

Elastofibroma dorsi: About six cases

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ABSTRACT: Elastofibroma dorsi (ED) is a rare, benign soft tissue tumor arising from connective tissue and usually found in the subscapular region. We conducted this retrospective study to contribute to a better understanding of this tumor, the pathogenesis of which is still unclear.

Methods: We reviewed the medical records of six patients (4women, 2men) treated for ED at our institution.

Results: The mean age was 48 (range, 33-63). The tumor was located on the right in one patient, on the left in three. One patient had a bilateral localization. One patient had three localizations. Two patients presented with symptoms of pain and clunking of the scapula on shoulder abduction. All patients underwent complete marginal resections. The resected tumors ranged in size from 47 mm to 85mm. two recurrence has been observed in follow-up of four months and six months in the same patient.

Conclusions: Elastofibroma dorsi should be considered in differential diagnosis of soft tissue tumors due to their specific location. As it exhibits benign behavior, it should be surgically removed only in symptomatic patients.

KEYWORDS: Elastofibroma ; soft tissue tumors ; scapula ; ct-scan ; surgery.

1 INTRODUCTION

Elastofibroma dorsi (ED) is a benign, slow-growing fibroelastic tumor that was first described by Jarvi and Saxen in 1961. In 99% of cases, EDs are localized to the infrascapular region between the thoracic wall, serratus anterior, latissimus dorsi muscle, and are often attached to the periosteum of the thoracic wall. Elastofibroma has been occasionally reported in other locations. It is bilateral in 10% of cases. The typical presentation of ED involves a subscapular mass associated with a long history of swelling, discomfort, snapping of the scapula and, in some cases, pain. Ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) can all be used for its radiologic diagnosis. However, in the symptomatic forms, or when there is doubt as to whether the tumor is benign, a biopsy or resection should be discussed.

We present 6 cases with elastofibroma dorsi and discuss the clinical presentation, diagnosis, and treatment options.

2 PATIENTS AND METHODS

The study included six patients, 4 females and 2 males, aged 33 to 63 years, who were diagnosed as presenting an elastofibroma dorsi. The mean follow-up period was 5 years. Four patients were initially presented at other clinics and were referred to our hospital with a large, unknown tumor of the back. On physical examination, a non-tender, firm mass was palpated in the dorsal region with functional disturbance. Radiographs were performed in all patients. Five patients were explored with computed tomography (CT), three of them with sonography and one with Magnetic Resonance Imaging (MRI).The MRI was indicated to control recurrence.

All patients were operated. We conducted an oblique approach on the inferior edge of the scapula under general anaesthesia and carried out complete macroscopic resection, adding a small muscle margin in cases of peripheral adhesion. All samples were sent to the anatomical pathology department for histological analysis. Postoperative immobilisation with a sling was maintained for 2 weeks. After this period, patients started assisted and passive exercises of the shoulder and

scapula, which then became active movements according to tolerance. The relevant follow up was performed by clinical, ultrasound tests and MRI for patient who presented recurrence.

3 RESULTS

The study included 6 patients (4 women, 2 men) with a mean age 48 (range, 33-63). No patients had a family history of elastofibroma. One patient was manualworker (tab.1). Clinical examination showed a firm, deep, swelling mostly in the infrascapular region (Fig. 1), which was fixed to the rib cage. The swelling was not tender on palpation. The swelling was more prominent on forward flexion of the shoulder due to the inferior angle of the scapula moving forward. Examination of surrounding lymph nodes was normal.

The main symptom was pain. Two patients presented with clunking of the scapula upon shoulder abduction. All patients were aware of a round mass at the inferior pole of the scapula that had been present for a mean 4 months (range, 1-12 months). The tumor was located on the right in one patient (Fig. 2), on the left in three. One patient had a bilateral localization.

One patient had three localizations (left subscapular, left apex of the chest, right infrascapular). This patient was operated 4 months ago, for a left subscapular chest wall tumor with free surgical margins. Histopathology study confirmed the diagnosis of elastofibroma tumor. She presented to the surgical department with a bilateral hard mass located in the left suprascapular and right infrascapular regions. In our patients, the sizes of the tumors were 47 mm to 85mm.

