

ORIGINAL ARTICLE

A thoracic surgeon's perspective on the elastofibroma dorsi: A benign tumor of the deep infrascapular region

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Keywords

chest wall tumor; elastofibroma; soft tissue tumor.

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Received: 23 March 2012;
accepted 13 April 2012.

doi: 10.1111/j.1759-7714.2012.00139.x

Abstract

Background: An elastofibroma is a benign, soft-tissue tumor and is important in the differential diagnosis of thoracic wall masses. Here, patients with elastofibromas who underwent thoracic surgery were retrospectively reviewed to elucidate elastofibroma formation and to facilitate the differential diagnosis.

Methods: This is a retrospective and descriptive study of a series of 30 patients with elastofibroma dorsi. The data was obtained by review of the hospital records.

Results: There were 27 female and three male patients (mean age, 55.13 ± 8.7 years) with a total of 42 elastofibroma dorsi tumors (12 bilateral cases, 18 unilateral cases) diagnosed between January 2004 and October 2011. Twenty patients (67%) underwent surgery as a result of subscapular swelling and pain. In 10 (33%) asymptomatic patients, elastofibromas were found incidentally during a thoracotomy. Imaging methods in symptomatic patients included computerized tomography (15 cases), magnetic resonance (three), and ultrasonography (two). For five patients, fluorodeoxyglucose uptake values were available and revealed mild metabolic activity in the elastofibromas. Elastofibromas were significantly larger in symptomatic patients (8.15 ± 1.9 vs. 6.2 ± 2.3 ; $P = 0.02$). Exposure to long-term repetitive micro-trauma was a precipitating factor in 23 (77%) patients. Seroma formation, the most common surgical complication, was observed in 40% of patients.

Conclusion: The differential diagnosis of elastofibroma dorsi is straightforward, and preoperative histology is unnecessary when the clinical, radiological, and metabolic characteristics are known. Repetitive micro-trauma may predispose to hyperproliferation of fibroelastic tissue, and genetics may also play a role. Surgical treatment can be reserved for cases with severe symptoms.

Introduction

Elastofibroma dorsi (ED) is a benign, slow-growing, soft-tissue tumor of the deep infrascapular region. Jarvi and Saxen first identified this rare entity in 1959 and published their findings in 1961.¹ Most EDs are asymptomatic or present with mild symptoms. Symptoms may include swelling, shoulder pain, and scapular snapping.² Although the prevalence of ED is unclear, it was found in 2% of elderly asymptomatic patients in a retrospective evaluation of computed tomography (CT) scans.³ Autopsy studies in patients older than 55 years of age showed a higher

prevalence, up to 24%.⁴ These results indicate that EDs are not as rare as previously assumed. Typical localization and characteristic imaging of the tumor allow definitive diagnosis in most cases. The importance of ED is in the differential diagnosis of thoracic tumors. In this study, the records of patients with ED who underwent thoracic surgery in our hospital were retrospectively evaluated. Demographic data, tumor characteristics, predisposing factors, complications, comorbidities, and radiological images were analyzed. To our knowledge, this is the largest single-center series of surgically treated EDs to date.

Patients and methods

Patients

Between January 2004 to October 2011, 30 patients with a total of 42 EDs underwent surgery at the Thoracic Surgery Department of Baskent University Hospital, Adana, Turkey. The tumors were diagnosed by physical examination and radiological imaging, and preoperative tissue biopsy was not required in any case. The patients' data was acquired by retrospectively reviewing radiological and clinical records, and current information about the patients was obtained by phone calls and outpatient visits. The study population consisted of two clinically different groups. Group 1 comprised 20 patients (67%) who underwent surgery for a symptomatic subscapular soft-tissue mass, and group 2 comprised 10 patients (33%) in whom EDs were found incidentally during 1150 thoracotomies for miscellaneous pulmonary diseases.

