UNICENTRIC CASTLEMAN DISEASE: AN UNUSUAL LOCALIZATION IN THE BREAST

MALADIE DE CASTELMAN MONO-CENTRIQUE : LOCALISATION INHABITUELLE DANS LE SEIN

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Abstract

Castelman disease (CD) is a rare benign lymphoproliferative disorder. Lymph node involvement is common, particularly in the mediastinum. Extranidal localizations are rare. We report the case of an unusual localization in the breast of CD hyalinized vascular type occurring in a 19 years old healthy female. We discuss also clinical findings and treatment options for this disease.

Key words: Castelman ; Breast ; Women.

Résumé

La maladie de Castelman, est une affection rare, caractérisée par un désordre lympho-prolifératif bénin. La localisation ganglionnaire est classique, particulièrement au niveau du médiastin. Les localisations extra-ganglionnaires sont rares. Nous rapportons le cas d'une localisation inhabituelle au niveau du sein d'une maladie de Castelman type hyalino-vasculaire chez une femme de 19 ans sans histoire pathologique connue. Nous discutons aussi les aspects cliniques et les options thérapeutiques de cette maladie.

Mots clés : Castelman ; Sein ; Femme.

ملخص

يعتبر داء كاستيلمان هو حالة نادرة تتميز باضطراب تكاثري ليمفاوي حميد. موضع العقدة هو كلاسيكي، ولا سيما في المنصف الصدري. أما التموقع خارج العقدة هو نادر. قمنا بالتبليغ عن حالة مرض بموقع الثدي غير المعتاد لمرض كاستلمان من النوع الزجاجي و الوعائي الدموي لدى امرأة تبلغ من العمر 19 عامًا بدون سوابق مرضية معروفة.
نناقش أيضا الجوانب السريرية والخيارات العلاجية لهذا المرض.

الكلمات المفاتيح : كاستيلمان ; الثدي ;الإمرأة.

INTRODUCTION

Castleman disease (CD), or angiofollicular lymphoid hyperplasia, is a rare lymphoproliferative disorder that was first described in 1956 [1].

It may occur anywhere along the lymphatic chain, but it is frequently found in the mediastinum and then in intra-abdominal [2]. The breast is rarely affected, and only few cases have been reported [3, 4].

CD is categorized into two clinical entities with adverse pathological outcomes: unicentric (localized) and multi-centric (systemic) forms. Pathologically, it is classified as hyaline vascular, plasmacytic, or mixed cellularity types. The hyaline vascular type is found in most unicentric cases with a favorable prognosis. The systemic form is mainly a plasma cell (PC) type with poor prognosis [3]. We present the fifth published case of localized breast CD in our knowledge.

CASE REPORT

A 19-year-old female patient presented with a 6-month history of a right-breast nodule discovered by self-examination. Physical examination revealed the presence of a mobile mass situated in the superolateral quadrant measuring approximately about 40 mm diameter. The patient had no other systemic symptoms or abnormal clinical findings.

Breast-Ultrasound revealed well-defined rounded, solid nodule in the superolateral quadrant of the right breast, hypo echoic with a hyper echoic span. Sonographic findings were compatible with fibroadenoma (Fig 1).

The resection of the breast nodule and its histological study showed an angio-follicular hyperplasia, also known as giant lymphoid hyperplasia or CD of the hyalinized vascular form (CD/HVF) (Fig 2).

Laboratory analyses were normal. Thoracic and abdominal CT showed absence of another lymphadenopathy. Along 6 months follow-up, the patient was well and free of local or distant lymphadenopathy.

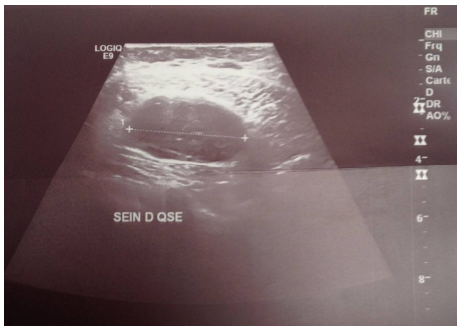

Fig 1: Rounded hypoechoic solid nodule with hyperechoic span.

