Unicentric castleman's disease: an uncommon cause of chronic cough

Maladie de Castleman unicentrique : une cause rare de toux chronique

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RÉSUMÉ

La maladie de Castleman encore dénommée hyperplasie lymphoïde angio-folliculaire est une affection très rare, caractérisée par une hyperplasie lymphoïde ganglionnaire. La maladie de Castleman peut se développer sous deux formes : l'une localisée ou unicentrique bénigne, et l'autre multicentrique de pronostic généralement réservé. Nous rapportons le cas d'une maladie de Castleman unicentrique intrathoracique chez une patiente âgée de 50 ans, non tabagique révélée par une toux sèche chronique. Le scanner thoracique avait montré une masse hilaire droite homogène, se rehaussant de façon intense après injection de produit de contraste. Une lobectomie supérieure droite associée à un curage ganglionnaire était réalisée. L'étude histologique avait conclu à une maladie de Castleman de type hyalino-vasculaire. Le suivi clinique et radiologique n'avait révélé aucune récidive sur une période de 6 mois et la patiente demeurait asymptomatique.

Mots-clés

Maladie de Castleman ; intrathoracique ; unicentrique; hyperplasie ganglionnaire angiofolliculaire; toux chronique

SUMMARY

Castleman disease (CD) or angiofollicular lymph node hyperplasia is a rare lymphoproliferative disorder characterized by lymph node hyperplasia of uncertain etiology. CD is divided clinically into unicentric (localized to one region of the body) considered as a benign disease and multicentric with less favourable prognosis. We describe a case of intrathoracic unicentric CD revealed by a chronic non-productive cough in a 50-year-old non-smoker female. Chest computed tomography revealed a bulky right hilar mass with intense homogenous contrast enhancement. The patient underwent a right upper lobectomy and mediastinal lymphadenectomy. Histopathology was consistent with hyaline-vascular (HV) type CD. The patient remained asymptomatic throughout the subsequent 6-months of follow-up.

Key-words

Castleman disease; intrathoracic; unicentric; angiofollicular lymph node hyperplasia; chronic cough

Castleman disease (CD) or giant lymph node hyperplasia is a rare lymphoproliferative disorder characterized by localized lymph node hyperplasia of uncertain etiology (1). CD is divided clinically into unicentric (localized to one region of the body) and multicentric (involved lymphoid structures in more than one region). Furthermore, there are three histological subtypes: the hyaline vascular (HV) type (90% of cases), the plasma cell (PC) type (8-9% of cases) and the mixed type (1-2% of cases) (1, 2). We describe a case of unicentric CD reveled by chronic cough in a non-smoking 50-year-old female.

CLINICAL CASE

A non-smoking 50-year-old female initially presented to our department with a history of chronic non-productive cough. She denied any history of fever, weight loss, or fatigue. Physical examination showed no enlargement in superficial lymph nodes. Laboratory results were within normal limits. HIV-serology was normal. Chest radiography showed enlarged right hilum. Flexible Bronchoscopy showed no abnormalities.
Chest computed tomography (CT) revealed a 4.8-cm bulky right hilar mass with intense homogenous contrast enhancement (figure 1).

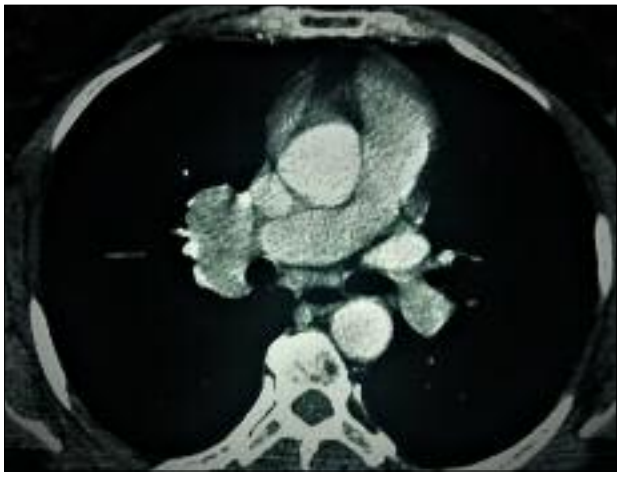

Figure 1: Chest computed tomography revealed a well-defined and homogenous enhanced mass in the right hilum.

