

Elastofibroma: clinical results after resection of a rare tumor entity

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Abstract

Elastofibroma (EF) is a benign proliferation of connective tissue and is typically located at the dorsal thoracic wall. Most patients complain about pain during motion in the shoulder girdle. The aim of our study was to evaluate the outcome after surgical treatment of EF. This study provides an overview of typical clinical findings, diagnostics and pathogenesis of this rare entity. In this retrospective study we analyzed data of 12 patients (6 male, 6 female) with EF treated in our institution between 2004 and 2012. The mean follow-up was 4.7 years (range: 5 months to 7.5 years). All tumors were found to be unilateral and all patients had a negative medical history for EF. Visual analogue scale and range of motion (ROM) was documented pre- and postoperatively. In all patients indication for surgical resection was pain or uneasiness during movement. There was no statistically significant difference in ROM of the shoulder between pre- and postoperatively but all patients reported significantly less pain after surgical resection. Patients benefited from tumor resection by a significant reduction of pain levels and improvement of the motion-dependent discomfort.

Introduction

Elastofibroma (EF) dorsi, which was first described by Jarvi in 1961,¹ is a rare and slowly growing benign lesion being typically located at the posterior thoracic wall. The reported incidence is 0.23/100,000.² Computed tomography (CT) studies have revealed a prevalence of 2%.^{2,3} Patients with EF are mostly female and over the age of 50.⁴ Manifestations in children

are extremely rare.⁵ EF is histologically characterized by an increased fibroblastic proliferation with an accumulation of elastic fibers (Figure 1). The World Health Organization classified the lesion in 2002 as a *benign fibroblastic/myofibroblastic tumor*. Etiological features of this entity are still under discussion and remain uncertain. Geographic and familial accumulations point towards a genetic component,⁶ while some authors postulate that repetitive mechanical micro-injuries may cause an increased proliferation of myofibroblastic fibers. Progressive loss of range of motion (ROM) of the shoulder caused by pain and uneasiness eventually becomes functionally debilitating. Due to mechanical irritation patients with EF often complain about sleep disorders. During clinical examination, a palpable and movable tumor, which usually does not show distinct margins to the surrounding soft-tissue, is the typical finding. Due to the rareness of this disease, patients often experience a delayed diagnosis.

This study presents the surgical outcome of 12 patients diagnosed with EF. Clinical features as well as diagnostic and therapeutic options are discussed. A critical review of the literature is given.

Materials and Methods

In this retrospective study, all patients diagnosed with EF were identified from our institutional tumor database. Over a period of 8 years, twelve patients, including six males and six females, were treated by surgical resection of this tumor entity (Table 1). Mean follow-up was 4.7±2.5 years (range: 5 months - 7.5 years). The mean age at the time of surgery was 59.7±8.2 years (range: 50-74 years). Of these patients 58.3% (n=7) were manual workers (either currently or retired), 41.7% (n=5) patients had no history of manual hard work.

Patients reported a slowly growing mass on their back during a period of several years, except two patients (No. 3 with pelvic location, No. 12 with EF of the hand). Furthermore, all patients reported a loss of ROM in their shoulder joint with uncomfortable mechanical irritation. Notably, one patient had experienced an incomplete resection under local anesthesia elsewhere (patient No. 5). In another patient, the tumor has developed near the iliac crest following a local abscess-operation more than 20 years ago (patient No. 3).

All patients underwent plain radiography of the shoulder (or pelvis/hand), magnetic resonance imaging (MRI), and sonography. Radiographically, there were no signs of malignancy in any of our patients. All lesions were located unilaterally. There was no positive medical history. After detailed informa-

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tion, all patients opted for a surgical resection of the tumor.

Statistical analyses

All data were analyzed with SPSS 21 software (SPSS Inc., Chicago, IL, USA). For descriptive statistics values were reported as the mean, standard deviation and range. The Shapiro-Wilk-Test was used to test for normality. To analyze statistical differences between the pre- and postoperative status, we used the t-test and the Mann-Whitney Rank Sum Test, respectively. The level of significance was set at P≤0.05.

