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Bilateral elastofibromadorsi : From the clinic to the treatment (about a case with literature review)

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Abstract: Elastofibromadorsi is a rare, slow-growing, ill-defined soft tissue tumor of the chest wall, most commonly located beneath the rhomboid major and latissimus dorsi muscles. This lesion is usually seen in patients over the age of 50 years. The symptoms depend on the size and location of the lesion. Clinical presentation is usually swelling, discomfort, snapping of the scapula, and occasionally pain. Its etiology has not been defined yet.The anatomic location, clinical symptoms, and typical radiologic features of elastofibromadorsi may not easily distinguish this rare benign tumor from malignant soft tissue tumors. A biopsy should be performed to rule out sarcoma in all cases, but the definitive treatment only requires simple surgical excision. Surgery should be recommended only for symptomatic patients having complaints. We report a case of bilateral elastofibromadorsi witch is un common specially when it occurred in very old age, the clinical presentation, radiology findings,surgical indications and complicationsare reviewed and compared with literature.

Keywords: Elastofibromadorsi; Soft tissue tumor; Surgery; complications

INTRODUCTION:

Elastofibromadorsi is an uncommon benign, soft tissue tumor found in the sub scapular and infra scapular region between the thoracic wall, serratus anterior, and latissimus dorsi muscle. It was first described in 1961 by Jarvi and Saxen. The autopsy incidence rate has been reported to range from 13% to 17%. It generally occurs unilaterally in women over the age of 50, bilateral involvement may also be observed by 10%. Clinical presentation is usually swelling, discomfort, snapping of the scapula, and occasionally pain. Its etiology has not been defined yet. In some studies, it has been attached to mechanical friction between the chest wall and the scapula, as a result of repetitive traumas. The differential diagnosis should be done with subcutaneous tumors such as lipomas, fibrolipomas or more aggressive tumors. In the orthopedic literature, there are few case reported. We present our experience with this lesion less frequent in African countriesthrough a case report with severe thoracic pain.

CLINICAL CASE:

A 73-year-old patient with a history of osteoarthritis in their hands presented with bilateral pain in the scapular region of 2 years duration, which was exacerbated by physical activity, referring the pain to the scapular area of the shoulder after movement. Physical examination found a hard consistency, elastic mass approximately 12 cm in diameter in the right subscapular region and 8 cm in the left side. Ultrasonography of the affected regions reported a solid mass of the chest wall, which was below the serratus anterior. CT scan showed a unen capsulated occupying mass with density similar to muscle, mixed with adipose tissue bands in both intra scapular region of 10 by 6 cm. Hematological and biochemical analyzes were normal. Our orthopedics department suggested excision of the lesions starting with the most painful side.

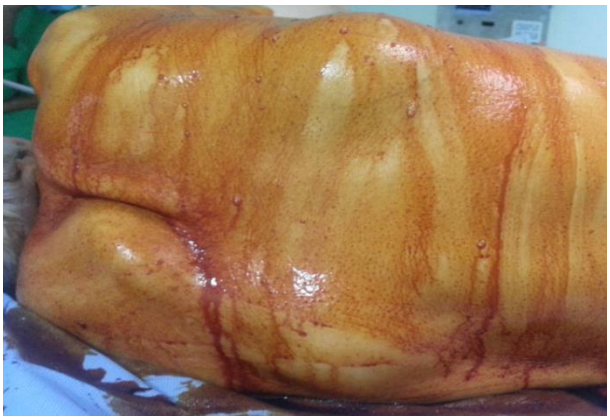

Fig-1:Bilateral elastofibromadorsi: pre operative aspects.