All patients underwent imaging of the tumor by computed tomography (CT) scanning. One patient was explored with Magnetic Resonance Imaging (MRI) (fig. 3). Ultrasound was performed in two patients. Ultrasound of the soft tissues showed hypoechogenic oval lesions with ill-defined margins on both the superficial and deep planes. Color-Doppler not revealed intralesional vascularization and transducer compression did not detect any morphologic variation whereas the lesions were always fixed to the deep costal plane and mobile to the superficial soft tissue. Thoracic CT-scan showed masses that were slightly heterogenous with a well-circumscribed fatty component suggesting a connective tissue tumour with no parietal invasion or costal lysis. In the woman who presented with recurrence after 4 months, Ultrasonography showed the same features for both tumors which were oval (fig. 4), with ill-defined margins and inhomogeneous consisting in an alternation of hyper and hypo- echogenic bands. Doppler not revealed an intrinsic vascularization (fig. 5). Thoracic and abdominal computed tomography demonstrated 2 solid masses, with ill-defined margins, muscular-like density and heterogeneous aspect. The first mass measured 47X22mm (fig. 6) and was located profoundly within the left trapezius and levator scapulae muscles in contact with left supraspinous fossa, the first three ribs and clavicle. The second tumor measured 70X25mm and located between the subscapular muscle and right rib cage (5-8th ribs) deep to serratus anterior muscle (fig. 7-8). After use of contrast agent, enhancement was low and heterogeneous. There was neither endothoracic extension, nor regional lymphadenopathy or metastatic localizations.

One patient had a trucut biopsy. All patients were operated. Surgical exploration confirmed radiological features tumors were indistinct to bone structures and the resection razes ribs and scapula. It was a well-circumscribed tumor, firm oval shaped unencapsulated, surrounded by subscapular, latisimus dorsi, and serratus anterior muscles, and in contact with the rib cage. Marginal resection of the tumour was performed into healthy tissue. A suction drains was Kept inside surgical sites for 48 hours. The postoperative course was uneventful.

The histopathological study of the surgical tumour sample showed degenerative elastic, highly eosinophilic fibers, fragmented into a series of successive globules or scattered irregularly. These elastic fibers were held together by a fibromyxoid network, associated with entrapped areas of mature adipose tissue, suggesting an elastofibroma. There were no malignancy features. One patient had a second recurrence 6 months after complete resection of bilateral localization. The tumor of 5 x 3 cm had the same scannographic features previously described. On MRI, tumor was ill-defined with inhomogeneous muscular-like signal intensity both on T1 and T2-weighted sequences and separated from surrounding tissue by a fat layer. After gadolinium injection, tumor displayed enhancement. Despite this radiologically documented recurrence, Surgery was deferred. First clinic and MRI control was realized at 6 months then the patient was monitored clinically every 6 months. After 24-month of periodic assessment, the patient is still asymptomatic with stable tumor size.



