



Elastofibroma Dorsi

Our experience with 11 lesions



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Elastofibroma dorsi. Our experience with 11 lesions

Elastofibroma is a rare benign lesion that typically arises on the posterior chest wall beneath the scapula. The etiopathogenesis is still unclear and the knowledge about its diagnosis and management mainly comes from small case series.

We collected all the data related to 11 Elastofibroma Dorsi treated at our institution between January 2003 and July 2014. The definitive diagnosis was made by histological examination of the mass. We analyzed the characteristic of the patients, (i.e. age, sex, dominant hand, and occupation), the presenting symptoms and signs, the preoperative investigations, surgical management and complications comparing our findings with the current medical literature on the subject.

We found that preoperative investigations are of limited value and when clinical presentation is typical the diagnosis could be made presumptively. Surgery should be performed under general anaesthesia since local anaesthetic infiltration and sedation are often inadequate to reduce intraoperative discomfort.

KEY WORDS: Elastofibroma

Introduction

Elastofibroma is a rare, slow-growing, benign soft tissue lesion characterized by fibroblastic proliferation and accumulation of abnormal elastic fibers, described for the first time by Jarvi and Saxen in 1961¹. Since then, elastofibroma has been described in different anatomic regions²⁻⁵ but it typically arises between the ribs and the muscles of the posterior thoracic wall beneath the scapula (hence Elastofibroma Dorsi, ED)⁶. It usually occurs in the elderly (aged over 50 years⁷), with a predilection for female patients⁸, it is extremely rare in children⁹

and it is often bilateral. Most often it is asymptomatic. When present, symptoms may include the presence of a mass, shoulder stiffness, scapular "snapping" and pain¹⁰. The etiology is a source of ongoing debate¹¹⁻¹³: whether ED is a true neoplasm or a reactive fibrous lesion remains uncertain. What is known about elastofibroma mainly comes from limited case reports and small series found in medical literature. We present a series of 8 patients (10 tumors) who were treated at our institution. We analyze the patient's clinical characteristics, features of the lesion, imaging findings, surgical approach and discuss issues surrounding its management while comparing our experience to the literature.

Patients and Methods

We reviewed the medical records of all Elastofibromas histologically diagnosed and surgically treated at our institution (Plastic Surgery Department) between January 2003 and July 2014. Eleven tumors were found in 9 patients. We considered the patient's age, gender, occupation, dominant hand as well the tumor size, location

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and associated symptoms. Preoperative investigations, surgical management and complications are also evaluated. We also called back and re-examined patients to assess any recurrence.

There was no external funding source for this study.

Results

All the patients came to our observation with a bulging mass under the scapula. Of these, seven tumors caused different degrees of discomfort ranging from simple “feeling of a mass” to pain during movement of the shoulder. The remaining four lesions were asymptomatic and were removed because of the patient’s will (Table I for details). The tumor was located on the right side in three patients, on the left side in four, and on both sides in two. The patients ranged in age from 58 to 75 years at the time of surgery (mean 68.2 years); six were women and three were men. None of them reported a family history of the lesion. Their occupations were respectively: a cleaner, a greengrocer, an hairdresser, three office workers, an housewife, a craftsman and a farmer. The craftsman, the cleaner, the greengrocer and the farmer reported daily hard and repetitive movements of their arms, especially the dominant hand. The remaining patients reported different degrees of manual activity in their lives more or less related to their job but the efforts weren’t noteworthy. One of the patients whose lifestyle was quite sedentary reported two falls down the stairs on two different occasions in which she hit the scapular region of the right side and left side respectively. The mild traumas happened some months before noticing the masses on the back.

The lesion involved the dominant hand side in four of the patients with single lesion (four out of seven). The location was the same in all patients (i.e. at the lower

scapular angle). On physical examination the lesion was almost completely covered and hidden by shoulder blade when the ipsilateral arm was adducted and became apparent when the arm was elevated and/or internally rotated (Fig. 1-2). Most of the times this “popping” of the lesion from underneath the scapula when rising the arm was accompanied by a snap felt by the patient. In those patient with scant subcutaneous adipose tissue the elevation of the scapula could be appreciated when the arm was hanging relaxed and close to the body (Fig. 1)

All the lesions underwent sonography (US) and six ultrasound-guided fine needle aspiration biopsies (FNAB) were performed.