Magnetic resonance imaging findings

In those ten patients with EF around the scapula, the tumor was found *loco typico* at the inferior pole of the scapula adjacent to the posterior thoracic wall, between the latissimus dorsi, the serratus anterior, and the rhomboid muscles ventrally. Usually the lesion was poorly defined rather than a capsulated tumor. In T1 (TR 400-700/TE 15-40) and T2 (TR 2500-3000/TE 80-120) weighted magnetic resonance imaging (MRI) sequences, the tumors revealed a characteristic signal similar to skeletal muscle with regions of alternating high and low signal intensities (Figure 2). Bilateral MRI or contrast enhancement was not performed routinely. Static and dynamic sonography was added to the diagnostic routine for preoperative localization of the tumor mass and planning of the surgical approach.

Results

In all patients, the lesion was removed with histologically tumor-free margins (Figure 3). Four patients had a biopsy prior to resection, while eight patients were operated on without a pre-operative biopsy. After the resection of the tumor mass, two patients developed a mild local seroma at the thoracic wall. These patients were treated with puncture under sterile conditions, elastic compression and temporary immobilization of the shoulder for 7 days. In all other patients the postoperative course was uneventful without wound infection or other complications. Although the ROM was improved, there was no significant difference in ROM of the shoulder girdle between the pre- and postoperative status. Mean flexion increased from 164° to 169°, mean extension increased from 27 to 28°, mean abduction increased from 165° to 169°, and mean external rotation increased from 71° to 72°. The patients' pain level was significantly decreased when compared to pre-operatively (preoperative VAS of 4 versus postoperative VAS of 0.4, $P \leq 0.001$) (Table 2). Tumor size seems to have no effect to preoperative symptoms or surgical outcome in our group. There was no local recurrence or new occurrence at the time of evaluation.

Discussion and Conclusions

EF is a benign tumor, which may significantly affect the wellbeing of the affected patients due to its periscapular localization. As it is a rare entity, diagnosis is often delayed. Ninety-nine percent of elastofibromas are located in the area between the inferior angulus and medial margo of the scapula ventral to the serratus anterior muscle between the 6th and the

8th rib (Figures 2 and 4).⁷⁻⁹ Most of the patients are able to shift the tumor with an individual maneuver (elevation and/or abduction of the shoulder). The *hidden* tumor dislocates from the thoraco-scapular space towards the medial margo of the scapula to a more medio-dorsal position and impresses as a mobile subcutaneous mass (Figure 4). The decreasing ROM of the shoulder is a result of the mechanical irritation and discomfort caused by the tumor.^{9,10} Though, some patients have a free ROM.^{3,8} In our patient cohort, there was only a slight difference between the range of motion pre- and postoperatively without statistical significance. This might be due to the fact that most patients in our study presented already preoperatively with a relatively free ROM.

Although the most common area is periscapular, other localisations have been reported: tricuspid valve,⁷ intraspinally,¹¹ ligamentum flavum,¹² axilla,⁷ hand,¹³ (Figure 5) intra-articular,¹⁴ ischial tuberosity, foot,¹⁵ deltoid,⁷ olecranon,⁷ and greater trochanter.¹⁶ In our study cohort, one patient had an EF near the iliac crest after surgical resection of an abscess 20 years ago (Table 1). In the elderly patient population (>60 years), the prevalence of elastofibroma dorsi diagnosed by computed tomography (CT) is 2%.³ Notably, an autopsy series revealed a much higher rate of EF (11.2% in males, 24.4% in females) in patients over the age of 55 years, whereas no case was reported under the age of 55.¹⁷ Pre-elastofibroma changes, which are defined as mild elastophilic fibrillar accumulation without a clear elastic tissue formation, were observed in 81% in an autopsy series of 100 patients.¹⁸ However, many of these cases revealed only mild streaks of abnormal elastic tissue. These variations in the prevalence of EF are most probably due to sensitivity limitations of CT and related to insufficient contrast resolution and an inability to differentiate streaks of nor-