Fig. 1. The swelling was more prominent on forward flexion of the shoulder



Fig. 2. Right infrascapular tumor

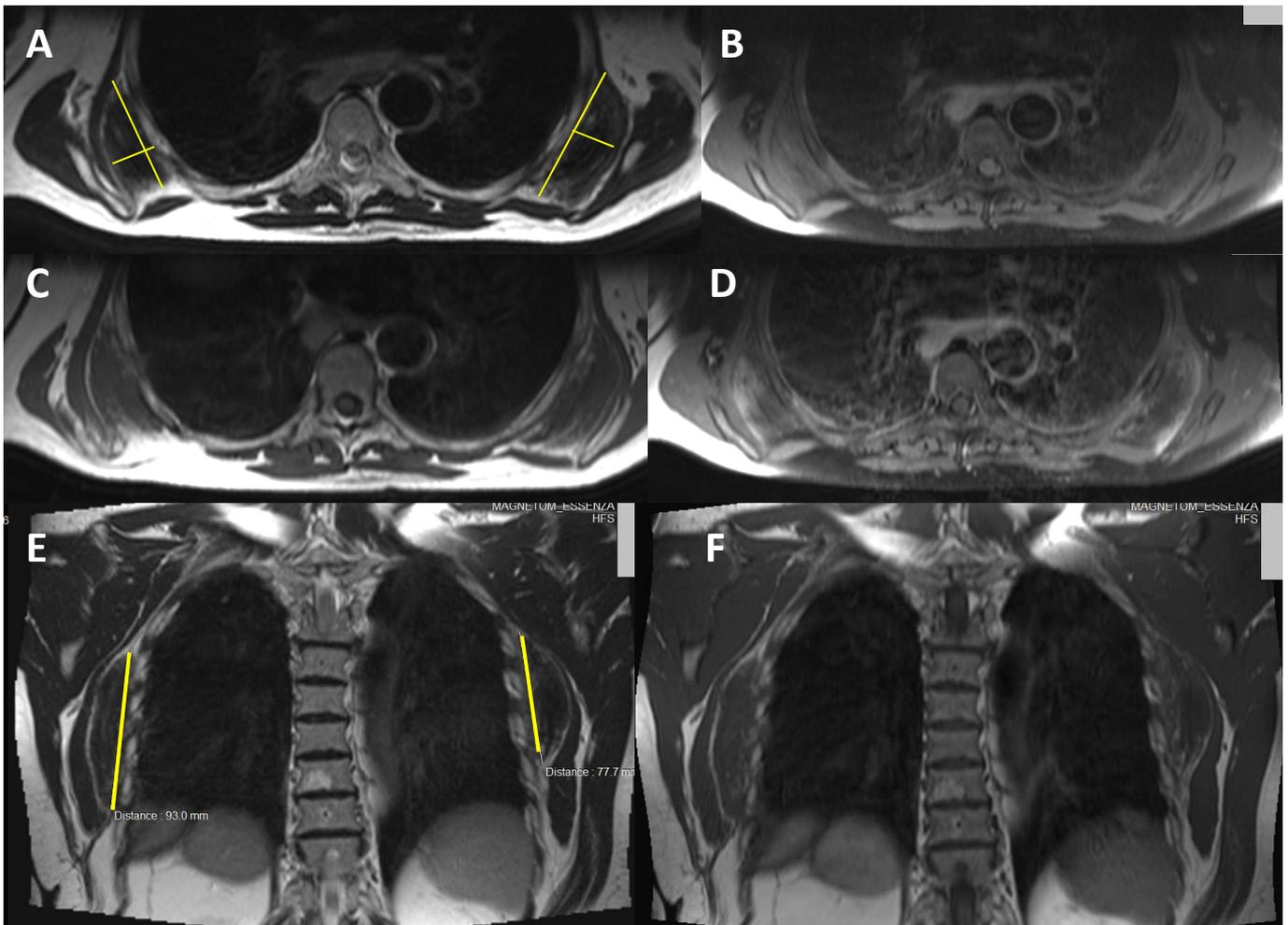


Fig. 3. MRI showing bilateral infra scapular masses having a comparable signal with the muscle (A: T2 ; B: T1 FS ; C: T1 ; D: T1 FS Gado ; E : T2 ; F : T1)