The US appearances of the lesions were quite different from one another: some lesions looked more hyperechoic, other was mainly hypoechoic, some showed sharp margins, some had poorly defined boundaries with tissues surrounding them. In all US reports the lesions were described as a mass with “inhomogeneous echogenicity” with benign characteristics, two were suspicious for fibrolipoma, four were suspicious for lipoma and the remaining four were not specified.

Computed tomography (CT) has been performed in just one case and an Magnetic Resonance Imaging (MRI) in another one. The CT revealed a mass with “fatty features compatible with lipoma” while the MRI described a “well-demarcated solid mass with patchy hyper intense signal, most likely an elastofibroma”.

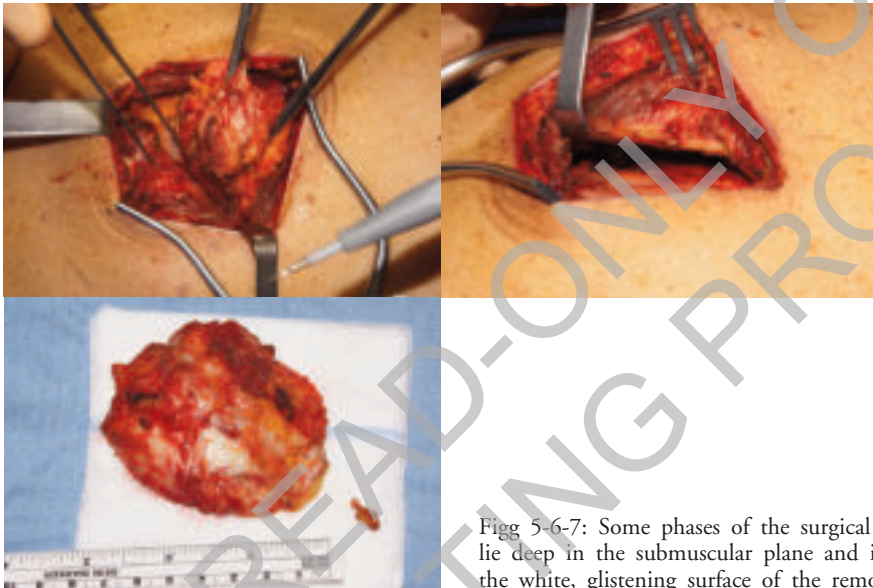
The mean long axis of the mass was 5.2 cm (range 3-11 cm). Four of the cytology reports showed “fibroadipose elements and scattered adipose cells”, the remaining two were referred as “mesenchymal proliferation”. Low cellularity of the specimens was a common feature. Seven interventions were performed on an outpatient basis (Day Surgery) under local anesthesia while in four occasions the patient was hospitalized and the excision was performed under general anesthesia. The masses were



Figg. 1-2: Patient with right sided elastofibroma dorsi. Elevation of the scapula is a characteristic finding in thinner patients with quite large tumor (Fig. 1). The mass becomes apparent when the upper limb is abducted (Fig. 2).



Figgs. 3-4: Left sided elastofibroma. In this patient no masses can be seen in upright relaxed position. Elevation of the scapula is hidden by thick subcutaneous tissue (Fig. 3). Note the bulging mass when the patient's shoulder is internally rotated (Fig. 4).



Figgs 5-6-7: Some phases of the surgical excision of the ED showed in fig 3-4. The mass lie deep in the submuscular plane and is partially covered by the overlying scapula. Note the white, glistening surface of the removed mass.

found in the submuscular plane at the level of the lower scapular angle, deep to the serratus anterior muscle, rhomboid muscle, and latissimus dorsi. They were not encapsulated, strongly attached to ribs and surrounding tissues. Ten lesions underwent complete surgical excision, one underwent only partial removal. The wounds were closed by multiple-layers interrupted sutures in layers over a suction drain. The mean operating time was 78 minutes (range 50- 110 minutes). There have been no true intraoperative complications but the efforts required to eradicate the tumors and the extent of the operations were greater than predicted due to the strong attachment of the masses to surrounding tissues deep in the submuscular plane. Every specimen has been sent to

histopathologic examination which then proved to be elastofibromas. Three of the pathology reports were very concise giving the diagnosis of elastofibroma without adding any other information. The remaining eight described a gray-white tumor with fibro-elastic, rubbery texture on gross examination and mature fibrous tissue with adipose lobules interspersed between thick collagen strands on histology slides. One pathology report specified that the tumor was not encapsulated; other reports added no further information concerning the presence or absence of a capsule.