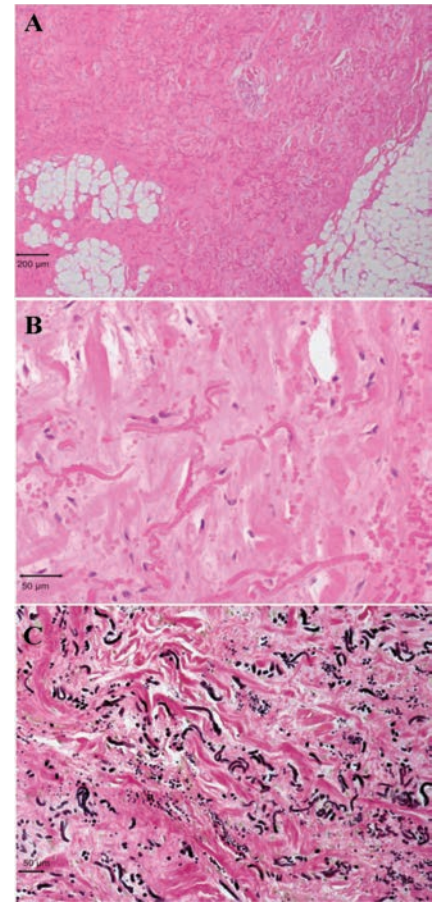


Figure 1. A) Paucicellular tissue composed of mesenchymal cells, fibrous, collagenous strands and elastic fibers, with irregular interdigitation into mature adipose tissue (hematoxylin-eosine-staining). B) Collagen bundles alternate with large, thick eosinophilic elastic cylinders with a dense central core interspersed with mesenchymal cells without atypia (hematoxylin-eosin-staining). C) Elastic stain demonstrates the presence of abundant elastic fibers and associated detached elastin globules (Elastica-van-Gieson).

Table 1. Patients' characteristics.

ID	Sex	Location	Tumor size, cm	Side	Therapy	Complication	Age at surgery	Follow-up, months
1	M	Periscapular	8.4×6.7×2.6	Left	Excision	-	60	87
2	M	Periscapular	6.6×5.7×2.0	Left	Excision	-	54	74
3	F	Iliac crest	10.0×7.1×7.3	Left	Excision	-	67	52
4	F	Periscapular	8.4×5.6×1.6	Right	Excision	Seroma	74	12
5	F	Periscapular	4.8×3.8×1.4	Left	Excision	Seroma	51	6
6	F	Periscapular	8.5×6.3×2.3	Right	Excision	-	62	82
7	F	Periscapular	7.2×6.1×3.1	Right	Excision	-	52	90
8	M	Periscapular	6.2×4.1×1.4	Left	Excision	-	56	76
9	M	Periscapular	5.2×4.7×2.0	Left	Excision	-	50	71
10	F	Periscapular	4.0×2.4×2.2	Left	Excision	-	51	64
11	M	Periscapular	9.0×6.2×2.5	Left	Excision	-	67	58
12	M	Hand	7.3×6.1×2.7	Left	Excision	-	72	5

mal tissue from abnormal tissue, as EF can only be detected via CT-scans if enough elastic tissue is present to appear as a mass.¹⁷ In a Spanish study, the incidence of clinically relevant elastofibroma was reported to be 0.23/100,000/year for both genders.²

An extensive Pubmed search revealed that in 57 years (January 1955 to April 2013), 283 studies on EF were published. Most of these studies are case reports or small case series, except for the study of Nagamine *et al.*⁹ in 1982, with a study sample of 170 cases and the study of Lococo *et al.*¹⁹ with 71 cases (Table 3). The pathogenesis of this tumor entity remains uncertain. Two main theories have been discussed: i) the origin of this lesion is a repetitive mechanical microtrauma in the periscapular region; ii) it is a *real* tumor that is caused by genetic alterations.

The tumor seems to develop within areas that are exposed to increased mechanical stress. Some authors postulate that friction between the thoracic wall and the scapula during movement in the shoulder girdle is a main pathogenic feature. This recurrent damage leads to local tissue reaction and the production and accumulation of elastic fibers.^{6,7,10,20} The main localization of this tumor at the inferior pole of the scapula is supporting this theory and studies have demonstrated that patients with increased physical activity are affected.¹⁰ However, this hypothesis does not explain the occurrence of these lesions in atypical sites (hand, foot, elbow, intraoral, intraspinal, ischial tuberosity, etc.). In addition, most of the case series reported in the literature provide limited information on physical activity (as a predisposing factor), and hence, this theory cannot be proven.

Others reported genetic alterations associated with EF, which lead to the conclusion that this lesion is a true neoplasm. Based on microarray *comparative genomic hybridisation* (CGH), Hernandez *et al.*²¹ found a modification in DNA-sequences at the chromosomal locations 1p, 13q, 19p and 22q. Nishio *et al.*²² reported an increase of DNA copy numbers on the long arm of the X chromosome using CGH. This hypothesis is supported by studies on the

local geographic accumulation of EF *i.e.* in the prefecture of Okinawa in Japan.⁹

The most commonly used diagnostic methods are ultrasound, CT-Scans and MRI. Some authors have reported an incidental detection of EF during a Positron-Emission-Tomography (PET)-scan. The soft tissue mass of EF is characterized by a diffuse uptake and low intensity of Fluorodeoxyglucose (18-FDG), demonstrating a benign lesion with a low level of metabolic activity.^{7,23,24} However, due to its costs and radiation exposure to the patient, PET scans are not a first line diagnostic modality.

