Localized Castleman’s disease

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Section: Chest imaging
Area of Interest: Thorax
Procedure: Diagnostic procedure
Procedure: Screening
Imaging Technique: CT
Imaging Technique: Conventional radiography
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 44 years, female

Clinical History:

Female patient, 44 years old, asymptomatic, with no relevant history of disease underwent a routine thoracic radiography.

Imaging Findings:

Chest radiography showed a nodular medium-density opacity, located in the superior segment of the left lower lobe (Fig. 1).
To further investigate the mass, chest CT was performed. CT showed a well-defined paravertebral mass. The mass showed a rough central calcification and enhanced homogeneously and avidly after intravenous administration of contrast material. The mass invaded the adjacent rib (Fig. 2).
The patient underwent mediastinoscopy and surgical biopsy was obtained.
The histological examination classified the lesion as hyaline vascular Castleman’s disease.
A successful surgical resection was performed.

Discussion:

Castleman’s disease, also known as angiofollicular or benign giant lymph node hyperplasia, is a rare benign lymphoproliferative disorder of unknown etiology [1-3].

It can be classified morphologically in unicentric/localized and multicentric/disseminated disease based on the extent of local lymph node involvement. Histopathologically two major categories can be distinguished: hyaline vascular, accounting for 90% of cases and plasma cell type. The former manifests, in approximately 90% of cases, as unicentric disease and the latter usually shows multicentric involvement [1-4].

Pathologically the hyaline vascular type is characterized by lymph node hyperplasia, involuted germinal centers and capillary proliferation with endothelial hyperplasia.

Hyaline vascular type occurs predominantly in young adults (3rd or 4th decade) with female predominance and has a significant predilection for the thorax, especially mediastinum and hilum but unusual locations can be involved, including pericardium, pleura, intercostal space and lung [3-5].

The typical radiographic appearance of hyaline vascular Castleman disease is an asymptomatic solitary rounded
mediastinal or hilar mass lesion [2-6].

The classic CT findings include a solitary mass or lymphadenopathy with avid homogeneous enhancement after intravenous administration of contrast material.

Based on CT, three different patterns have been described: 1) a solitary noninvasive mass; 2) an infiltrative mass with lymphadenopathy in the same compartment and 3) lymphadenopathy in a single compartment without mass. Characteristic hypervascularity of lesions is verified by their homogeneous and intense enhancement after administration of contrast material. Indeed the mass may show feeding vessels in its proximity. About 10% of the lesions can show internal calcifications, which are typically coarse and central or branchlike. Systemic involvement of hyaline vascular type is uncommon compared with plasma cell type [1, 3-8].

Mediastinal masses may simulate lymphoma, sarcoma, thymoma and neurogenic tumors, although hilar masses can mimic bronchial adenomas. Pericardial disease may resemble pericardial cyst. Intercostal disease may show rib erosion and can be confused with other chest wall masses. Pleural disease can manifest as pleural effusion or well-defined interlobar mass [4-8].

Definitive preoperative diagnosis usually requires mediastinoscopy or surgical biopsy.

Complete surgical resection is the gold standard for unicentric disease. If it is not possible or if multicentric involvement is present, radiotherapy and/or steroid therapy may be indicated [3-5].

**Differential Diagnosis List:** Hyaline vascular Castleman's disease, Neurogenic tumours (ganglioneuroma; paraganglioma; nerve sheaths tumours), Lymphoma, Neurenteric cyst, Meningocele, Extramedullary haematopoiiesis

**Final Diagnosis:** Hyaline vascular Castleman's disease

**References:**


Description: Axial contrast-enhanced CT shows a well-defined hypervascular left paravertebral mass, which enhances avidly after intravenous administration of contrast material. Origin: Department of Radiology, IPO Porto.
Description: The mass shows central rough calcification and invasion of the adjacent rib. Origin: Department of Radiology, IPO Porto
Figure 2

Description: The posteroanterior radiography of the chest shows nodular medium-density opacity obliterating the left upper heart edge. Origin: Department of Radiology, IPO Porto, Portugal
Description: The lateral radiography of the chest shows nodular opacity located in the superior segment of the left lower lobe. Origin: Department of Radiology, IPO Porto, Portugal