Elastofibroma dorsi management and outcomes: review of 16 cases

Ozgur Karakurt§, Tevﬁk Kaplan¶, Nesimi Gunal†, Gultekin Gulbaharµ, Bulent Kocer†, Serdar Han¶, Koray Duralc¶ and Unal Sakinci¶

§ Department of Thoracic Surgery, Ankara Numune Teaching and Research Hospital, Ankara, Turkey
¶ Department of Thoracic Surgery, Ufuk University School of Medicine, Ankara, Turkey
† Department of Thoracic Surgery, Kirikkale University School of Medicine, Kirikkale, Turkey
µ Department of Thoracic Surgery, Sincan State Hospital, Ankara, Turkey

* Corresponding author. Department of Thoracic Surgery, Ufuk University School of Medicine, Dr. Ridvan Ege Teaching and Research Hospital, Mevlana Bulvar (Konya Yolu) No: 86-88, 06520 Balgat - Ankara, Turkey. Tel: +90-312-2044201; fax: +90-312-2872390; e-mail: tevikkaplan@yahoo.com (T. Kaplan).

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Abstract

OBJECTIVES: Elastofibroma dorsi (ED) is a rare, benign lesion arising from connective tissue, usually found at the inferior pole of the scapula. To date, only a few small series have been reported in the English literature and there are few data about the long-term outcomes after surgery. Our goal is to contribute a better understanding of this tumour and to determine the long-term outcomes after surgery.

METHODS: Sixteen patients with a diagnosis of ED were identiﬁed from the unit’s database. The clinical presentation, diagnosis, pathological evidences and long-term outcomes were evaluated.

RESULTS: There were 11 females and 5 males with a mean age of 61.1 years (range 38–78 years). The tumour was located on the right in 5 (31.2%) patients, on the left in 6 (37.5%) patients and bilaterally in 5 (31.2%). Six patients had painful scapular swelling resulting in restriction of movement of the shoulder whereas 10 reported only painful scapular mass. All 16 patients underwent complete resections. The tumour size ranged from 3 to 15 cm. The mean hospital stay was 3.1 ± 1.4 days with a morbidity of 18.75% (seroma observed in 3 patients). The mean follow-up was 58.4 ± 29.5 months (range 11–92 months). In 2 patients (12.5%) a new occurrence on the contralateral side was observed at the follow-up.

CONCLUSIONS: Elastofibroma dorsi is a rare, ill-deﬁned, pseudotumoural lesion of the soft tissues. Surgical treatment can be proposed if the lesion is symptomatic. Furthermore, at the follow-up, the possibility of new occurrences on the contralateral side should be kept in mind.

Keywords: Chest wall • Benign tumour • Treatment • Outcomes

INTRODUCTION

Elastofibroma dorsi (ED) is a benign tumour of the thoracic wall, which is characterized by proliferation of the elastin component of the encapsulated fibrous tissue [1]. The lesion usually arises beneath the rhomboid major and latissimus dorsi muscles subjacent to the inferior angle of the scapula [2]. The pathogenesis of these lesions remains unclear. Some authors suggested that elastofibroma is a reactive process initiated by mechanical friction of the scapula to the chest wall and hyperproliferation of fibroelastic tissue caused by recurrent microtraumas [1]. Since its ﬁrst description, this neoplasm has received little attention in the modern literature and only a few studies have analysed the aetiology and the clinical behaviour of this tumour. In this study, we describe a series of 16 patients with ED and on the basis of our data we discuss the main features and the long-term outcomes of this entity.

PATIENTS AND METHODS

All ED patients who underwent surgery in Ankara Numune Teaching and Research Hospital between January 2003 and December 2012 were retrospectively reviewed. The clinical features including patients’ diagnostic methods, treatments, pathological characteristics of ED and the follow-up records of the outpatient clinic were evaluated. Ultrasound, computed tomography (CT) of the thorax and magnetic resonance imaging (MRI) were performed alone or in combination during the preoperative diagnostic work-up evaluation. This study protocol was approved by the Medical Ethics Committee of Ankara Numune Teaching and Research Hospital.