Bianchi *et al.*, who performed a sonographic characterization study of EF, revealed that this lesion is characterized by a well-defined, distinct, multi-layered echo signal, caused by fibro-elastic streaks and fat. Thus, the authors assume an EF if: i) these sonographic patterns are observed in ii) female patients above 50 years of age iii) in the subscapular area.²⁵ These observations are similar to those of other studies and describe pathognomonic criteria.²⁶⁻²⁸ In this study, the authors recommend a CT or MRI if the typical multi-layered pattern is missing on ultrasound.²⁵

Nevertheless, daily practice has shown that, not ultrasound or CT, but rather MRI scans are a suitable diagnostic tool to closer identify the entity and dignity of a tumor. By means of MRI, EF has been classified into various subtypes: i) inhomogeneous, fasciculated type (type A), ii) inhomogeneous nonspecific type (type B), and iii) homogeneous muscle isodense or iso-intense type (type C).² The authors reported a sensitivity of 100% and a mean positive predictive value of 93.3%.

As previously mentioned, it is our standard to perform MRI studies in all patients with unclear soft tissue tumors especially if a malignant lesion cannot be excluded with certainty and an excision without prior biopsy is planned.²⁹⁻³¹ We are not aware of any reported case of metastasizing EF. Therefore, if metastases are present, EF may not be the correct diagnosis and further investigations are necessary to analyze the primary tumor.

Indication for surgical resection is always an individual decision and a preoperative dis-

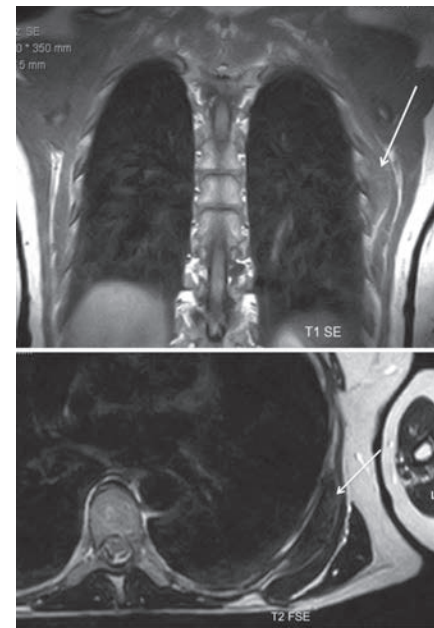


Figure 2. Magnetic resonance imaging of a 51 y/o female patient (No. 5) in T1 (coronal plane) and T2 (transverse plane) sequences showing the subscapular location of the elastofibroma. The tumor is located between the M.serratus anterior and M.latissimus dorsi and the thoracic wall.

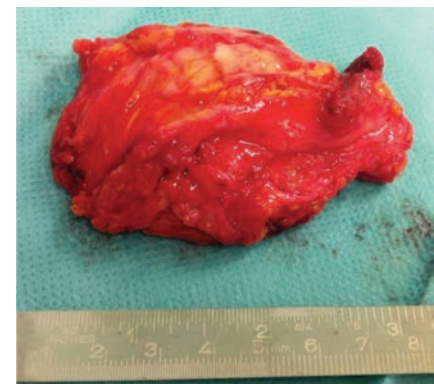


Figure 3. Macroscopic aspect after excision: mild yellow colored elastic fibers cover the tumor.

Table 2. Mean range of motion and visual analogue scale.

	Pre-operatively	Post-operatively	P
Flexion	164 (± 9.2)	169 (± 8.3)	0.241
Extension	27 (± 6.8)	28 (± 6.0)	0.934
Abduction	165 (± 8.1)	169 (± 7.0)	0.276
Adduction	25 (± 5.0)	26 (± 6.6)	0.865
External rotation	71 (± 9.4)	72 (± 8.7)	0.872
VAS	4 (± 0.7)	0.4 (± 0.4)	<0.001

VAS, visual analogue scale.