Chest magnetic resonance imaging (MRI) revealed T2 hyperintense lesion on T2-weighted images and avid enhancement on contrast T1-weighted fat-saturated sequence images (figure 2 a, b, c). Thoracotomy was performed and a 7 cm hypervascular mass was identified in the hilar region of the right upper lobe (Figure 3). The patient underwent a right upper lobectomy and mediastinal lymphadenectomy. No complications were observed in the postoperative period and the patient's cough dissolved. Histopathology was consistent with HV type CD characterized by lymphoid follicular hyperplasia with interfollicular vascular proliferation (figure 4). On immunohistochemical staining, the hyperplastic cells were positive for CD3, CD20. Histopathology of resected mediastinal lymph nodes was inflammation. The patient remained asymptomatic throughout the subsequent 6-months of follow-up and her initial coughing did not recur.

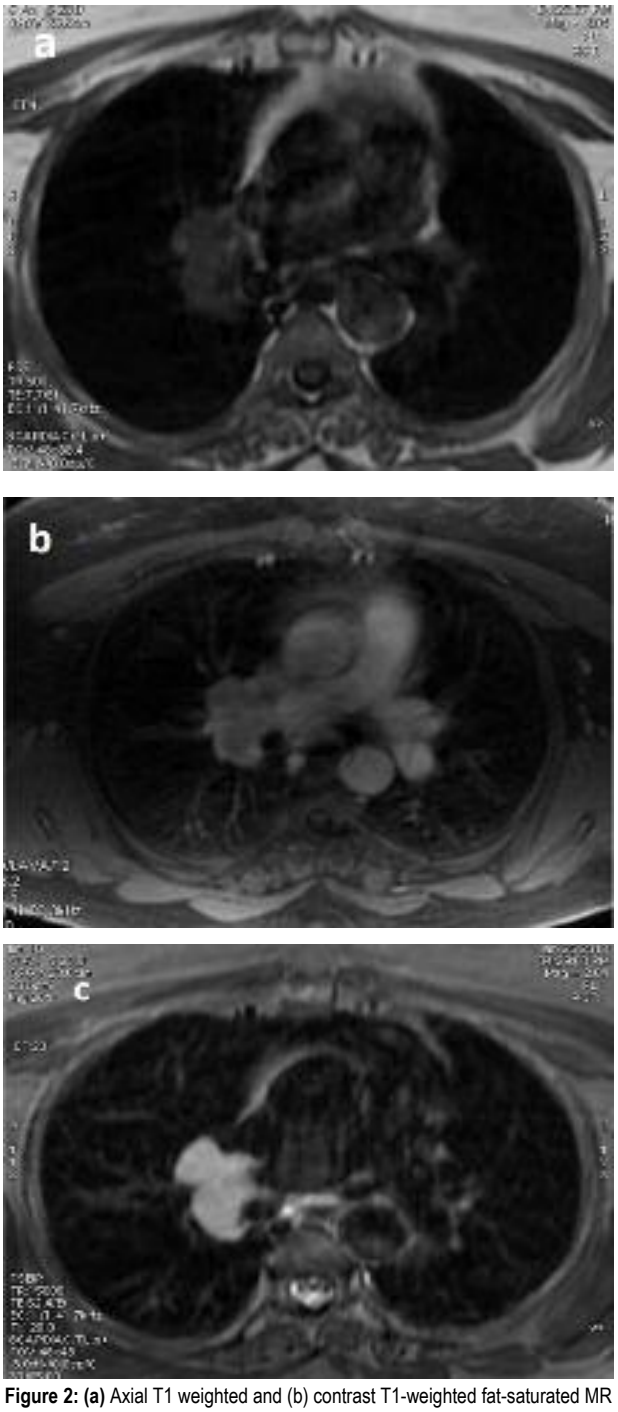

Figure 2: (a) Axial T1 weighted and (b) contrast T1-weighted fat-saturated MR imaging demonstrates the right hilar mass with avid enhancement. (c): Axial T2 weighted MR imaging shows homogenous T2 hyperintensity.

Figure 3: Photograph of right upper lobectomy specimen shows a 7 cm Castleman's disease mass with prominent vasculature.

