a great mimicker as it may present in a similar way to other, more common mediastinal lesions. There is no characteristic finding on thoracic imaging, but the tumour is a hypervascular lesion that may intensely enhance on a contrast CT scan of the chest, where even a feeding artery may occasionally be seen. Calcification and central necrosis are less common findings.¹¹ Fine needle aspiration biopsy guided by either ultrasonography or CT scanning may confirm diagnosis preoperatively.¹² Since the most common posterior mediastinal lesions are neurogenic tumours, MRI of the chest are frequently performed preoperatively. The tumour is usually of isointense or hyperintense on T1-weighted images and heterogeneously hyperintense on T2-weighted ones.⁵

Surgery is the treatment of choice for a localised HV subtype and offers 100% cure in this group of patients. Although there are reports of video-assisted thoracoscopic resection of posterior mediastinal CD, the majority of these tumours are removed by thoracotomy. The hypervascularity of the tumour and occasional tight adhesions to the chest wall may make thoracoscopic resection less feasible. Although surgical resection offers a near 100% cure in patients with unicentric CD, there are documents of cases that were treated by radiation alone or in combination with surgery because of either incomplete surgical resection of the primary tumour or when the patients were poor surgical candidates. In such cases, total radiation doses of 40–50 Gy (single daily dose of 1.8–2 Gy) were used successfully. However, late complications such as oesophageal and tracheal stenosis are not uncommon and patients need to be followed up carefully.^{13–16}

Other clinical subtypes of the tumour, including MC disseminated disease, localised CD of mixed cell type and HH-8/HIV-associated CD, are less amenable to surgery and are treated by chemotherapy. Occasionally, surgery may be necessary to relieve the patient's symptoms, for example, a painful splenomegaly. The chemotherapy regimen for CD resembles that of

lymphoma. Mild cases may respond to monotherapy with cyclophosphamide, vinblastine or etoposide. In more severe cases, a combination therapy with cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) is recommended. However, caution must be exercised when using such a regimen for patients with HIV because of probable interactions with antiretroviral drugs and an increased risk of infection.¹⁷ There are also reports of sustained remission associated with monoclonal antibodies such as rituximab (against CD20) and altizumab (against interleukin-6 receptor) either in patients infected with HIV or in patients not having HIV. These antibodies may be used as monotherapy in less aggressive disease or in combination with other chemotherapy drugs such as etoposide for more aggressive cases. Rituximab has been used in doses of 375 mg/m² weekly for 4 weeks.^{18 19} However, to date, the data are insufficient to recommend antiherpes drugs in the treatment of HH8 associated CD.

Learning points

- Castleman's disease (CD) may present as a mediastinal mass.
- Surgery is the treatment of choice for a localised mediastinal CD.
- Mediastinum is the most common location for unicentric Castleman's disease.

Competing interests None.

Patient consent Obtained.

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