Four cases were complicated postoperatively by a seroma, drained by large-bore syringe 14 days after surgery in one case and at 13 days in the other three cases.

Three of these seromas recurred five times and were abundant. The mean follow up time of nine out of ten lesions was 5,5 years (range 6 months- 8 years).

In this period the patient with bilateral elastofibroma had a recurrence on both sides but she refused further operations.

One tumor (left shoulder in patient n 8, see table) has just undergone surgical excision at the time of writing so data on post-operative complications and recurrence are not available.

Discussion

ED is a slow-growing benign tumor, asymptomatic in most of patients ¹⁴. Therefore the excision of an Elastofibroma is needed just in symptomatic cases and/or if the patient is concerned about it, or wants to remove it for a cosmetic reason. In our eight-patient series we encountered most of the elastofibroma typical clinical features. The mean age of the patients (68.2 years at the time of intervention) and female predominance are in

keeping with the literature ^{15,16}. The location deep to the serratus anterior muscle, rhomboid muscle, and latissimus dorsi at the level of the scapular angle is so distinctive that some authors consider it almost pathognomonic ¹⁴ and was constant in our cases. On the other hand, the association between the presence of the tumor, handiwork and hand dominance as reported in other series ^{11,17} is not clear. Just five of the patients reported some repetitive efforts of upper limbs and just four out of seven patients with monolateral ED had the lesion on the dominant side. It is interesting to note the similarity between the patient number 9 (see table) and the case reported by Heck et al ¹⁸ in which bilateral contusion to shoulder girdle preceded by a few months the expansion of the masses.

With regards to preoperative investigations, our analysis revealed that all the tests (but one) made in our eight cases before the histopathology slides missed the diagnosis and most of them were suspicious for other lesions rather than Elastofibroma, which demonstrates the lack of a reliable tool able to give an accurate diagnosis pre-operatively in our institution. Such inability to reach a

TABLE I - Details of patients and tumours

Patient	Age/ sex	Side of the lesion	Hand dominance	Occupation- level of arm activity	Time before surgery	Symptoms	Intraoperative pain (NRS)	Complications
1	63/ f	left	Right	Cleaner. Heavy manual labour	About 8 years	Asymptomatic	8	seroma
2a	75/f	left	right	Greengrocer. Heavy manual labour	About 4 years	Asymptomatic	9	none
2b	77/f	right				Feeling of a mass	5	none
3	66/m	right	right	Craftsman. Heavy manual labour	1 year	Asymptomatic	7	none
4	70/f	right	right	Office worker. Frequent activities involving arms	2 years	Feeling of a mass. Scapular snap while elevating the arm. Pain to right-side ribcage irradiating anteriorly to the right breast	10	none
5	65/f	left	right	Hair dresser. Mild manual labour.	About 3 months	Feeling of a mass. Shoulder stiffness. Scapular snap while elevating the arm	6	seroma
6	58/f	right	right	Housewife. Mild manual activities	8 months	Asymptomatic	General anesthesia	seroma
7	70/m	left	right	Office worker. average manual activities	About 5 months	Feeling of a mass. Scapular snap	8	none
8	58/m	left	Left	Farmer, hard manual activities	About 1 year	Feeling of a mass. Scapular snap	General anesthesia	Missing data
9a	73/f	left	right	Office worker. average manual activities. Two falls down the stairs with mild shoulder trauma.	Few months	Feeling of a mass. Scapular snap	General anesthesia	seroma
9b	75/f	right			Few months	Mild pain	General anesthesia	

* Patient number 2 and patient number 9 had bilateral metachronous elastofibromas (a: left side; b: right side)

diagnosis preoperatively is probably due to the little interest and limited knowledge about this lesion, given its benign nature, its rarity and its overall low impact on a patient's life.