RESULTS

A total of 16 patients were treated within the study period. There were 11 females and 5 males (F/M ratio = 2.2). The mean age at presentation was 61.1 years (range 38–78 years). Six patients (37.5%) were manual labourers and the remaining 10 (62.5%) patients did not report any signiﬁcant information on heavy manual labour or sport activity. No genetic tendency that could be an aetiological factor was determined.

Elastofibroma dorsi appeared in the subscapular region in 13 (81.2%) patients and parascapular in 3 (18.7%). The tumour was
located on the right in 5 (31.2%) patients, on the left in 6 (37.5%) patients and bilaterally in 5 (31.2%).

In all patients, the major symptoms were pain and swelling but in 6 patients, painful scapular swelling resulted in a restriction of shoulder movement. The time from the onset of the pain to discovery of the lesions ranged between 3 and 42 months. First line radiology comprised ultrasound scan and thorax CT (Fig. 1). Magnetic resonance imaging was performed only in second instance to add information relevant to diagnostic clarification. The patients’ data are summarized in Table 1.

In all patients, complete excision was performed following frozen-section examination demonstrating benign soft tissue tumours. A complete surgical excision was done via muscle sparing technique, which requires preparation of latissimus dorsi and serratus anterior muscle flaps. The level of incision varied between the sixth and eighth ribs, depending on the size and location of the tumour. Tumours were usually fixed to the periostium of the ribs and tip of scapula. Our resection does not include resection of muscle and bone structures of the chest wall. As a result, after excision of the tumour the integrity and rigidity of the chest wall is protected. After complete excision of the encapsulated tumour, a suction drain was placed and muscle flaps were proximated with running sutures. The surgical wounds were closed and we used compression bandage. The drain was removed after 24–48 h. The patients with bilateral ED underwent complete excision of both sides during the same session (Fig. 2). The resected specimen ranged from 3 to 15 cm in diameter. A postoperative seroma developed in 3 patients (18.75%) requiring needle aspiration. Mean hospitalization was 3.1 ± 1.4 days (range 1–7 days).

In general, the lesions macroscopically appeared as irregular masses with indistinct borders and hard elastic consistence (Fig. 2). Microscopically, the tumours were composed of dense collagen fibre bundles and elastic fibres (Fig. 3).

The mean follow-up was 58.4 ± 29.5 months (range 11–92 months). At the end of the follow-up, all the patients were alive and well with no sign of recurrent disease. Surgery achieved excellent pain relief as well as a good range of shoulder movement. Interestingly, 2 patients (12.5%) developed a new ED on the opposite site of the operation. The time for the new occurrences after the operation was 13 months for the first patient and 19 months for the second one. The 2 patients were manual labourers. Re-evaluation of the initial thorax CT of the patients revealed no lesion on the other site. These 2 patients underwent surgery again and they are well at the follow-up.

DISCUSSION

Elastofibroma dorsi is a rare, benign tumour of the thoracic wall. It was first defined by Jarvi and Saxen [1] in 1961. Elastofibroma dorsi could be defined as a subscapular tumour clinically and it is also characterized by distinct elastosis histopathologically. Pathophysiological determinants are still not clear and several hypotheses have been put forward. Repeated microinjuries between the chest wall and the scapula, the source of excess elastin production and collagen degeneration could play a physiopathological role in this rare lesion [1, 3]. This view has been supported by the higher ED prevalence particularly among individuals who work at hard manual labours. However, patients who have never been involved in hard manual work, as well as those with elastofibromas in different locations, have undermined this view. In this study 6 patients (37.5%) were manual labourers. A new ED on the contralateral side developed in 2 of these patients at the follow-up. So this supports the hypothesis of microinjuries between the chest wall and the scapula in the pathogenesis of this rare proliferative lesion.

Elastofibroma dorsi is more commonly seen in older women, with a reported female: male ratio of 5/1 and a mean age at diagnosis of 65–70 years and there is one case report of a 6-year-old child with ED [4–6]. In this study, the female:male ratio was 2.2 and the mean age at presentation was 61.1 years (range 38–78 years).

