Massa Esternal: Um Caso Raro de Linfoma Hodgkin Primário do Osso

Sternal Mass: A Rare Case of Primary Hodgkin Lymphoma of The Bone

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Resumo:

As massas esternais são lesões que constituem um verdadeiro desafio na prática clínica. Apresentam vários diagnósticos diferenciais que por vezes dificultam a abordagem do linfoma de Hodgkin ósseo primário, sendo frequentemente diagnosticado como tuberculose ou granuloma eosinofílico. Os autores relatam o caso de um doente de 32 anos, com dor torácica esternal inespecífica, com duração aproximada de um ano, como apresentação inicial de linfoma de Hodgkin primário do esterno, tratado com quimioterapia, sem recidiva da doença após dois anos.

Os autores apresentam este caso para sensibilizar os médicos sobre a importância dos estudos imuno-histoquímicos no estudo de massas esternais, uma vez que permitem um rápido diagnóstico diferencial e consequentemente uma melhoria substancial do seu prognóstico, apesar da raridade do linfoma de Hodgkin primário do esterno.

Palavras-chave: Doença de Hodgkin; Dor; Neoplasias do Osso.

Abstract:

Sternal tumour lesions remain a challenge in clinical practice. Due to multiple etiologies, it can be difficult to diagnose primary bone Hodgkin lymph, often misdiagnosed as tuberculosis or eosinophilic granuloma. The authors report the case of a 32-year-old patient with non-specific sternal chest pain that had lasted approximately one year as the initial presentation of primary Hodgkin lymphoma of the sternum, treated with chemotherapy, without recurrence of the disease after two years.

The authors present this case to raise awareness among clinicians about the importance of immunohistochemical studies in the differential research of sternal masses, allowing a rapid differential diagnosis and consequently a substantial improvement in the prognosis of these patients, despite the rarity of primary Hodgkin's lymphoma of the sternum.

Keywords: Bone Neoplasms; Hodgkin Disease; Pain.

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https://doi.org/10.60591/crspmi.211

Introduction

Sternal tumour lesions are rare, with an incidence of 0.65%, almost one sixth of chest wall tumors.[1] The sternum is an uncommon site for neoplastic involvement (whether benign or malignant) and metastases are much more common than primary neoplasms, which are frequently malignant.[1,2]

Sternal masses remain a challenge in clinical practice, due to a myriad of possible diagnoses, such as osteomyelitis, osteosarcoma, intrathoracic thyroid thymoma, germ cell tumours, metastatic bone tumour, tuberculosis, eosinophilic granuloma and lymphomas.[2-5]

Sternal involvement is common in non-Hodgkin's lymphoma but seldom associated with Hodgkin's lymphoma (HL).[2] HL is typically a systemic disease involving multiple lymph nodes, generally painless, with cervical and supraclavicular location.[1,2,4] Extranodal presentation is uncommon. Bone involvement is rarely found at clinical presentation and is generally associated with advanced stages of HL, occurring in 10%-20% of cases. Although there are several reported cases of HL with initial skeletal symptoms, most have concurrent the involvement of other structures (breast, supraclavicular, axillary, lymph node). Thus, isolated bone involvement (primary bone HL) is extremely rare (1%-4%).[4,5] Primary bone HL is characterized by a lymphoma limited to the bone, with or without an associated soft tissue component, without regional lymphadenopathies at the time of diagnosis and six months later. In other words, it behaves like an early stage (IE) according to Ann Arbor and, therefore, has a good prognosis if diagnosed in a timely manner.[3,5]

Nonetheless, as it is a very rare lymphoma and due to the highly complex differential diagnosis, around 40% of cases are incorrectly diagnosed on histopathological examination (tuberculosis or eosinophilic granuloma), delaying treatment, with ominous impact on prognosis.[2,5] In addition to the various imaging tests that allow the etiological study of sternal masses, such as chest radiography, computed tomography enables the evaluation of sternal lesions and magnetic resonance determines the exact extent of bone involvement and mediastinum invasion. Carrying out an immunohistochemical study is essential for the definitive diagnosis of this entity, allowing the exclusion of several entities that commonly lead to diagnostic errors.[3,5]