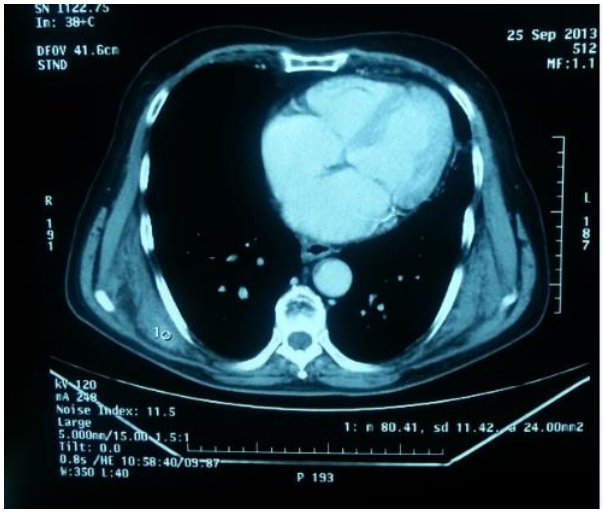

Fig-2: MR Image of Elastofibroma Dorsi.

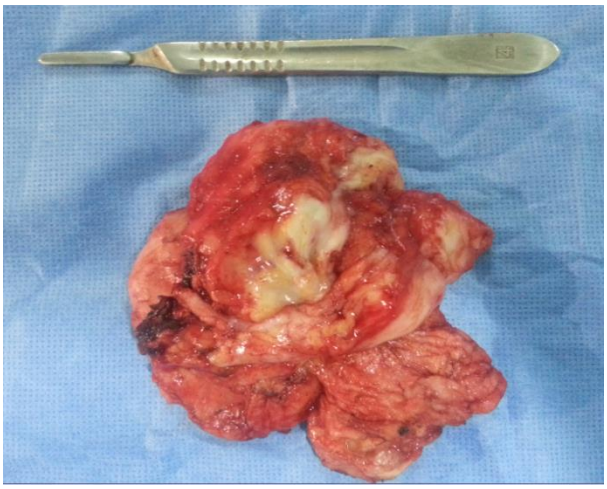

Fig-3: The excised mass.

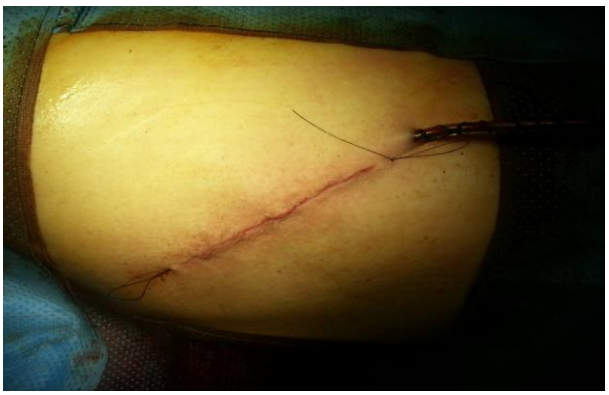

Fig- 4: Final scar: suction drainage in place.

DISCUSSION:

Elastofibromadorsi is a rare, benign, slow-growing soft tissue tumour with typical localization in the sub scapular and intra scapular region, between the thoracic wall, serratus anterior, and latissimus dorsi muscle, often connected to the thoracic wall periosteum [1]. Nag amine and Nohara have described in their study of 170 cases, unusual locations of elastofibromadorsisuch as orbit, maxilla, mediastinum, and greater omentum [2]. They reported a rage of 32% of genetic predisposition. To the same effect,in a 2002 study, Nishio et al [3] detected Deoxyribonucleic acid (DNA) copy number changes involving 1 or 2 chromosomes in 33% of 27 their patients. The most common recurrent gains were at bands Xq12-q22 and 19.Some authors [4, 5] state, in addition of the mechanical theory, that circulatory insufficieny caused by degeneration in the fibrous tissue the main cause of elastofibroma [6, 7]. All the larger series of elastofibroma reported in the literature, showed that this tumor occurs mainly in women over 55 years, with a mean age of 60 years at diagnosis [1, 8]. In our case, it was a man aged over 70 years.