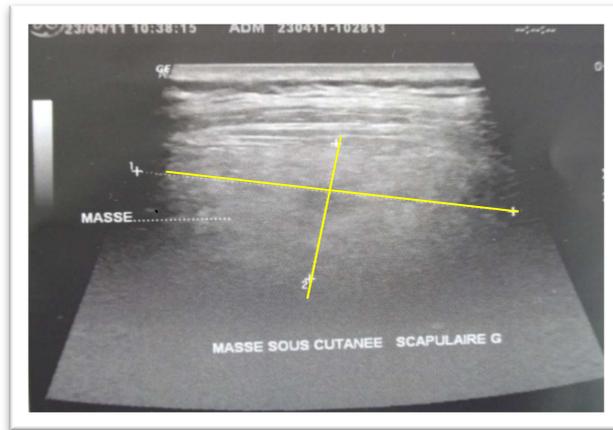


Fig. 4. Ultrasonography showed oval tumor

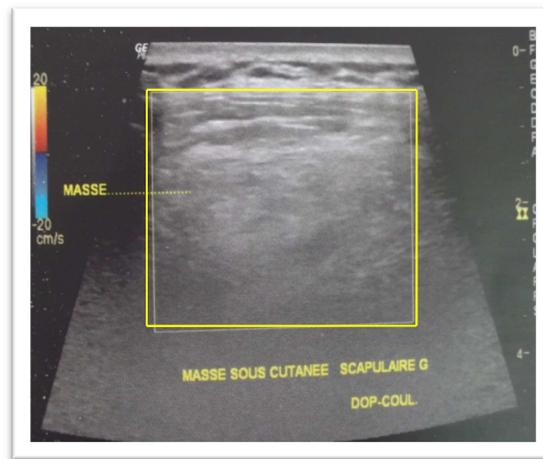


Fig. 5. Doppler not revealed an intrinsic vascularization

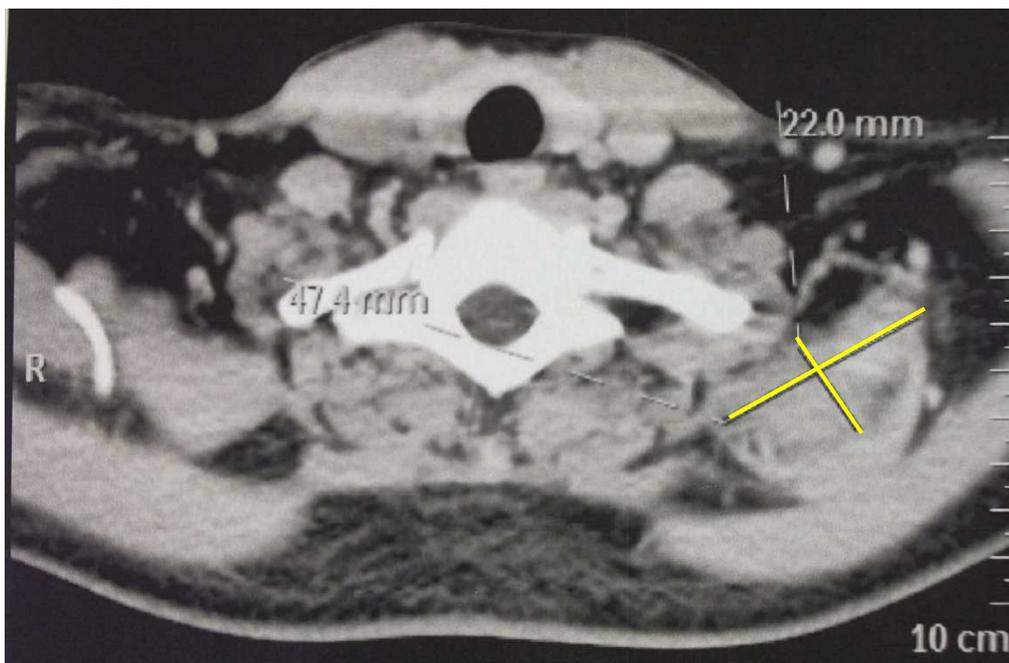


Fig. 6. Thoracic CT-scan showed heterogeneous tissular tumor

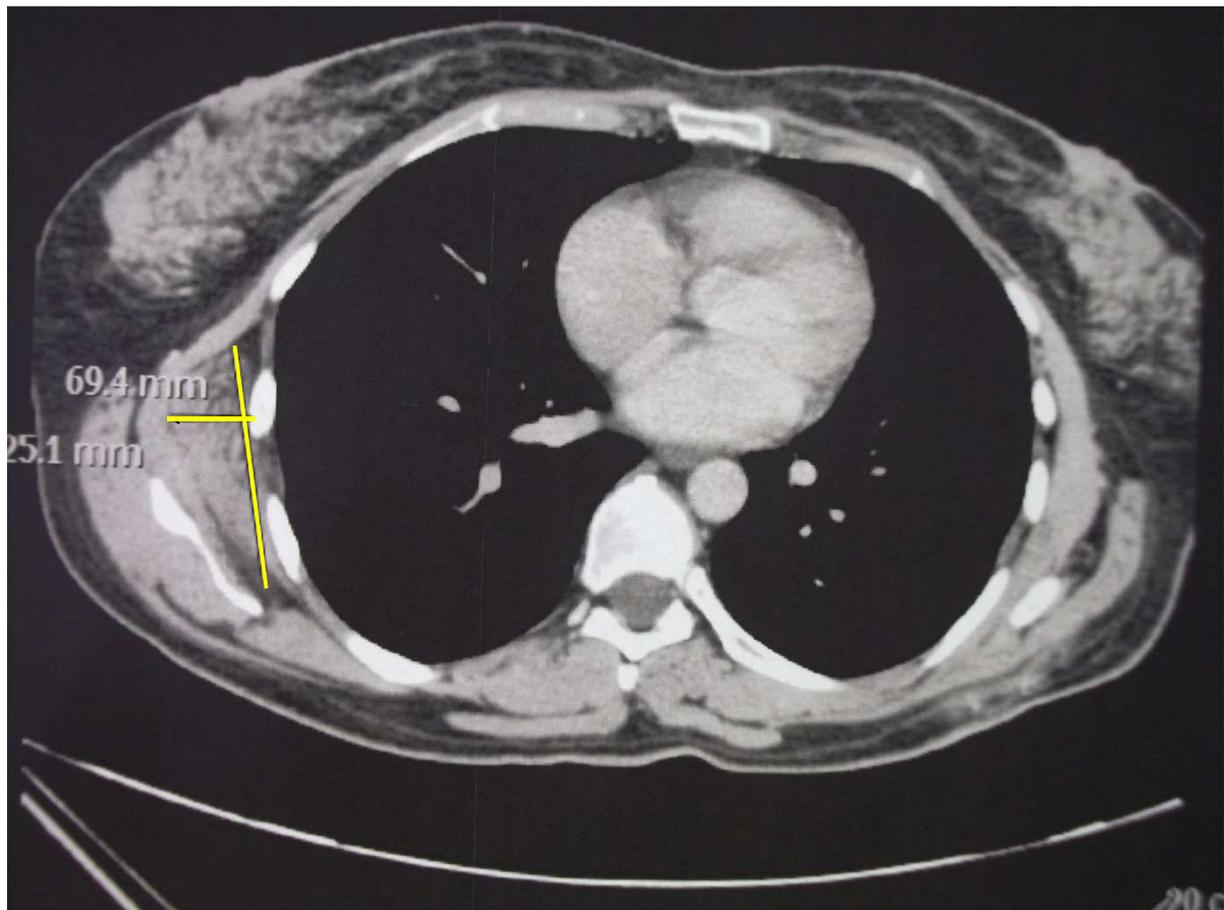


Fig. 7. Thoracic CT-scan showed the right localization

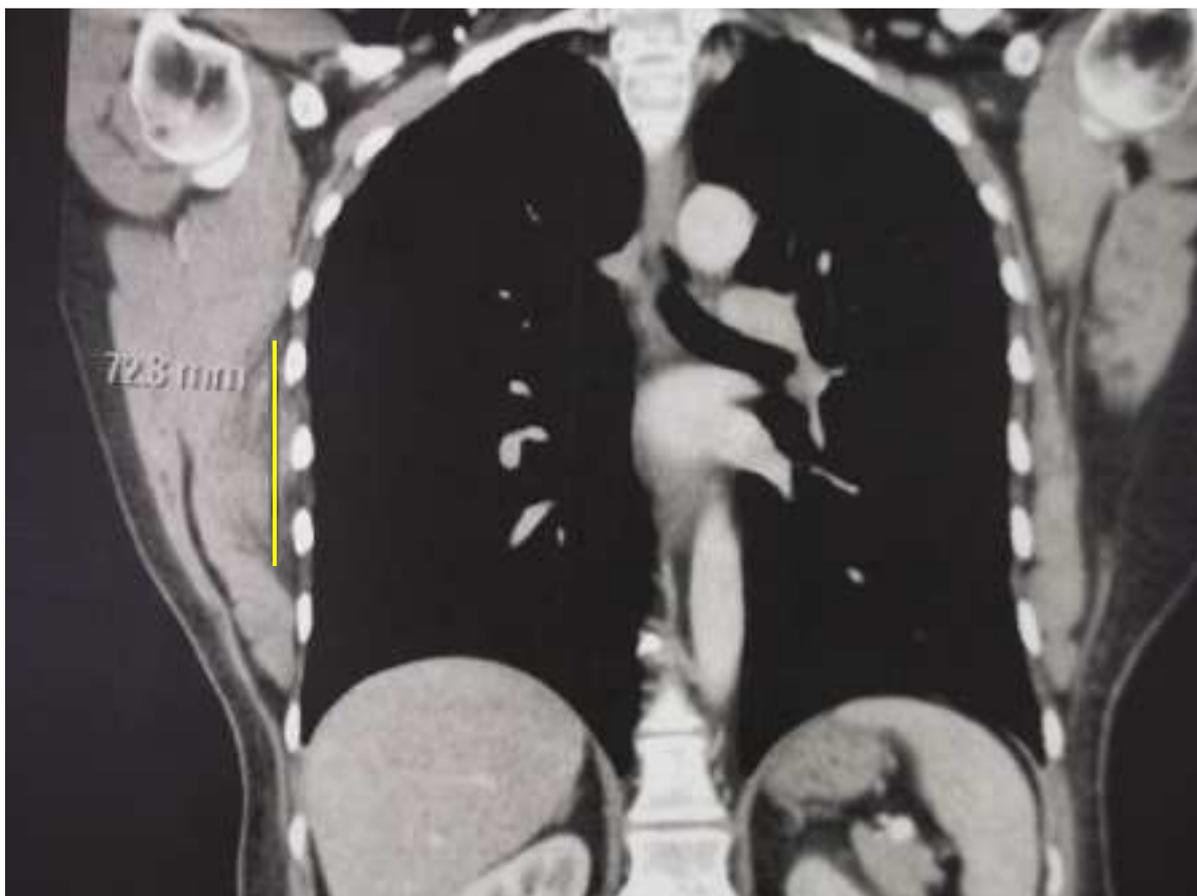


Fig. 8. Thoracic CT-scan showed infra scapular tumor

Table 1. Detailed summary of patient data

Patients	Gender	Age	Profession	Tumor extension in cm	Location	Symptom	Comorbidity	Duration of onset (months)	Basis of diagnosis	Treatment	Recurrence
1	M	48	farm worker	70mm	1-left subscapular tumor	pain and clunking of the scapula	No	1 month	CT scan	complete marginal resection	no
2	F	33	housewife	85mm	1-left subscapular tumor	pain+swelling	no	4 months	Ultrasound +CT scan	complete marginal resection	no
3	M	42		70X20mm	1-left subscapular tumor	pain+swelling	No	12months	CT scan	complete marginal resection	no
