Case report

Castleman’s disease: unusual intrathoracic localization

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Abstract

Chest-wall is a rare localization of Castleman’s disease. The tumour is often diagnosed after onset of non-specific thoracic symptoms but can be occasionally detected in asymptomatic patients. Surgical removal is curative and should be conservative with no recurrences. We report a new case and we review the international literature.

Keywords: Castleman’s disease; Mediastinal tumours; Surgical excision

1. Introduction

Giant lymph node hyperplasia (Castleman’s disease) is a disease characterized by a peculiar form of lymph node hyperplasia [1].

It can be found whenever lymph nodes are present but 71% of the cases are located in the chest, along the tracheobronchial tree in the mediastinum or lung hilus but it also can occur in other spaces: it also has been reported in the pelvis, neck, retroperitoneum and muscle [2].

We reported the sixth case from the literature of Castleman’s disease from the intercostal space, and we review the literature.

2. Case report

A 36 year old Caucasian female teacher was referred to us after the incidental finding during radiographs carried out for routine occupational control of an homogeneous, well-marginated left paraortic opacity of the chest. She presented to hospital without symptoms. She was a lifelong non-smoker and denied any important remote pathologies.

On examination no pathological signs were noted. Laboratory investigations showed normal blood count and full biochemical screen.

The chest X-ray and computed tomography (CT) scan of the chest (Fig. 1) confirmed a left-sided paraortic opacity of 6 × 5 × 4 cm diameter. The lesion presented as densely enhancing, homogeneous, well-marginated soft-tissue mass located in the left costovertebral sulcus; minimal quantity of pleural fluid was detected. The differential diagnosis of this lesion was either a lymphoma, a neurofibroma or a soft-tissue tumour.

Fine-needle aspiration biopsy was carried out but no diagnostic conclusion was obtained: cytology revealed only blood-stained fluid with no evidence of malignant cells.

Left lateral thoracotomy was performed: 200 ml of pleural fluid had cumulated; they were aspirated and sent for cytology and microbiology. Inspection of the left hemithorax confirmed a 6 × 4.5 × 4.5 cm capsulated mass at the costovertebral sulcus covered by the parietal pleura, extending from the third to the fifth intercostal space and firmly adherent to the chest-wall. No pleural adhesions between the tumour and lung surface were detected. Partial parietal pleurectomy was performed and the mass, totally extrapleural, was completely excised with soft surrounding tissues. The lesion showed hypervascular feature and conspicuous bleeding had to be controlled: many vessels arising from the intercostal arteries had to be clipped or ligated. No perioperative or postoperative complications occurred and the patient was discharged on day 6.

Microscopically the mass had morphologic features of the classic hyaline vascular variety of Castleman’s disease with small to medium sized lymphoid follicles. Inter-follicular stroma was characterized by numerous hyperplastic hyalized small vessels and an admixture of plasma cells, eosinophils and immunoblasts. Cytology and cultures of the pleural fluid were all negative.
3. Discussion

Castleman’s disease is an eponymous which was first given to a lymphoid tumour of the mediastinum [1] also referred as lymph node hyperplasia, or angiofollicular lymph node hyperplasia [2,3]. Although the majority of lesions occur within the chest, less commonly other sites including neck, pelvis, retroperitoneum and axilla may be involved [2].

There are no significant sex predominance or identifiable risk factors in the development of the disease [2]. Three histological types have been described: hyaline vascular, plasma cell and mixed type [4]. Patients are usually asymptomatic or have non-specific complaints. Symptoms are apparently due to tracheobronchial compression such as cough, dyspnoea, chest pain, respiratory infection and back pain [2].

Chest-wall involvement by Castleman’s disease is a rare manifestation of this pathology and the international literature reports five cases during the last eleven years, in addition to our current observation: we review about these six cases [5–9] (Table 1).

Radiological evaluation detected in every case an homogeneous well-marginated highly vascularized mass densely enhancing, associated with massive pleural effusion in two cases [6,9]; one case was characterized by a little intratumoural calcification [5].

Preoperative aspiration biopsy was carried out in four

<table>
<thead>
<tr>
<th>Author</th>
<th>Sex</th>
<th>Age</th>
<th>Presentation</th>
<th>Surgery</th>
<th>Diameter (mm)</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Our (2001)</td>
<td>F</td>
<td>36</td>
<td>IRF</td>
<td>Thoracotomy</td>
<td>60</td>
<td>H-V</td>
</tr>
</tbody>
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*IRF, incidental radiological finding; H-V, hyaline-vascular.
cases and resulted inconclusive in all: all patients underwent surgical exploration and subsequent removal of the mass by thoracotomy or median sternotomy; one patient was submitted to partial parietal pleurectomy because of multicentric diffusion of the tumour [6].

Microscopic examination after surgery revealed unique localization of hyaline vascular variety of Castleman’s disease in five cases (83%) [5,7±9] and multicentric mixed variety in the other [6]. Pleural effusion was noted both in the hyaline vascular and mixed type. Cytology and cultures of the pleural fluid were negative in both patients referred in the literature [6,9] and in our current case. Development of pleural fluid seemed not related to the tumour’s diameter: at surgery the tumours had quite the same dimensions but only three patients developed pleural effusion.

Symptomatic release occurred after surgery in all cases, all patients survived surgery and no authors referred about recurrences or malignancies after excision of Castleman’s disease involving the chest-wall.

Chest-wall localization is a rare manifestation of Castleman’s disease often diagnosed after onset of non-specific thoracic symptoms such as dyspnoea, cough, chest-wall pain or generalized malaise; occasionally patients could be asymptomatic.

Tissue diagnosis is mandatory to avoid mismanagement: needle biopsy has low diagnostic accuracy and thorascopic biopsy is dangerous because of the high vascularization of the tumour increasing risk of bleeding.

Surgical removal managed with care to avoid immediate or perioperative bleeding is probably the best diagnostic and also curative procedure: symptoms disappear after surgery. Radical excision is mandatory but often the tumour is not easily removed from the underlying tissues: some subtotal excision had been performed without short-term recurrences reported [6].

References