Table 3. Review of the literature showing diagnostic procedures, complications and recurrence rates.

Author	Year	No.	Diagnostics	Therapy	Complication	Recurrence	Bilateral
Battaglia	2009	43	US, CT, MRI	Excision	-	-	54%
Chandrasekar	2008	15	MRI (3), biopsy (3), trucut biopsy (8), open biopsy (1)	Excision	-	-	13%
Cota	2006	1	US	Excision	-	-	0
Gao	2010	6	CT (4), MRI (6)	Excision	?	-	17%
Haykir	2006	2	CT (1), MRI (2)	Excision	-	-	0
Lococo	2013	71	CT (47), MRI (44), US (37)	Excision (66), observation (5)	hematoma (6%), seroma (7%), fever (4%), other (1%)	4.5%	16.9%
Muratori	2007	8	CT (8), MRI (8), US (8)	Excision	-	-	12%
Mortman	2007	6	CT, MRI	Excision	?	17%	0
Nagamine	1982	170	Biopsy (136)	Excision	?	0.006%	73%
Nishio	2012	11	MRI, biopsy (1)	Excision (5), observation (6)	seroma (10%)	-	18%
Marino	2012	14	MRI	Excision	-	10%	50%
Parrat	2010	15	MRI, CT-guided biopsy (4)	Excision (12), observation (3)	hematoma (26%), seroma (20%)	7%	40%

CT, computed tomography; MRI, magnetic resonance imaging; US, ultrasound.



Figure 4. With a combination of rotation and abduction of the right shoulder, this female patient (No. 4) was able to dislocate the tumour from the scapulo-thoracic space towards the medial edge of the scapula.

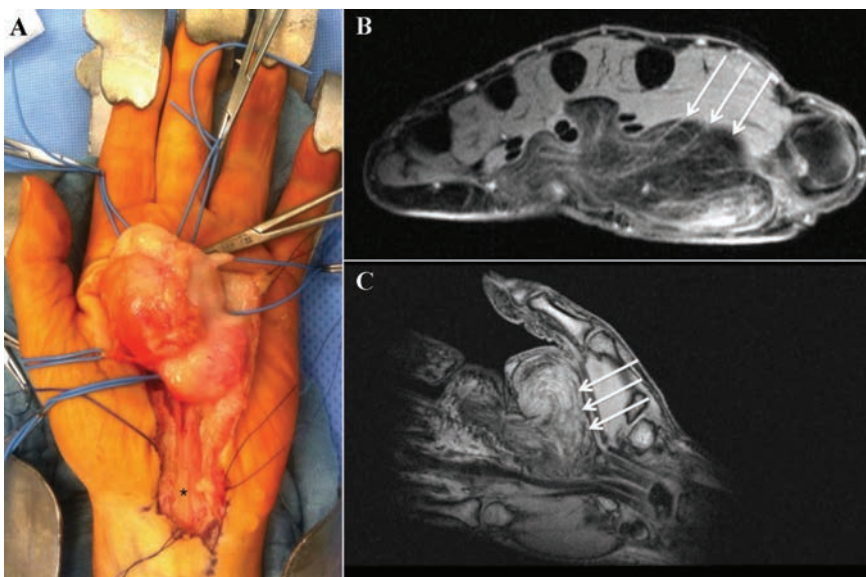


Figure 5. A) Elastofibroma of the hand (Pat No. 12). After resection of the flexor retinaculum the median nerve and its branches are visible (*). B) and C) MRI of the hand (T1 weighted, coronal and sagittal). Elastofibroma is marked with white arrows.

discussion with each patient is necessary especially when the patient is relatively symptom-free. In our case series, the resection of the tumor mass did not show a statistical improvement of ROM in the shoulder girdle. However, there was a significant pain reduction postoperatively (mean VAS 4 preoperatively to 0.4 postoperatively). Although long-term complications are rare, as noted in our study sample, temporary complications such as postoperative seroma or hematoma may occur especially if the resected tumor mass had large dimensions.^{2,6,10} The tumor size has no correlation to symptoms or surgical results in our group. Maybe this is due to the small number of patients (n=12).

In conclusion, in EF-patients with typical motion-dependent discomfort and pain in the shoulder girdle, surgical resection is safe. A biopsy prior to tumor resection is not necessary if all pathognomonic criteria are present and a soft tissue sarcoma is excluded via MRI.

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