Methods

Surgery

All patients received general anesthesia before surgery. The patients in group 1 were placed in the prone position, with both arms stretched forward. This allowed lateral movement of the scapula, giving better visibility. The skin was incised over the lesion, parallel to the medial margin and inferior angle of the scapula, with the latissimus dorsi preserved or dissected parallel to the muscle fibers. Due to the non-encapsulated structure of EDs, they were removed along with surrounding fatty tissue. The tumors were tightly adhered to the periosteum of the ribs and the muscles of the scapula, as if having originated from these sites. Following removal, Hemovac drains were routinely used to prevent seroma. In group 2, the incidental EDs were excised in much the same fashion, but the procedure was somewhat simplified by the wider surgical field of the posterior or posterolateral thoracotomy.

Radiological evaluation

Radiology of an ED typically shows a subscapular or infrascapular soft-tissue mass with a lenticular, poorly circumscribed, and non-encapsulated structure. CT and magnetic resonance imaging (MRI) reveal a soft-tissue mass with muscle attenuation or signal intensity interspersed with strands of fat attenuation (Fig. 1). On ultrasonography (US), EDs typically appear as an array of linear strands against an echogenic background, with neither a clear cleavage surface nor a specific vascular pattern readily visible. In group 1, EDs were diagnosed by CT in 15 patients, MRI in three patients, and US in two patients. CT and MRI were used together to

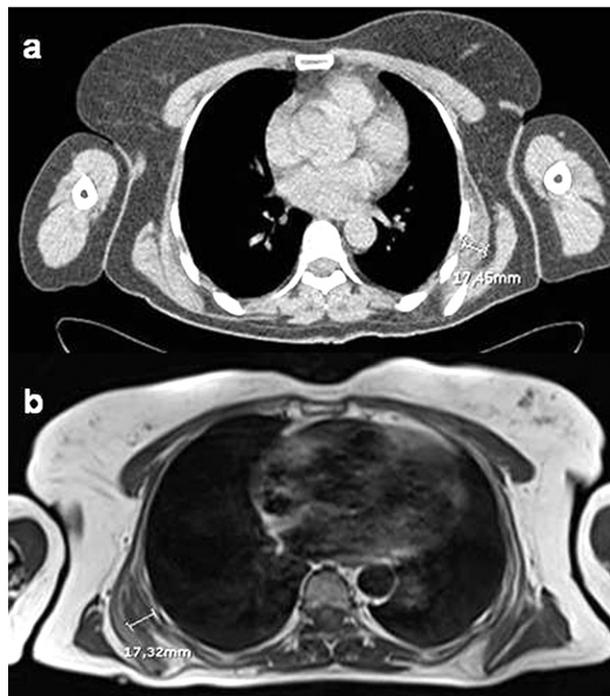


Figure 1 Radiologic images of a 56-year-old woman with bilateral asynchronous elastofibroma dorsi (ED). (a) Chest computed tomography (CT) scan demonstrates a left subscapular unencapsulated mass (February 2006). (b) Magnetic resonance imaging (MRI) of the same patient four years later. Demonstrates a right subscapular soft tissue mass with muscle attenuation compatible with an elastofibroma (June 2010). Both EDs were symptomatic.

confirm the diagnosis in two patients. In group 2, the patients were evaluated by CT for miscellaneous pulmonary diseases, but all of the EDs were overlooked in these imaging studies. Fludeoxyglucose (FDG) uptake values were available in the records of five patients (Table 1). Fludeoxyglucose-positron emission tomography (FDG-PET) was used to evaluate a painful subscapular mass suggestive of metastasis in one breast cancer patient in group 1. The other four patients were in group 2 and were evaluated with FDG-PET for staging of pulmonary cancer (Fig. 2).

Pathology

EDs are benign dermal tumors composed of branched and unbranched elastic fibers, eosinophilic collagen bundles, and scattered fatty tissue. The elastic fibers have a degenerated, beaded appearance or are fragmented into small globules or droplets arranged linearly. Immunohistochemically, EDs stain positive for vimentin and frequently stain positive for CD34 and lysozyme. EDs stain negative for smooth muscle actin, S-100, desmin, and p53.

