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Mediastinal Castleman's disease : report of case

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Abstract: *Castleman's disease, a nonclonal lymphoproliferative disorder, is one of the most common causes of nonneoplastic lymphadenopathy. Because of its wide variety of manifestations and ability to affect any part of the body, Castleman's disease closely resembles benign and malignant abnormalities of the neck, chest, abdomen, and pelvis. Castleman disease most commonly presents as unicentric Castleman disease with nodal mass enhancement and should be considered in the differential diagnosis of lymphoma, metastatic lymphadenopathy, and infectious and/or inflammatory disorders causing lymphadenopathy to consider. We present a case report of a patient who presented non-specific respiratory symptomatology of cough and dyspnea. A chest CT scan revealed a single right mediastinal-hilar mass, with intense enhancement after contrast and without any other lesions. The anatomopathological examination revealed the hyaline-vascular subtype of Castleman's disease.*

Keywords: *Castleman, disease, CT scan, unicentric form, mediastinal*

1. INTRODUCTION

Castleman's disease (CD) is a rare benign lymphatic disorder with a variable clinical course. It usually affects the mediastinum but can occur anywhere there is lymphoid tissue. It was first described in 1956 by Dr. Benjamin Castleman. The two major histologic subtypes of CD are hyaline vascular and plasmacytic, and the two clinic radiological entities are unicentric and multicentric CD (1). This condition is difficult to diagnose preoperatively, and an accurate diagnosis is usually only made after surgical removal of the tumor (2).

2. CASE PRESENTATION:

We report the case of a 35-year-old patient, without any previous history, a chest X-ray was performed as part of a pre-employment check-up, which revealed a right para-hilar opacity.
An interrogation and a clinical examination were carried out and returned negative.
The patient was lost to follow-up for 10 years, and returned to the pneumology clinic with non-specific respiratory symptomatology of cough and dyspnea, of moderate intensity.
The clinical examination was normal.
A chest CT scan revealed a single right mediastinal-hilar mass, well-limited with intense enhancement after contrast and without any other lesions (figure1)
The CT characteristics of the mass led to three main diagnoses: a pulmonary artery aneurysm, a right hilar lymphadenopathy, or a carcinoid tumor.
Bronchoscopy and endobronchial biopsy were inconclusive.
The case was discussed at a multidisciplinary consultation meeting, leading to the decision to perform a surgical removal.
The anatomopathological examination revealed Castleman's disease.

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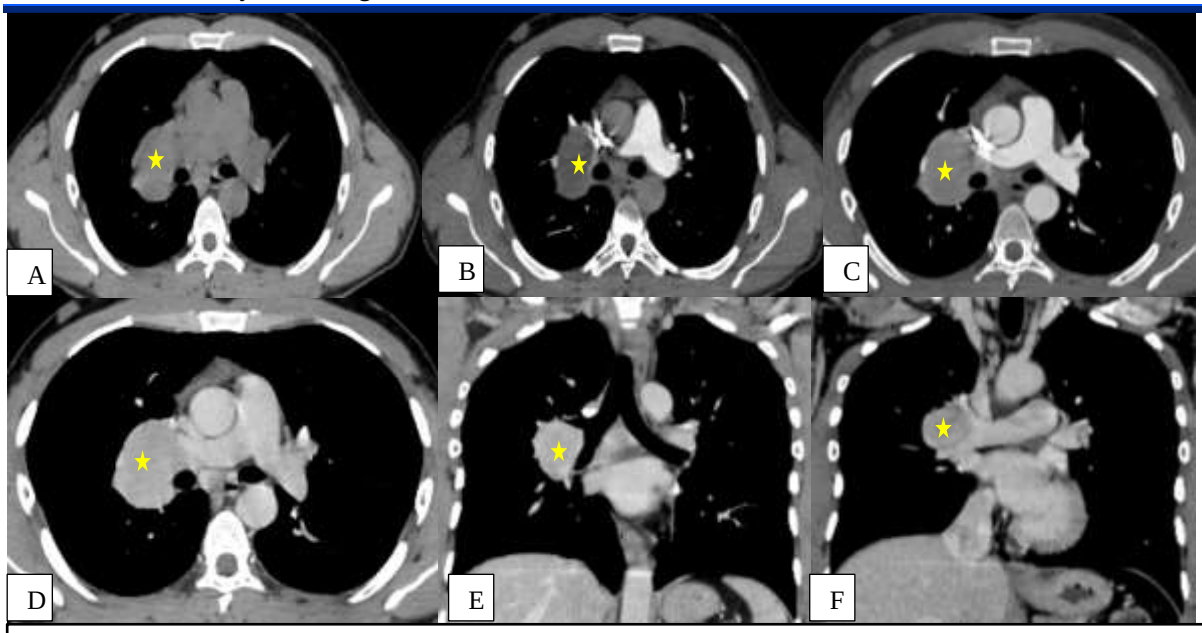

