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Dorsal Elastofibroma: About Two Cases

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Abstract **Case Report**

Dorsal elastofibroma is a rare benign tumor or pseudotumor of the soft tissues, typically located under the tip of the scapula, often found in elderly people. Its etiopathogeny is still not elucidated. Imaging, by computed tomography (CT) or magnetic resonance imaging (MRI) allows to carracterize the lesions. We report two observations to specify the clinical and paraclinical characteristics of this type of tumor as well as the modalities of management; this entity would gain to be known by any practitioner; to allow a well-codified therapeutic attitude.

Keywords: Elastofibroma; Chest wall; Scapula; imaging.

INTRODUCTION

The DE is a benign tumor not encapsulated of fibroblast origin, on the plan it is characterized by the association of dystrophic elastic fibers, dense clusters of collagen fibers, adipose tissue and spindle cells, tumor lesions, and this lesion hamartomatous [1]. It occurs mainly in people over 55 years of age with a female predominance [2-4]. It electively affects the dorsal thoracic wall at the caudal angle of the scapula, bordered by the dorsal major muscle and the serratus anterior muscle. It has an origin triggered by tissue friction found in the inferior angle of the scapula and the chest wall (99%) [3]. It is bilateral in 10% to 66% of cases. It presents with mild symptoms to pain, discomfort and functional impotence in the affected area. It is a benign tumor that requires surgical treatment due to its symptomatology and differentiation from malignant tumors [4]. We communicate two rare cases of presentation in daily clinical practice of EFD to highlight its status and our experience of management.

Observation 1

F.A., 48 years old, without any particular pathological history, was admitted for two right and left prescapular masses, of progressive appearance, painless, evolving for six years. The clinical examination revealed firm masses, mobile in relation to the two planes. Ultrasound examination of the soft tissues showed infrascapular masses with a fibrillary appearance and hypoechoic stratified echostructures (Fig 1). The thoracic CT scan shows a hypertrophy of the large dorsal muscles with invasion of the surrounding structures. These masses are opposite the inferior angles of the scapulae and contain some linear hypodense images without underlying lesions. The patient underwent elective surgery with mass resection of the scapular tumors. The anatomopathologic examination was in favor of a dorsal elastofibroma. The postoperative course was simple, with no recurrence over a period of five years.

Observation 2

Mrs. M. N., 56 years old, with no known pathological TCA. Admitted for two swellings of the right and left scapular region, evolving for two years, progressively increasing in volume and becoming more and more annoying when mobilizing the shoulder. The clinical examination revealed two well-limited swellings of 7 cm in length on the right and 5 cm on the left, opposite the tip of the left and right scapula and extending into the subscapular area. These masses were best seen in abduction and antepulsion of the upper limbs, firm in consistency, painful to palpation, mobile in relation to the superficial and deep plane of the tip of the scapula. No palpable axillary adenopathies. Soft tissue ultrasonography showed bilateral, fibrillary, stratified echo-structured infra and pre-scapular masses. The thoracic CT scan showed unencapsulated masses of lenticular shape, well delimited, of fibrous type tissue (Fig 2). Allowing to retain the diagnosis of elastofibroma. The treatment was surgical, consisting of resection of both masses. The anatomopathological examination showed that it was indeed a dorsal elastofibroma. The postoperative course was simple, with no sign of recurrence after two years.