Ninety-nine percent of EDs are located in the subscapular region and more commonly on the right; however, 10–66% of cases are bilateral [4, 7]. In our study, the tumour was located on the right in 5 (31.2%) patients, on the left in 6 (37.5%) patients and bilaterally in 5 (31.2%). Also ED appeared in the subscapular region in 13 (81.2%) patients and parascapular in 3 (18.7%).

In the physical examination, ED is in the form of well-defined lesions that do not adhere to the skin and can easily be palpated on the thoracic wall. The scapula may sometimes mask the lesion. When both arms of the patient are strongly pulled forward and the body is bent forward at an angle of 10–15°, the lesion becomes more evident. Pain is the most common clinical complaint; however, it is mild in almost all patients. There may be limitations in the motions of the upper extremities. While some authors [4, 8] have reported that in most patients, ED progresses asymptomatically, in this study, 6 patients had painful scapular swelling resulting in a movement restriction of the shoulder and 10 reported only painful scapular mass.

Elastofibroma dorsi tumours were initially considered a rare and slowly growing lesion; however, this lesion should be known to differentiate it from malignant tumour and to avoid wide or radical surgery [8, 9]. In this study, the strategy was to use frozen-section histological evaluation during the operation, and in the case of benign histology, to perform complete resection.

Figure 1: Thorax CT of a case with bilateral elastofibroma dorsi. CT scan demonstrates an 8 × 8 cm on the left and 9.5 × 7 cm on the right side regular soft-tissue masses (arrowheads) posterior to both scapulas. The masses have attenuation similar to that of the adjacent skeletal muscle.
On ultrasound ED usually appears as a hypoechoic mass with interspersed linear echogenicity resembling muscle [4, 10]. On computed tomography ED appears as a heterogeneous soft tissue mass with muscle-like density and, depending on the fat tissue, contains areas of low density [8, 11]. This appearance on CT is diagnostic for ED. On MRI, a typical feature of ED is that the interposed areas of decreased signal intensity also appear as low signal intensity on T2-weighted sequences [9, 11]. Computed tomography and MRI images are not pathognomic for ED, but the image of a lesion located in the subscapular region of a middle-aged or older individual is highly suggestive of ED [5, 12].

Some researchers believe that imaging features are not sufficient to diagnose ED and believe that biopsy is indicated to exclude more aggressive tumours [7]. But fine needle aspiration is

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**Table 1: Characteristics of the patients**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Heavy manual labour</th>
<th>Location</th>
<th>Size (cm)</th>
<th>Symptom</th>
<th>Complication</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
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<td>No</td>
<td>Right subcapular</td>
<td>3 × 2</td>
<td>Painful mass</td>
<td>No</td>
<td>No recurrence</td>
</tr>
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<td>F</td>
<td>No</td>
<td>Right subcapular</td>
<td>8 × 6</td>
<td>Painful mass</td>
<td>No</td>
<td>No recurrence</td>
</tr>
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<td>3</td>
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<td>Right subcapular</td>
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<td>Movement restriction</td>
<td>No recurrence</td>
</tr>
<tr>
<td>4</td>
<td>57</td>
<td>M</td>
<td>No</td>
<td>Left parascapular</td>
<td>5 × 3</td>
<td>Painful mass</td>
<td>Movement restriction</td>
<td>No recurrence</td>
</tr>
<tr>
<td>5</td>
<td>64</td>
<td>F</td>
<td>Yes</td>
<td>Bilateral subcapular</td>
<td>10 × 6</td>
<td>Painful mass</td>
<td>Movement restriction</td>
<td>No recurrence</td>
</tr>
<tr>
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<td>73</td>
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<td>7 × 6</td>
<td>Painful mass</td>
<td>No</td>
<td>No recurrence</td>
</tr>
<tr>
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<td>78</td>
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<td>Left subcapular</td>
<td>8 × 5</td>
<td>Painful mass</td>
<td>No</td>
<td>No recurrence</td>
</tr>
<tr>
<td>8</td>
<td>54</td>
<td>M</td>
<td>Yes</td>
<td>Bilateral subcapular</td>
<td>11 × 8</td>
<td>Painful mass</td>
<td>Movement restriction</td>
<td>No recurrence</td>
</tr>
<tr>
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<td>67</td>
<td>F</td>
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<td>Bilateral parascapular</td>
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<td>Painful mass</td>
<td>No</td>
<td>No recurrence</td>
</tr>
<tr>
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<td>Movement restriction</td>
<td>Seroma No recurrence</td>
</tr>
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<td>Movement restriction</td>
<td>New occurrence Contralateral side</td>
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<td>Painful mass</td>
<td>Movement restriction</td>
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</tr>
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<td>Painful mass</td>
<td>Movement restriction</td>
<td>Seroma No recurrence</td>
</tr>
<tr>
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<td>No</td>
<td>No recurrence</td>
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<td>62</td>
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<tr>
<td>16</td>
<td>78</td>
<td>F</td>
<td>No</td>
<td>Left subcapular</td>
<td>6 × 5</td>
<td>Painful mass</td>
<td>No</td>
<td>No recurrence</td>
</tr>
</tbody>
</table>