4	F	44	housewife	50mm 47X22mm 70X25mm	1-left subscapular, 2-left apex of the chest, 3-right infrascapular	pain and clunking of the scapula	diabetes	1month	Ultrasound+ CT scan+ trucut biopsy	complete marginal resection	yes
5	F	63	housewife	60mm	right infrascapular	pain+swelling	left breast carcinoma	4 months	CT scan	complete marginal resection	no
6	F	60	housewife	64x23mm 70x30mm	1- right infrascapular 2-left infrascapular	pain+swelling	hypertension	6 months	Ultrasound+MRI	complete marginal resection	no

4 DISCUSSION

Elastofibroma dorsi is an uncommon soft tissue tumor that was first described by Jarvi and Saxen [1] in 1961. It is a benign hyperplastic soft-tissue pseudotumor consisting of fibroelastic tissue and fat. The site is characteristic and is most commonly (93%) the deep dorsal region between the thoracic wall and the lower third of the scapula beneath the serratus anterior and the latissimus dorsi muscles. Other locations are much rarer (the hand, the foot, greater trochanter, the deltoid muscle, the ischiatic tuberosity, the cervical region, the stomach, the omentum and the rectum [2]). It may be bilateral in 10–66% of cases, such as our patient. Its onset occurs at an average age of 60 years (range 41–80 years) [3]. The mean age in our series was 48 (range, 33-63). Less than 330 cases were reported in the literature before 2005. In the autopsy series of Giebel and colleagues [4], elastofibroma was encountered in 13% of patients, whereas preelastofibroma-like changes were observed in 81% of autopsies.

The ED is more likely to be found on the same side as the dominant arm, although we have not encountered any publication supporting this view. On the contrary, Daigeler et al. reported in their seven-case series that there was no correlation between the dominant hand and the side of the ED [5, 6]. When etiopathogenesis is linked to the use of arms only, it would be expected that the ED incidence might be higher in men, who often carry out more manual jobs than women.