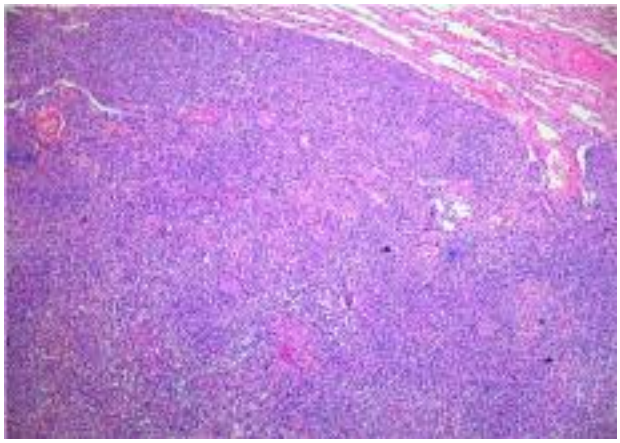

Figure 4: Pathological micrograph showed follicular hyperplasia with involuted germinal centers and marked interfollicular vascular proliferation consistent with hyaline-vascular Castleman's disease (haematoxylin and eosin staining; x400).

DISCUSSION

The unicentric variant of CD is the most common type and frequently localized to the mediastinum. Other intrathoracic locations (pleura, lung parenchyma, and chest wall), extrathoracic lymph nodes, abdomen, pelvis and head may be involved (3). In adults, 90% of unicentric CD cases are HV-type and predominantly occurs in young female patients. HV follicles with interfollicular lymphocytes and capillary proliferations characterize HV-type CD. Unicentric CD is often asymptomatic and diagnosed incidentally. Nevertheless, nonspecific symptoms related to local compression by the CD mass such as cough, chest pain, dyspnea and palpitations were sometimes reported (1-6). The diagnosis is challenging and can mimic lymphoma, thymoma, sarcoidosis or metastatic tumor (7). Multicentric CD is less common (10-20% of cases), frequently PC-type and occurs in an older age population. Histologically, large follicles with intervening sheets of plasma cells characterize PC-type CD. Lymphadenopathies, hepatomegaly, splenomegaly, fever, fatigue, and weight loss are the common clinical features. Abnormal laboratory findings including anemia, elevated erythrocyte sedimentation rate, thrombocytopenia, abnormal liver function and hypoalbuminemia may be observed. Thoracic imaging shows hypervascular lesion with intense enhancement on contrast CT scan due to the abundance of blood vessels, where a feeding artery may occasionally be seen (7,8). A central hypodensity and calcifications can also be seen on CT (8). On MR imaging, lesions are hyperintense on T2-weighted images with avid enhancement on contrast sequence and prominent flow voids reflecting the vascularity and feeding vessels on T1- and T2-weighted images. The imaging features are correlated with the histological type, with Homogenous high enhancement typically observed in HV-type CD due to the abundance of blood vessels. However, PC-type CD is typically associated with absent or mild homogenous enhancement (8). Unicentric CD is considered a benign disease. Nevertheless, there are few reports of lymphoma and solitary plasmocytoma associated with unicentric CD. Malignancies including Kaposi sarcoma, plasmocytoma, non-Hodgkin lymphoma, glomeroid hemangioma and carcinomas of the colon, kidney and thyroid were reported in the multicentric CD. An excisional biopsy is recommended for the pathological diagnosis of CD and is useful to rule out differential diagnosis such as lymphoproliferative disorders, metastatic disease and other hypervascular masses. Complete resection is the gold standard for the treatment of unicentric CD and is curative in most of cases with an excellent prognosis (6,9). Therapeutic options are chemotherapy, radiotherapy, interferon, corticosteroids, anti-IL6 receptor and anti-CD20 antibody (rituximab) for multicentric CD. The prognosis is poor with a median survival ranging from13 to 30 months (10).

Conclusion: CD should be considered when a single persistently enlarged node, nodal mass or multistation lymphadenopathy is associated with moderate to intense post contrast enhancement and especially when data indicated a low probability of malignancy in a low-risk population group. In addition, unicentric CD presents commonly as an asymptomatic or incidental thoracic mass but nonspecific symptoms related to compression by the CD mass such as chronic cough in this case could be rarely seen. Curative surgical resection is the treatment of choice for unicentric CD.

Disclosure of interest: The authors declare that they have no competing interest.

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