Table 1 Positron emission tomography-computed tomography (PET-CT) findings of the patients with elastofibroma dorsi (ED)

Patient	Age (year)	Gender	Side	Malignancy	Size (mm)	FDG uptake value	Symptom
1	56	Male	Bilateral	Lung	60 × 50 × 30	1.8	Yes
2	58	Female	Left	Lung	50 × 40 × 25	2.7	No
3	58	Female	Right	Lung	60 × 45 × 35	2.2	No
4	60	Male	Right	Lung	70 × 45 × 30	2.1	No
5	50	Female	Bilateral	Breast	10 × 80 × 30 60 × 50 × 20	1.6 1.9	Yes

FDG, Fludeoxyglucose

Results

In our series, 27 women (89%) and three men (11%) with EDs underwent excisional surgery. The mean age was 55.13 ± 8.7 (range, 34–75 years). The most common symptom of ED was swelling, which was observed in 20 patients, although 10 patients (33%) were asymptomatic. Back and shoulder pain were present in 15 patients (50%). In addition, scapular snapping was noted in three patients. The overall mean (\pm SD) size of the EDs (long axis) was $7.50 (\pm 2.21)$ cm. The mean size of the EDs in group 1, which included symptomatic EDs, was significantly larger than that in group 2, which included EDs incidentally found during a thoracotomy (8.15 ± 1.9 vs. 6.2 ± 2.3 cm; $P = 0.02$). EDs were located bilaterally in 12 patients (40%), on only the left side in 10 patients (33%), and on only the right side in eight patients (27%). Only four patients (13%) were left-handed, while 26 (87%) were right-handed.

Seroma was the most common postoperative complication. It occurred in 12 patients (40%), and eight of these (27%) were readmitted because of swelling, fluctuation, and



Figure 2 Axial positron emission tomography (PET) image of a patient with non small cell lung cancer on the right lower lobe. PET-computed tomography (CT) demonstrates a high metabolic activity in the lung parenchyma (Suv max:16), and a moderate activity in left posterolateral chest (Suv max = 2,8). A smaller and less metabolically active lesion (arrow) is noted contralaterally (Suv max = 1.8).

pain at the surgical site. Chronic pain and discomfort were observed in five of the 20 patients (25%) in group 1; all five had undergone bilateral excision of large EDs. Group 2 was excluded from the pain analysis because pain due to ED excision might have been confused with chronic post-thoracotomy pain. During the 38-month mean follow-up period, no recurrence was observed.

Several possible predisposing or precipitating factors were identified (Table 2). First, ED was most commonly observed in middle-aged or older women, with the majority (24 patients, 80%) being menopausal. However, no hormonal disorder or menopausal drug use was recorded. Second, most of the patients had either a long-term (>10 years) manual labor occupation (four patients, 13%) or were housewives who knitted as a hobby (19 patients, 63%).

Discussion

EDs are connective tissue tumors classified as fibroblastic-myofibroblastic, benign, soft-tissue tumors. For most soft-tissue tumors, a relationship exists among the tumor type, symptoms, and location, and the patient's age and gender; EDs are no exception. Because of the nature of the tumor and its location on the chest wall, the differential diagnosis includes sarcomas and desmoid tumors. The most common symptoms are discomfort in the shoulder and back, which accounts for the interest of orthopedic surgeons in this topic. The current study evaluated EDs in patients who were referred to the thoracic surgery department, as well as EDs that were found incidentally during a thoracotomy.

