CLINICAL REPORT

Bilateral elastofibroma dorsi. A case report and review of the literature

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Summary Elastofibroma is a rare benign soft tissue lesion, typically located deep under the lower pole of the scapula. It is characterized by a fibrous and adipose tissue proliferation and most frequently affects older females. Its characteristic location and its specific aspect in imaging studies most often provides the diagnosis following an incidental discovery. Nevertheless, anatomic and pathologic confirmation is necessary to formally rule out a malignant tumor diagnosis. We report a 66-year-old woman original observation; this lady’s occupation involved a number of strenuous manual activities; she consulted for chronic pain related to a left subscapular mass. MRI demonstrated, in fact, two symmetrical tumor masses under each scapula. The only symptomatic lesion was surgically excised.

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Introduction

Elastofibroma dorsi (EFD) is a rare, benign, nonencapsulated lesion characterized by the proliferation of elastic fibers within a stroma of collagenous adipose tissue. Since it was first described by Jarvi and Saxen in 1961 [1], only 320 cases have been reported in the literature [2,9,10]. In the vast majority of cases, it is located at the posterior chest wall, usually in the periscapular soft tissues at the inferior tip of the scapula, bordered by the subscapular, rhomboid, latissimus dorsi, and serratus anterior muscles (99% of cases) [2,9]. Elastofibroma is often asymptomatic and discovered inadvertently. The imaging data, particularly MRI, seem sufficient for diagnosis and suspending further investigations [7,8]. However, in the symptomatic forms, or when there is doubt as to whether the tumor is benign, a biopsy or resection should be discussed [2,15].

Observation

A 66-year-old female, with no particular history, consulted for pain when using her left shoulder, which had been progressing for 5 months, associated with a dull sound from the scapula during use. She had exercised a number of repetitive manual activities for 45 years, notably traditional bread kneading and baking (Tabouna). The physical examination demonstrated a well-circumscribed mass that was mobile, sensitive, and located under the inferomedial angle of the

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left scapula. The tumor was better visualized in abduction or in antepulsion. No adenopathy was found. The neurological examination of the left upper limb was normal. The biological and thoracic radiological tests showed no abnormalities. Ultrasound showed a solid mass that was relatively well circumscribed, measuring 9 cm in greatest dimension. It was located against the posterior side of the rib cage partially covered by the scapula and the muscle layers, was isointense relative to skeletal muscle, but showed a few streaks of increased intensity within. Because of suspected malignity, an MRI was performed immediately, demonstrating a mass located behind the inferior pole of the scapula, seated deep to the chest wall, raising the serratus anterior muscle with which it formed a cleavage. It was oblong in shape, fairly well circumscribed, nonencapsulated, showing a heterogeneous signal with a fibrillar structure with a fibrous component showing low signal intensity on T1- and T2-weighted sequences (Fig. 1), and an adipose component showing high signal intensity on T1- and T2-weighted sequences, with slight enhancement after gadolinium injection. The same lesion was viewed on the opposite side, measuring 4 cm in greatest dimension. The diagnosis of elastofibroma was made based on these imaging and clinical findings. Given the persistence of the progressively worsening functional discomfort, the lesion was explored surgically under general anesthesia via a lateral 5-cm intermuscular incision. The tumor was excised totally. Macroscopically, the tumor was firm, ovoid, well-circumscribed, nonencapsulated, and measured 10 × 8 × 3 cm (Fig. 2). Histological analysis showed a mesenchymatous tumor made up of thick bands of collagen and many elastic fibers with an abnormal aspect arranged in wide rows or in irregular-shaped areas separated by mature adipose islets (Fig. 3). The elastic fibers, clearly demonstrated with orcein staining, were highly dystrophic: some were long and had a serrated border, others were branched and presented as small beads with scalloped contours that were sometimes organized linearly (Fig. 4). There was no capsule at the lesion's periphery. This dense, fibrous, connective tissue that was rich in dystrophic elastic fibers admixed with adipose tissue gave a histological

**Figure 1** Transversal T2-weighted MR image showing an elastofibroma as a soft tissue mass deep in the muscles and adjacent to the chest wall.

**Figure 2** Macroscopic appearance of elastofibroma showing grayish-white fibrous areas admixed with adipose tissue.

**Figure 3** H&E stain showing branched and unbranched coarse elastin fibres admixed with collagen and mature adipose tissue.

**Figure 4** Orcein stain showing deeply staining branched and unbranched elastin fibers.
diagnosis of elastofibroma. The patient was doing well after 6 months.