The symptoms depend on the size and location of the lesion; in its dorsal form, it may present with shoulder pain, snapping feeling in shoulder motions, and patients start sweeling on their backs while lying. The physical examination may reveal a palpable, firm mass at the lower corner of the scapula, more prominent on abduction of the arm, usually immobile and probably due to its adherence to the surrounding tissue [1;5;9]. Another mass may be present on the opposite shoulder up to 50% of the cases [6]. Although the majority of cases are asymptomatic and the diagnosis is incidental during thoracotomy or post-mortem studies [2,10]. Giebel et al [11], in a series of 100 autopsies, found 13 patients with elastofibroma dorsi. In our patient, the clinical presentation was typical and bilateral.

In imaging, chest radiographs may show an elevation of the scapula from the chest wall. In sonographic, elastofibroma has a typical appearance consisting of arrays of linear strands against an echogenic background. However, in some cases, the sonographic pattern of elastofibromadorsi may be very similar to that of the surrounding muscular tissue, and neither a clear cleavage surface nor a specific vascular pattern can be seen [12]. In these cases, both CT scan and IMR, may show as described Kransdorf [13] a non encapsulated heterogeneous soft tissue mass with poorly defined margin and a lenticular form with its long axis in craniocaudal orientation. Elastofibroma appears as heterogeneous soft tissue attenuation mostly similar to the skeletal muscles, with linear interlaced low density streaks suggesting mature fat. Elastofibromas are very frequently bilateral, a finding that has not been strongly emphasized in the literature. The presence of similar contra lateral per scapular lesion is of great help in making the radiologic diagnosis of elastofibroma, because this finding virtually eliminates malignancy from the differential diagnosis. Therefore, if the diagnosis of elastofibroma is suspected clinically, it is important to image both sides of the chest wall [14].Biopsy is reserved for suspicious cases that do not exhibit a characteristic pattern.

Treatment of elastofibromadorsi involves complete excision of the tumor, something which is not

always possible due to the fixation of the tumor to muscles, periosteum of ribs and scapula. In the past, we recommended resection for symptomatic and asymptomatic patients with tumors suspicious for elastofibroma, in order to pathologically confirm the diagnosis [10, 15]. However, as the diagnosis of elastofibroma is now easily made based on its clinical and imaging characteristics, we only perform tumor resection in symptomatic patients or in those who desire resection of the tumor [16,17]. Elastofibroma is usually described as a slowly growing lesion, but there are scattered reports of rapid growth and one report of spontaneous regression of elastofibroma without treatment. Recurrence following incomplete surgical excision was reported once. No reports of malignant transformation exist [2, 16,17]. In the case we report, the two masses appeared simultaneously with a slow and progressive evolution over 2 years. Pain was the main symptom that prompted us to operate the patient to this advanced age.

The high incidence of postoperative hematoma which varies from 38 to 87% discourages surgical treatment of elastofibroma specially in asymptomatic cases [18,19]. In a study of Satochi Nagano [20], the author demonstrated tumor size and duration of wound drainage to be associated with the incidence of postoperative hematoma. Although rare, the occurrence of postoperative seroma is a complication to know and for which Satochi proposed combining quilting sutures and fibrin sealant, a technique adopted by many plastic surgeons in the donor site of a latissimus dorsi muscle flap. These complications leads to additional treatment and longer hospital stay, it is for this reason the majority of authors recommend a week to ten days of immobilization of the upper extremity on the affected side with longer suction drainage. Postoperatively, our patient came in consultation about the tenth day with a swelling at the site of resection on the right side, in relation to moderate plenty hematoma that required surgical evacuation.In the literature, no postoperative neurologic abnormalities or muscle weakness of the shoulder or the arm has been reported [2,13,19,20].

CONCLUSION:

Elastofibroma dorsi is very rare, the typical anatomical lactation, coupled with symptoms, strongly suggest the diagnosis, which can be con-firmed by core biopsy. Knowledge of this entity could avoid useless procedures especially in elderly and asymptomatic patients where a simple follow up could be sufficient. Surgery should be recommended only for symptomatic patients having complaints.

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