At present in the literature there are no widely accepted investigations to confirm the clinically suspected ED: Some authors favor US^{19,20}, which describes characteristic echogenic features of the mass, others promote CT and MRI^{21,22}. MRI seems to be the most reliable but a cost-benefit analysis hasn't been done yet. In our personal experience we saw that the US failed to recognize the EDs. The US were performed almost each time by different operators so we can rule out a single-operator inability to identify it and we couldn't find any common highly characteristic sonography pattern among different lesions. On the other hand all US reports confidently described a lesion "with benign features" (e.g. vascular pattern, lack of surrounding tissues infiltration) allowing us to give the correct priority to the problem and described quite precisely the localization of the mass deep to the muscular planes. US were followed by Fine Needle Aspiration Biopsy in six tumors. The results of cytology haven't been able to suspect elastofibroma and didn't add any precious information in making differential diagnosis if compared to imaging alone. There are few publications supporting Fine Needle Aspiration Biopsy in the diagnosis of ED²³. The tumor has low cellularity and the cell sample is often inadequate²⁴. Furthermore cytology cannot easily differentiate some of high-grade soft tissue malignant tumors from their benign counterparts (e.g. high grade malignant liposarcoma - benign lipoma) making the FNAB in our opinion an useless exam in this particular field. Again, the only CT report described the position, size and shape of the lesion but missed the diagnosis. The MRI - as previously cited - was the only exam that clearly confirmed the suspect of elastofibroma.

Some considerations must be made about the surgical approach to these tumors. For this retrospective analysis on ED, we contacted all patients whose tumor had been removed under local anesthesia (all cases but three operated under general anesthesia) and using the Numerical Rating Scale we asked about the pain felt during the intervention. The mean value was 7,42 with a range of 5-9. In some cases a iv sedative has been administered intraoperatively in addition to local anesthetic to help pain control. These data associated with a mean operating time of 78 minutes highlight that the complexity of the excision was underestimated and a general anesthesia could be a better choice. The difficulty of dissection is mostly given by the tenacious adhesion of the tumor to the periosteum of ribs and surrounding muscles.

The high- rate of post-operative complications seen in our series and reported in literature¹⁰ suggest that all efforts should be made to prevent fluid collections (hematomas and seromas): accurate hemostasis is mandatory, dead spaces should be obliterated by multilayered

suture and a highly compressive dressing, a suction drain and immobilization of the involved arm are strongly advisable.

Conclusions

The main problem emerging from our review of ten cases is obtaining an accurate and reliable diagnosis preoperatively. In our opinion the physical examination is the first and the most important step in the diagnosis of ED. Ultrasound proved to be a cost-effective tool in confirming the subscapular and submuscular location of the lesion, giving its size and roughly suggesting its benign nature. When the clinical presentation of these benign neoplasms is typical (as we observed in most of our patients) we can consider the fine needle aspiration biopsy to be a unnecessary procedure. Further imaging and tissue sampling using a core-biopsy to get a representative tissue specimen is advisable when there are any concerns about the nature of the mass. Surgical excision, as revealed by mean operative time and intraoperative pain reported by patients, need to be performed under general anesthesia in a hospitalized patient.

Riassunto

L'elastofibroma è una lesione benigna rara che tipicamente insorge sulla parete posteriore del torace al sotto la scapola. L'etiopatogenesi è ancora poco chiara e la conoscenza sulla sua diagnosi e gestione deriva principalmente dalle piccole serie di casi.

Abbiamo raccolto tutti i dati relativi ad 11 casi di Elastofibroma Dorsi trattati nel nostro Istituto tra gennaio 2003 e luglio 2014. La diagnosi definitiva è stata fatta mediante esame istologico della massa. Abbiamo analizzato le caratteristiche dei pazienti (come età, sesso, mano dominante, occupazione), i sintomi ed i segni all'esordio, gli esami preoperatori, la gestione chirurgica e le complicanze, comparando i nostri dati con la letteratura medica corrente sull'argomento.

Dalla nostra analisi emerge che gli esami preoperatori sono di valore limitato e quando la presentazione clinica è tipica la diagnosi può essere fatta presuntivamente. L'intervento chirurgico dovrebbe essere eseguito sotto anestesia generale, poiché l'infiltrazione anestetica locale e la sedazione sono spesso inadeguate per ridurre il dolore intraoperatorio.

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