EDs appear to be relatively rare, although the frequency may be higher than reported because EDs can be easily overlooked in many patients who are asymptomatic. In a survey study by Brandser, the prevalence of ED revealed by CT was 2% in an elderly population, with most of the masses having been missed on physical examination.³ In a similar study, a prevalence of 1.66% was found in patients who underwent positron emission tomography (PET)/CT for malignancies.⁵ In an autopsy series, Jarvi and Lansimies found EDs in 24% of women and 11% of men older than 55 years of age.⁴ This percentage was higher than expected. In another autopsy study, elastophilic tissue changes, which may be a precursor of ED,

Table 2 Characteristics of patients with elastofibroma dorsi (ED)

	Group 1 <i>n</i> = 20; 67%	Group 2 <i>n</i> = 10; 33%	Total/mean <i>n</i> = 30	
Patients				
Age	53	59	55.13 ± 8.7	
Gender				
Female	19	8	27	90%
Male	1	2	3	10%
Hand Knitting	15	4	19	63%
Menopause	17	7	24	80%
Concomitant diseases				
Diabetes mellitus	3	3	6	20%
COPD	1	3	4	13%
Lung cancer	∅	4	4	13%
Coronary artery disease	1	2	3	10%
Rheumatic diseases	2	1	3	10%
Hydatid disease	∅	2	2	7%
Bronchiectasis	∅	2	2	7%
Tuberculosis	∅	1	1	3%
Breast cancer	1	∅	1	3%
Tumor size (long axis;cm)	8.15 ± 1.9	6.2 ± 2.3	7.50 ± 2.2	
Tumor side				
Bilateral	11	1	12	40%
Left	6	4	10	33%
Right	3	5	8	27%
Dominant hand				
Right	18	8	26	87%
Left	2	2	4	13%

COPD, chronic obstructive pulmonary disease.

were identified in 81% of the patients.⁶ In our study, among 1150 patients who underwent a posterior or posterolateral thoracotomy for miscellaneous pulmonary diseases from 2004 to 2011, only 10 cases of ED were identified incidentally, representing a 0.8% prevalence of ED. Despite meticulous physical and radiological examinations, none of these EDs were noted before surgery. Naylor reported that among 16 patients, nine underwent surgery for ED, and EDs were incidentally found during thoracotomy in three patients.⁷ Kara *et al.* explained the importance of positioning during physical examination in order to diagnose an ED.⁸ Although an ED is palpable, often only proper positioning of the arm can make the lesion visible.

Elastofibromas are characterized by an accumulation of abnormal elastic fibers and are generally considered a reactive process. The tumor probably occurs as a result of repetitive trauma.^{9,10} Familial predisposition with an underlying enzymatic defect and hereditary factors may also be responsible for the development of EDs.^{11,12} Nevertheless, the initiating factor is unclear. Even though the surface of the scapula and the chest wall are exposed to continuous friction, neither is mechanically suitable to act as a joint. We believe that minimal degenerative changes in the subscapular region predispose to elastofibroma formation in response to repeated micro-trauma. The degenerative changes may be attributable

to various factors such as insufficient vascularity or genetic characteristics.^{4,13} In our study, the most common precipitating or predisposing factors were gender (females were over-represented), age (more common in elderly), and exposure to repetitive micro-trauma related to an occupation or hobby.

Nearly all EDs are observed attached to the deep musculature of the shoulder and very rarely occur in other parts of the body such as the hand.¹⁴ In our series, all of the elastofibromas were localized to the subscapular region. Preoperative tissue biopsy was not required in symptomatic patients, and the differential diagnosis of ED versus other chest wall tumors was not challenging, as the tumors showed typical radiological and clinical findings. Clinically, a unilateral or bilateral, subscapular, slow-growing swelling is typical. In one patient with bilateral asynchronous ED, the left ED had become visible and symptomatic over a 4-year period and was excised in 2006, after which a painful right subscapular swelling, about 6 × 4 × 2 cm in size, was observed and excised in 2010 (Fig. 1). Mild symptoms of an ED include discomfort of the back, arm, and shoulder, and snapping of the scapula. The size of an ED affects the degree of symptoms, and in our series, asymptomatic EDs were smaller than symptomatic EDs (6.2 vs. 8.2 cm, respectively; *P* = 0.02).