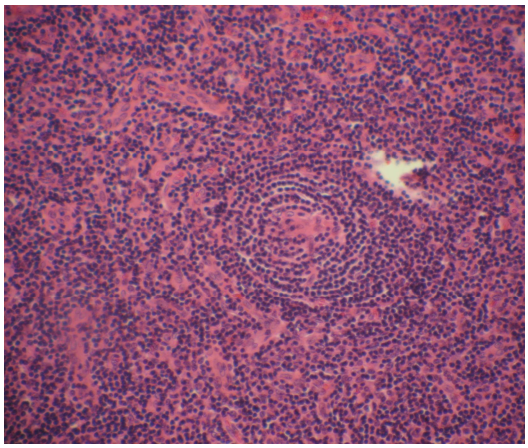

Fig 2: Angiofollicular hyperplasia

DISCUSSION

Castleman disease (CD), or angio-follicular lymphoid hyperplasia, is a rare lympho-proliferative disorder that was first described in 1956 [1]. It occurs equally for women and men, and it was divided into two subgroups: the unicentric form is more common and the multicentric form is less common. Unicentric and multicentric CD differ in their clinical presentation and their distribution of involved lymph nodes [5].

The etiology and pathogenesis are still not completely elucidated. But the immunologic mechanism hypothesis including over-production of IL-6 and human herpes virus type 8 infection (HHV8), is commonly supported by most authors. The main point in CD pathogenesis is that reactive follicular hyperplasia is a response to an unknown antigenic stimulus [6, 7].

CD was divided into two subgroups based on its histology, the hyalinized vascular type and the plasma cell type [8]. The first subtype is characterized by numerous small- to medium-sized

germinal follicles in the lymph nodes; the hyalinized vessels with the germinal follicles and a concentrically arranged mantle zone producing a characteristic 'onion-peel' appearance is usually found in unicentric CD. The histological appearances of the plasma cell type, typical of multicentric CD, are of an intense plasmacytosis in the inter-follicular areas of the nodes, again with a prominent increase in capillaries and post-capillary venules, which may be hyalinized. Plasma cells are identified by their clock-face nucleus and pale perinuclear cytoplasmic crescent. Mixed forms of CD exist with both hyaline-vascular and plasma cell elements present [8].

Clinically, unicentric CD tends to be present in the form of an enlarged, benign, painless lymph node that generally remains asymptomatic unless it begins to compress adjacent structures or is discovered fortuitously at the time of a routine physical examination [2].

Extra nodal CD, especially when localized in the breast, is extremely rare [3, 7]. Breast localization is clinically related to intra- mammary lymph node (ILN) disease or interpectoral lymph node with axillary involved lymph nodes [3, 9]. Breast CD differential diagnoses include inflammation, reaction to dermatitis, tuberculosis, lympho-proliferative disease, and breast cancer; that means that immunohistochemistry may be required to confirm the diagnosis. Chang et al. published a case report of a ductal-carcinoma in situ with micro-invasion associated with a synchronous axillary CD [10].

The pre-operative diagnosis of CD is still very difficult. The diagnosis of CD is established by biopsy with histological study, and treatment is often based on published case reports only. The best therapeutic approach remains controversial, and many treatment regimens have been proposed, including surgery [11].

For Unicentric CD complete surgical removal remains the best option for better prognosis and a long-term follow-up to detect disease progression and complications. Multi-centric CD has a poor prognosis, and many treatments have been discussed such as cortico-therapy, poly-chemotherapy and immuno-therapy [11].

CONCLUSION

CD is a rare and poorly understood disease that created both diagnosis and therapeutic dilemmas for physicians and researchers. Breast unicentric CD is present in the form of asymptomatic nodule.

Only histological exam can improve diagnosis. Surgical treatment is usually curative for the breast and seems to be the most important prognosis factor.

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