Elastofibroma dorsi generally manifests as a circular/oval, poorly demarcated, and slow-growing soft tissue mass in the subscapular area of the chest wall. Although it is frequently seen as an insensitive, asymptomatic swelling. The swelling may be accompanied by pain, discomfort, or tension induced by shoulder movement in the periscapular area, limited shoulder mobility, and a snapping scapula [7]. Mortman et al. reported in their six-case series that four cases were asymptomatic with ED diagnosed after thoracotomy for some other reason [8]. In our series, the primary complaints were swelling and pain while lying on the back. Two patients presented with symptoms of pain and clunking of the scapula on shoulder abduction and limited mobility when using the arms.

Radiologic examination is essential in the evaluation of ED. Ultrasound is the initial examination of choice. Solivetti et al have recommended the use of ultrasound imaging for conclusive diagnosis of elastofibroma. Ultrasound imaging allows a reduction in the costs of pathology and avoids unnecessary investigation [9, 10]. It often shows the fibrous, fasciculated appearance of the tumour with hyperechogenic strands parallel to its main axis [2]. The CT appearance of ED is a poorly defined soft tissue mass, which is isodense in relation to the adjacent muscle, with scattered areas of fat attenuation and internal striations. Fat attenuation is a finding that correlates with the histopathologic finding of plentiful mature adipocytes scattered throughout the ED [11].

On MRI, ED is seen as patterns of alternating fibrous and fatty tissue. The fibrous tissue is isointense relative to skeletal muscle in both T1- and T2-weighted images, whereas the fatty tissue is hyperintense in T1-weighted sequences. Contrast-enhanced images reveal areas with, and areas without enhancement [12, 13]. On PET scan, elastofibroma dorsi results in low grade diffuse F-18 FDG uptake, suggesting a benign entity with a low level of metabolic activity [2].

The differential diagnosis list for ED is limited, but it includes lesions similar in CT and MRI appearance to skeletal muscle, with decreased cellularity and abundant collagen, such as extra-abdominal desmoid, neurofibroma, cicatricial fibroma, and malignant fibrous histiocytoma. Atypical imaging features, including adjacent bone destruction or intense contrast enhancement, should raise suspicion of different tumors, such as liposarcoma or metastatic disease. In patients with no symptoms, simple excision of the tumor is recommended only if the maximum diameter exceeds 5 cm. Simple observation of lesions larger than 5 cm in patients with no symptoms is warranted if the histologic diagnosis from the biopsy specimen shows a benign character [2, 10, 14, 15]. .

Marginal total resection is the best treatment for symptomatic ED. Therapy seems to vary depending on whether or not there are symptoms. If the lesion is asymptomatic, simple observation suffices. If the symptoms are severe enough, marginal resection is sufficient [2, 15]. As for the necessity of biopsy, opinions vary widely. The authors of the oldest articles systematically recommend biopsy to establish the differential diagnosis with a sarcomatous lesion [16]. On the other hand, the authors of the most recent articles most often consider that imaging studies, particularly MRI, suffice if the lesion is typical [17]. As malignant transformation has never been reported [15]. Kransdorf reported a recurrence rate of 7% which was attributed to incomplete tumour resection [2]. We have found recurrence in one patient, 4 months after first resection and appearance of a second recurrence, with different localisation, 6 months after second surgery. In this case Surgery was deferred.

5 CONCLUSION

Elastofibroma dorsi, although rare, must be recognized to avoid diagnostic–therapeutic errors. The finding of a solid, slow-growing lesion, firmly adherent to the deep subscapular region in a patient between the 5th and 6th decade of life, with a typical fasciculated pattern is pathognomonic of elastofibroma dorsi, its bilateral location or its homolateral bifocality convalidate diagnosis. Ultrasound is the first examination and is sufficient to orientate diagnosis. CT and/or MR are reserved only in case of non-fasciculated ultrasound patterns, when the lesion has an atypical isolated site or to stage the precise location in candidates for surgery. Biopsy is reserved only in cases where integrated imaging is aspecific and unable to differentiate the disease from other malignant lesions. Unnecessary wide and radical resections in the symptomatic patient can be avoided because marginal resection has proven to be sufficient. We recommend postoperative wound drainage and compression garment, as well as shoulder immobilization for one week to reduce postoperative seroma.

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