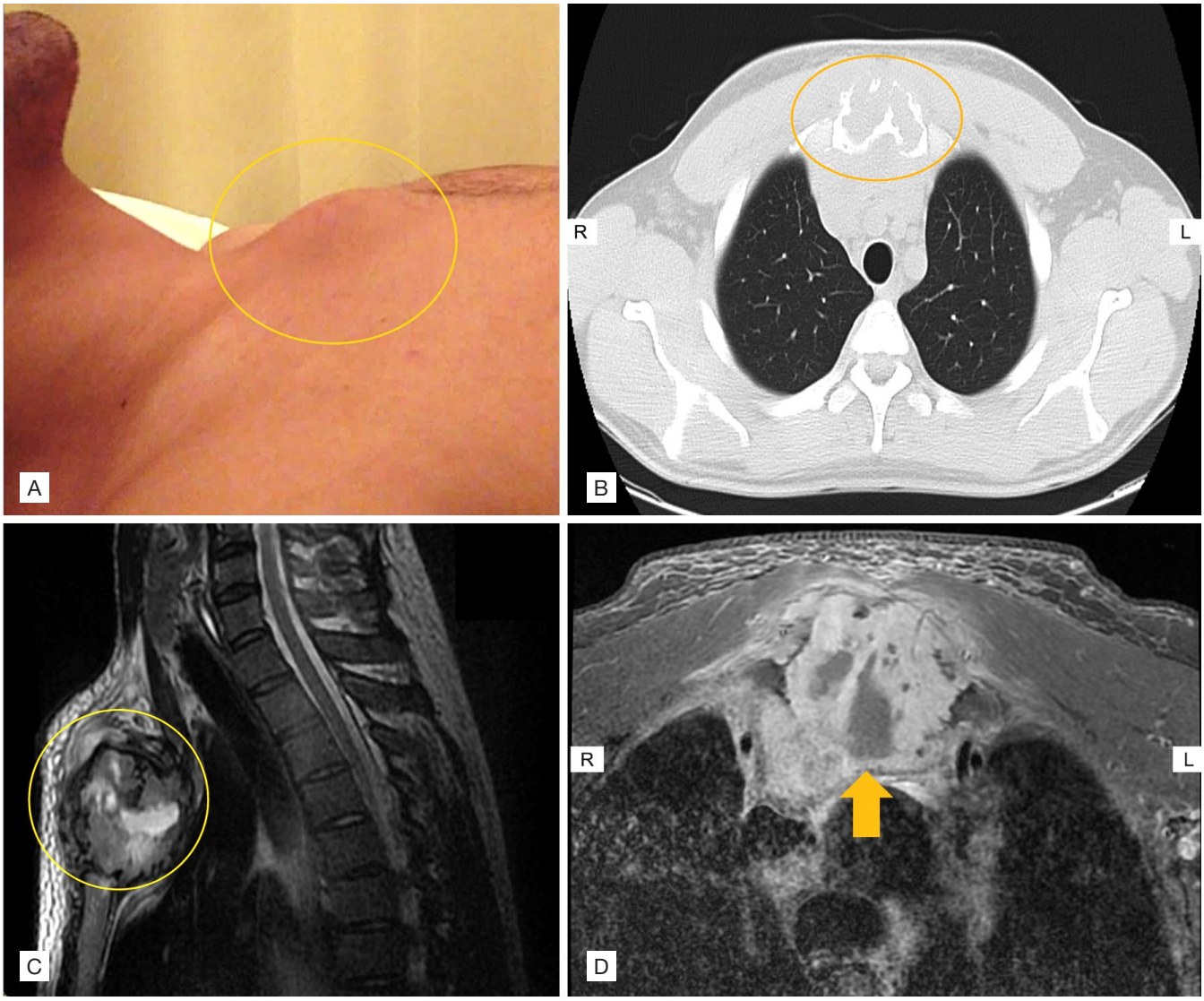

Figure 1: Sternal mass. **A** - Sternal mass detected on physical examination. **B** - Chest computed tomography - sagittal section - an anterior mediastinal soft tissue mass (76x46 mm) with bone invasion and lytic manubrium. **C** e **D** - Lytic mass centered on the sternal manubrium (6 x 5.5 x 5.6 cm), destructive, which ruptures the cortex and is associated with a soft tissue mass in the pre-vascular mediastinum and subcutaneous cellular tissue, longitudinal magnetic resonance image - T1 (C) and sagittal magnetic resonance image - T2 (D).

The authors present a primary bone Hodgkin's lymphoma case whose initial presentation was a sternal mass.

Case Report

The authors present the case of a 32-year-old male with a 1-year history of unspecific chest pain located to the xiphoid appendix. Several tests performed at general practice were unremarkable, and the patient was diagnosed with anxiety and started mirtazapine, with no improvement. In the following six months he noticed swelling over the anterior chest wall that worsened with physical activity and fatigue. He denied night sweats, weight loss or fever. Clinical examination was unremarkable except for his chest, revealing a midline mass of approximately 60 x 50 mm at the sternal manubrium. On palpation, the mass was fixed, firm and non-tender with smooth margins and slight overlying erythema (Fig. 1A). Blood tests revealed only an elevated sedimentation rate of 44 mm. Chest computed tomography showed an anterior mediastinal soft tissue mass (76 x 46 mm), with bone invasion and lytic manubrium, confirmed by magnetic resonance (Figs. 1B, C, D). There was no lymphadenopathy or organomegaly described. Biopsy was performed, and a histopathological study (with immunohistochemistry) revealed HL. The patient achieved complete remission after chemotherapy treatment (hyper CVAD protocol) and was followed for two years without tumour recurrence relapse or clinically detectable lymph node enlargement.

Discussion

The diagnosis of primary HL of the bone requires a high

index of suspicion, and an immunohistochemical study is essential for a definitive diagnosis.[4,5]