Discussion

Elastofibroma is a rare lesion that should be known and suggested in a patient presenting functional discomfort with scapula movement. Several pathogenic theories have been proposed. Repeated microinjuries between the chest wall and the scapula, the source of excess elastin production and collagen degeneration, could play a physiopathologic role in this rare lesion [9,10,13]. However, in the literature review, the notion of forced manual exercise is not found consistently, since only rarely do authors report patients’ occupations and the predisposing factors. A familial predisposition was reported by one author [2]. Recent research has suggested that EFD may not be a simple reactive fibroblastic pseudotumor, but rather a monoclonal neoplastic process with genomic instability. The main cytogenetic anomaly of EFD consists of substantial karyotypic instability responsible for structural modifications that can affect nearly all chromosomes [9,14]. After a study of 14 cases of EFD, Hisoaka and Hashimoto suggested that the mesenchymatous CD34+ cells are an integral part of the EFD process, meaning that this is a fibrous monoclonal proliferation [17]. Nevertheless, a controversy persists as to whether these tumors are neoplastic or reactive. Whatever the origin of the EFD, it is a tumor, or more exactly, a reactive process that develops very slowly. Nagamine et al. noted pretherapeutic delays of 1 day to 67 years, which they did not find to be exceptional [2]. Initially, EFD was considered a rare pathological entity, but autopsy series have shown the presence of subclinical EFD (< 3 cm) in 24% of women and 11% of men older than 50 [1,13].

In more than 80% of cases, EFD is located in the subcapsular region between the rhomboid, latissimus dorsi muscles, and thoracic wall from the sixth to the eighth ribs in 99% of cases. However, rarer locations have been described: the ischial tuberosity, foot, deltoid, axilla, great trochanter, olecranon, tricuspid valve, stomach, eye, hand, and finally the inguinal region and the greater omentum [2,3,4,11]. Subcapsular EFD is found more frequently on the right side (60%), but in 66% of cases, it is bilateral [1,2,5]. In this context, both tumors develop asynchronously. The second tumor is most often discovered on clinical or radiological examination, as in our observation. Two different locations can be discovered in the same patient: dorsal and olecranon [2] and dorsal and gastric [4].

From a clinical perspective, in more than half of the cases, EFD is asymptomatic, and in more than 90% of the cases, it is discovered inadvertently by patients who note a mass protruding into the scapular region [10,14]. An impression of discomfort or stiffness when using the shoulder, sometimes with clicking as in our observation, is found in 25% of cases [9]. Periscapular pain symptoms are only observed in 10% [2]. Exceptionally, neurological involvement of the upper limb is observed, suggesting cervicobrachial neuralgia [12]. Clinical examination often finds a firm mass, usually painless, under the inferior angle of the scapula, often bilateral, adhering to the deep layers, with no local signs of inflammation, masked by the scapula during retropulsion of the shoulder, which becomes prominent when the shoulder is displaced toward the front.

EFD is found most often in the subject aged 55 years or more; however, some cases have been described in younger patients [11,2]. A female predominance is most often noted, with a male:female sex ratio of 1:13 [5,10]. However, recent studies have reported an equal distribution between men and women [14], even a male predominance (12:3 male:female ratio) [10]. The biological constants always remain normal. In most cases, the thoracic x-ray is normal, but it can show a raised scapula or an opacity between the scapula and the chest wall [9,10]. Tumefaction in the soft tissues can suggest a number of diagnoses: lipoma, fibroma, schwannoma, desmoid tumor, hemangioma, even primary or secondary sarcoma [16].

Ultrasound is often the first-line examination. Although not specific, several characteristics allow a more precise diagnostic approach: a sub- and precapsular location of the mass, its fibrillar and fascicled aspect in relation to the hyperechoic streaks parallel to the lesion’s long axis, notably in its superficial portion, and finally bilateralism [7,16]. This bilateralism is an additional argument and therefore, the contralateral side should be systematically explored. The diagnosis suggested by ultrasound should be confirmed by CT or MRI. The typical aspect on CT is a nonencapsulated mass that is lenticular in shape, craniocaudal in its greatest dimension, isodense compared to the neighboring muscular structures, with hypodense streaks and the density of fatty tissue [7,16]. In addition, this exam clearly shows the characteristic feature of no bone abnormality in this tissue mass. Some articles have mentioned the inadvertent discovery of EFD during a PET scan. EFD leads to diffuse uptake and low intensity of 18-FDG, demonstrating a benign entity with a low level of metabolic activity [7,9,19]. MRI, the noninvasive technique that is the best adapted to diagnosing EFD, can demonstrate the dual fibrous and fatty tissue quality while showing the very characteristic location.