Typical imaging findings for ED have been described previously.⁷ Experienced radiologists have no trouble diagnosing

an ED by MRI or CT. Nevertheless, the diagnosis was missed in 10 of our patients. EDs were likely overlooked due to their lack of symptoms and because the radiologists and surgeons were focused on the primary pulmonary disorder. In bilateral ED, small lesions can be missed despite a careful evaluation.⁸ In recent years, FDG-PET has been used more routinely to evaluate malignancies. Although it is not necessary to perform a PET study to diagnose these lesions, it is important to identify EDs in order to avoid a misdiagnosis. In the few studies that addressed the role of PET-CT in ED identification, FDG uptake values of EDs were usually between 1.4 and 3.2. Uptake values in this range indicate mild or moderate metabolic activity that may predispose to an inflammatory or malignant process.^{4,15} In our study, FDG-PET was performed for pulmonary neoplasia in four patients, and the maximum standard uptake values ranged from 1.6 to 2.8. However, EDs in all four patients were diagnosed incidentally during a thoracotomy and were not mentioned until the cases were retrospectively reviewed. In one case, a woman with breast cancer was evaluated by PET-CT due to a subscapular swelling and back pain, which were attributed to possible metastases. Bilateral EDs with maximum standard uptake values of 1.8 and 2.0 were identified, and she underwent bilateral surgical excision. Our series and other reports have demonstrated mild metabolic activity in EDs. Although FDG-PET is controversial in the assessment of EDs, it may be useful in the differential diagnosis of malignant chest wall tumors.

Only a few reports have explored the role of radiotherapy for ED; the typical treatment is marginal excision.^{16,17} However, many clinicians believe that surgery is indicated only in severely symptomatic cases or when the diagnosis is in doubt.^{18–20} Fine-needle biopsy can be performed in patients who are not suitable for surgery. However, the diagnosis of ED by this method is difficult because the inherent hypocellularity of elastofibromas may appear histopathologically as non-specific fibromatosis.²¹ Thus, a core biopsy of adequate tissue size is usually needed to establish a specific diagnosis. Alternatively, marginal excision of the tumor may be preferred as a curative treatment. Wide surgical excision of an ED is not required, as neither malignant transformation nor metastases have been reported; marginal excision carries a very low risk for recurrence because of the benign nature of EDs.^{7,19} No recurrence was observed in our series during the mean 38-month follow-up period.

Seroma formation was the only postoperative complication observed in our study; it occurred in 40% of the patients. Although a small-bore Hemovac drain was routinely used following surgery, sterile fluid accumulation at the surgical site caused swelling and pain in some cases. Shoulder immobilization for one week and a compression garment were also recommended to avoid seroma.²¹ Most seromas improved spontaneously or were easily drained by fine needle aspiration during an outpatient visit. Postoperative chronic pain

was a serious problem in five (25%) out of 20 patients (group 1). In all patients with prolonged pain, EDs that were significantly larger than the other EDs had been excised bilaterally. A possible method of decreasing postoperative pain in such patients is to excise only symptomatic EDs, or divide the operation into two procedures.

The present study has some limitations because of its retrospective nature. However, this is one of the largest surgical series including both symptomatic and asymptomatic patients with ED, and it also provides insight into the mechanism of ED formation. In addition, PET-CT is evaluated as a new imaging modality for the diagnosis of ED.

Conclusion

Knowledge of the typical localization and imaging findings of ED is important for helping surgeons and radiologists diagnose this subscapular mass. Studies on predisposing and precipitating factors have indicated that this is an age- and gender-related process triggered by repetitive long-term micro-trauma. The mild metabolic activity of EDs in PET-CT can help to differentiate ED from other inflammatory and malignant diseases. Single-stage operations are feasible in patients with bilateral EDs, but are recommended only when both EDs are symptomatic or are very large.

Disclosure

No authors report any conflict of interest.

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