In practice, an immediate diagnosis at the early stage of the disease and timely treatment with systemic chemotherapy and local radiotherapy are pivotal, as patients with this presentation usually have an excellent long-term prognosis.[5,6]

However, diagnosing primary HL of the bone can be difficult, as clinical and radiological signs are not specific, histological findings are not always easy to interpret, and the clinician must maintain a high index of suspicion to perform the diagnosis and differential diagnosis of a sternal mass.[3,4]

In this sense, the authors present this case to demonstrate the importance and diagnostic challenge of this type of bone lymphohematopoietic tumours with unusual presentation.[4-6] ■

Declaração de Contribuição

CD, DA, TP, AIR - Elaboração do artigo, conceção, desenho, recolha de dados e revisão da literatura.
AP, JRM - Revisão do artigo.
Todos os autores aprovaram a versão final a ser publicada.

Contributorship Statement

CD, DA, TP, AIR - Preparation of the article, conception, design, data collection and literature review.
AP, JRM- Article review.
All authors approved the final draft.

Responsabilidades Éticas

Conflitos de Interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.
Fontes de Financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.
Confidencialidade dos Dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.
Consentimento: Consentimento do doente para publicação obtido.
Proveniência e Revisão por Pares: Não comissionado; revisão externa por pares.

Ethical Disclosures

Conflicts of Interest: The authors have no conflicts of interest to declare.
Financing Support: This work has not received any contribution, grant or scholarship.
Confidentiality of Data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.
Patient Consent: Consent for publication was obtained.
Provenance and Peer Review: Not commissioned; externally peer reviewed.

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Recebido / Received: 2024/01/16
Aceite / Accepted: 2024/04/15
Publicado online / Published online: 2024/07/05

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