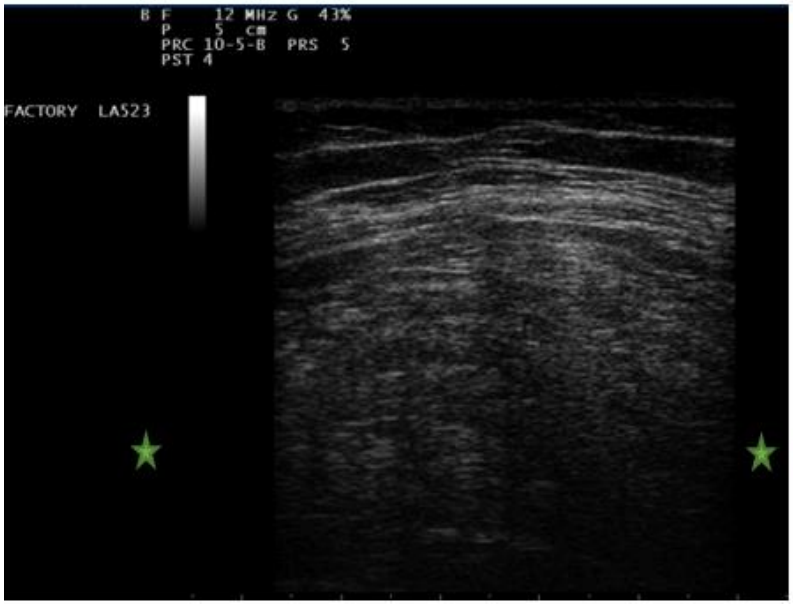

Fig 1: Soft tissue ultrasound showing alternating hiper and hipoechogenic striae

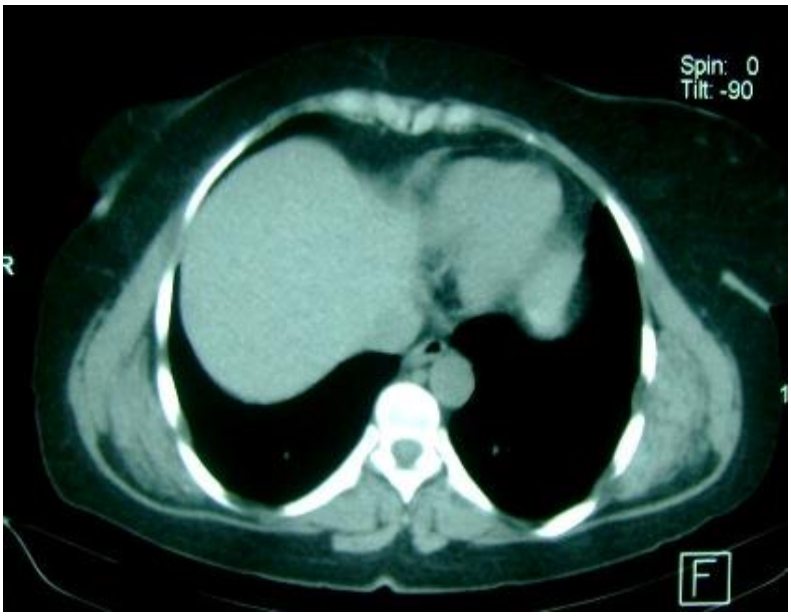

Fig 2: Chest CT scan showing bilateral masses (stars)

DISCUSSION

Bilateral dorsal elastofibroma is a benign, rare, slowly progressive tumor. It occurs in 2% of people over 60 years of age. Dorsal elastofibroma is a benign soft tissue pseudotumor lesion that is usually found in the subscapular region, as in our case, although they can be identified in other locations such as the deltoids, inguinal region, axilla, olecranon, with the spine being less frequent. One third of the presentation is bilateral. It is diagnosed more frequently in advanced ages, predilection in subjects over 55 years, although cases have been described in adolescence. It appears more frequently in the female sex, although in our case. Thus, the series of Nagamine et al, in an excellent study on closed communities in the Okinawa archipelago, an important family aggregation of the disease of 32%. His sample was 170 patients found 158 women for 12 men, which proves the female predominance of this entity that corresponds to our report [4].

The localization of this process and the age of the affected subjects strongly suggest that it is not a genuine tumor process but rather a hyperplastic reaction induced by microtrauma, following repeated friction between the scapula and the chest wall, this hyperplastic reaction being accompanied by degeneration of collagen fibers and excessive production of immature elastic tissue, derived from fibroblasts [5]. It should be noted, however, that this probably pseudotumoral reaction also appears to be constitutional, since it occurs abnormally frequently in certain families or populations. It should be noted that there was no family history in our case [6].

The asymptomatic presentation is present in half of the cases, as in our first case; sensation of discomfort in the order of 25% or stiffness during mobilization of the shoulder are the symptoms often described by the patients, clinical situation found in our second case. Painful scapular or subscapular symptoms were observed in only 10% of cases. Neurological involvement of the upper limb may be observed exceptionally, suggesting cervico-brachial neuralgia. Bilateral localization of the dorsal elastofibroma is uncommon with asynchronous development of the two masses. The possibility of two different locations in the same patient is about 3% [7]. Clinical examination reveals a firm mass, mobile with respect to the superficial planes and without signs of skin infiltration. It is often painless, palpable, and clearly visible in subscapular locations, especially when the upper limb is in antepulsion with abduction. The biological work-up is often normal; the frontal X-ray of the thorax may show an elevation of the scapula or an enlargement of the scapulothoracic space. An interscapulothoracic opacity may be demonstrated, but without associated bone lysis or calcification [8]. Thoracic computed tomography (CT) demonstrates a mass of the same density as the surrounding soft tissue, with areas of lesser density [9]. The tumor is often poorly limited and inhomogeneous. MRI shows a well-defined, often heterogeneous mass with two different T1-weighted signals, one of intermediate intensity equivalent to that of the skeletal muscle, the second of high intensity corresponding to the fat trapped within the mass. In T2, an increase in signal intensity is observed. Gadolinium injection does not enhance the signal [9]. Imaging aims to determine the measurements of the mass and its location in relation to adjacent muscle structures. The application PET / CT would be a behavior to implement in the current treatment [9]. Other authors recommend a tumor biopsy to confirm or establish another diagnosis. The histological diagnosis is based on the presence of elastotic-like fibers often fragmented within a collagenous matrix. In electron microscopy, this eosinophilic material frequently contains mature elastic fiber tissue at its center and appears to be secreted by activated fibroblasts. These confirm the thesis that the elastotic material originates from fibroblasts rather than from the degeneration of collagenous fibers. Dense cytoplasmic granulations of fibroblasts seem to correspond to elastin or elastin precursors [9]. Regarding differential diagnoses, we should consider the following entities such as lipoma, liposarcoma, hemangioma, hematoma, fibromatosis, desmoid tumor and malignant histiocytoma [9]. Complete surgical excision is the ideal treatment in symptomatic forms.

CONCLUSION

Elastofibroma is a rare tumor whose symptomatology can range from a mild gene to pain and functional impotence of the upper limb. The bilateral presentation is uncommon. MRI is the key examination for diagnosis. The differential diagnosis must be considered with malignant tumors of the chest wall. Complete surgical removal is the consensus treatment.

Conflicts of interest: The authors declare no conflicts of interest.

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