Elastofibroma is therefore a mass with clear or indistinct margins [7,20,21]. Its contents are heterogeneous and include a part of fibrous tissue that presents a low signal on T1- and T2-weighted images, very similar to that of the muscles. The low signal on T2-weighted images results from the low cellularity and the abundant formation of collagen tissue. Moreover, the foci of fatty tissue, with an intense signal in T1-weighted images and an intermediate signal in T2-weighted sequences, are demonstrated [7,10,20]. In the most typical cases, the fatty layers are distributed in strata alternating with fibrous layers of linear or curvilinear structures, more or less parallel to the chest wall, resulting in a superimposed layered aspect [7,16,20,21]. The discovery of a mass that produces little pain is bilateral and symmetrical, with MRI aspects on T1- and T2-weighted images as described above, provides the diagnosis of elastofibroma, without the need for gadolinium injection. Beyond these classical cases, atypical forms exist (unilateral or a highly asymmetrical lesion, painful, rapidly progressing in volume, with a hard consistency, and homogenous on imaging studies), which require gadolinium injection on MRI, so that a heterogeneous mass...
with very slight enhancement can be found. This last point provides the differential diagnosis with malignant lesions [20, 21]. The differential diagnosis of periscapular lesions with a low or intermediate signal on T1- or T2-weighted sequences includes tumoral lesions with low cellularity and abundant collagen such as desmoid tumors, neurofibroma, liposarcoma, aggressive fibromatosis, and malignant histiocytodesmoma [6,15]. However, enhancement of these tumors after gadolinium injection is usually substantial or heterogeneous in relation with tumor neovascularization that is often significant in this type of tumor compared to elastofibroma [7,9,16]. Moreover, the frequent onset of EFD in the older subject, particularly in women, its subscapular topography, its bilateralism and its aspect on slice imaging (CT and/or MRI) are sufficient for a positive diagnosis, thus avoiding systematic biopsies and unnecessary surgical resection [14, 15]. Therapeutic abstention is the rule in EFD discovered inadvertently. Occasionally, after imaging exams have been done, doubt persists, making it necessary to obtain certainty of benignity of the lesion on histological exam. Puncture biopsy under local anesthesia can be done in this case. If benignity is confirmed, no treatment is proposed [16, 20, 21]. The advantages of cytology have not been clearly established and little has been published on the subject in the literature [13,16]. Surgical treatment is reserved for highly invalidating forms or for esthetic reasons with voluminous masses (≥ 5 cm) [13,16]. Even though the subcapsular region is a richly vascularized anatomic area with frequent risk of hematoma, complete excision with healthy surgical margins can be easily achieved [8,9].

EFD presents a typical macroscopic and histological aspect, as did the case in our observation. Macroscopically, the tumor was firm and well circumscribed, nonencapsulated, measuring between two and 10 cm in greatest dimension [13,18]. The specimen was grayish-white in color alternating with zones with a fatty aspect. The microscopic study demonstrated a pseudotumoral lesion made up of elastic fibers that had a characteristic morphology within fibrous and adipose tissues. Electron microscope examination, performed in certain cases, specifies the anomalies of the elastic fibers, which appear finely granulous or fibrillar, occasionally with a moth-eaten aspect [13,16]. They are thick and branched in small globules with highly irregular contours admixed with wide bands of collagen and a few fibroblasts that appear active [8,9].

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 [14,15]. 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 [2,4]. 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 [7,16,20,21].

The clinical follow-up of patients who have undergone surgical excision or biopsy varies, depending on the study, between 1 month and 4 years, with a mean follow-up of 9 months with no complications [13,14,15]. Local tumor recurrence has been reported after incomplete excision [9,18]. No cases of malignant transformation have been described in the literature [8].

Conclusion

EFD is a frequent benign pseudotumor in the older subject, particularly in women, with subscapular topography that is quite often bilateral with a characteristic CT and/or MRI aspect comprising a stratified alternation of fibrous and fatty tissues. When the lesion is absolutely typical on imaging studies and it is asymptomatic, as it most frequently is, no complementary studies are necessary. However, surgical treatment can be proposed if the lesion is symptomatic or if doubt persists as to the lesion’s benign nature: complete resection with healthy surgical margins, thus allowing precise histological diagnosis.

Conflict of interest

None.

References