Figure.1 : Axial (A, B, C, D) and coronal (E, F) chest CT showing a single right mediastinal-hilar mass (Star), well-limited with intense enhancement after contrast.

3. DISCUSSION

Castleman's disease can affect anyone from adolescence to their seventies, with an equal gender distribution. All systemic lymph nodes including cervical (42%), mediastinal (31%), intra-abdominal (18%), and retroperitoneal (5%) lymph nodes were reported to be involved. Only 5% have extranodal presentation (3).

Three forms of the disease have been described histologically; the hyaline vascular form is usually seen in the mediastinum (approximately 90% of cases), the plasmacytic form affects extrathoracic sites, and the rare mixed form. Clinically, the CD can present as a benign, localized, resectable disease, as in our case, or as an aggressive multicentric variant with systemic symptoms and recurrence after treatment (4).

Typical CT findings in hyaline-vascular Castleman disease are single enlarged lymph nodes or focal nodular masses that show homogeneous strong enhancement after administration of contrast medium. Three patterns of involvement have been described, including solitary non-invasive mass (most common: 50% of cases), overtly invasive mass with associated lymphadenopathy (40% of cases), and dull lymph nodes without obvious mass Enlargement (10% of cases. cases) (5).

Castleman's hyaline vessel disease easily involves the chest and usually presents with greatly increased mediastinal lymphadenopathy. Mediastinal Castleman disease can mimic thymoma, lymphoma, sarcoma, hemangiopericytoma, paraganglioma, neurofibroma or schwannomas, and chest wall tumors. Hilar-Castleman disease may be mistaken for bronchial adenoma (6).

Approximately 10% of lesions have internal calcifications characterized by roughness or a pronounced branching pattern. However, nonspecific calcifications were observed more frequently (7).

On magnetic resonance imaging (MR), hyaline vascular Castleman's disease lesions often show heterogeneous T1 and T2 hyperintensity compared with skeletal muscle. The unique flow chamber can be seen to identify the feed container. MRI imaging is good at showing the extent of disease and its relationship to adjacent structures but has a limited assessment of calcification (8).

Castleman disease shows mild to moderate FDG uptake on PET scans. Histological diagnosis before surgical resection can be made by CT-guided biopsy (3).

Surgical resection is the method of diagnosis and treatment of unicentric Castleman's disease. However, corticosteroid therapy, chemotherapy, and monoclonal antibody therapy are indicated for multicentric Castleman disease. Surgical resection of the monocentric hyaline vessel type may not be easy due to its large vascularity. It can be associated with excessive bleeding during resection, and pneumonectomy due to massive bleeding has been reported (9). Preoperative embolization of the feeding artery may be considered to avoid intraoperative bleeding.

Surgical resection of unicentric Castleman's disease has an excellent prognosis, with a 5-year survival rate of 100% (10). In contrast, multicentric Castleman disease has a poorer prognosis (11).

4. CONCLUSION

In conclusion, Castleman's disease encompasses a wide range of pathological findings, presentations, and associations. Castleman disease most commonly presents as unicentric disease with hyperenhancing nodal masses. Next, an excisional biopsy is preferred to differentiate it from lymphoproliferative disease, metastases, and other hypervascular masses.

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