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**Figure 2:** Macroscopic view of excised material from a patient with bilateral elastofibroma dorsi. Regularly shaped, non-encapsulated dense masses.

**Figure 3:** Histopathological view of elastofibroma dorsi. Original magnification, ×100; haematoxylin-eosin stain reveals the fibrous bands to be composed of a mixture of collagen bundles and dense, eosinophilic elastic fibres (arrowheads). Mature adipose tissue is also seen.
not recommended due to the hypocellular structure of the tumour [13, 14]. Open biopsy or, at least, core needle biopsy is needed to obtain sufficient tissue samples [14]. In this study we did not perform fine needle aspiration biopsy or core needle biopsy because a complete tumour resection was proposed due to persisting symptoms and absence of malignancy evidences.

Complete excision is the recommended treatment for ED. Nevertheless, some studies advocate conservative treatment of patients whose diagnoses of ED are established radiologically and recommend surgical excision only when there are functional motion limitations, compression symptoms, pain or tumour size of 5 cm or larger [8, 15]. Surgical treatment is effective in elimination of ED-related complaints. In our study, all the patients had painful scapular swelling and in 7 of these patients the mass restricted movement of the shoulder. Surgery has achieved excellent pain relief as well as a good range of shoulder movement.

Post-surgical complications after the excision of ED include seroma, postoperative haematoma and wound infections [16, 17]. Seroma and haematoma were common in one series, which was attributed to insufficient immobilization [14]. In this study, we use postoperative wound drainage and compression bandage to reduce these complications. But even then a postoperative seroma developed in 3 patients requiring needle aspiration.

Pathologically, ED is in the form of a fibrous lesion of dirty white colour, is non-encapsulated and contains streaks of fat tissue. Some elastofibromas may have cystic degeneration. The histological appearance of the lesion is typical. In large areas, it contains hyalinized collagenous stroma and little amount of fat tissue in between. In hypocellular collagenous stroma, fibrils and globules that show eosinophilic staining are striking. In sections of haemotoxylen-eosin, the presence of fibrils and globules is important for determining the location of the lesion and its diagnosis [4, 10].

Nagamine et al. [7], however, reported 1 case with recurrence in their 170-patient series. Briccoli et al. [8] reported 1 patient with ED who had a second lesion on the other side 2 years after surgical resection. Marino et al. [18] reported 10% recurrence rate after a mean post-surgical period of 47 months. We followed up these patients 58.4 ± 29.5 months. We observed no recurrences on the operation site but interestingly 2 patients developed a new ED on the contralateral site of the operation and these 2 patients were manual labourers. Only a few studies [4-19] have reported on the outcomes of surgical treatment of ED (Table 2). In a series of 71 patients who underwent complete excision of ED, with similar outcomes, with a tumour size between 3 and 14 cm, complications of 16.7% and no recurrence at follow-up, have been reported [19].

In conclusion for symptomatic ED, surgery should be the choice of treatment because it is easy, has few complications, and there are no recurrences after complete excision. Furthermore, at follow-up, the possibility of new occurrences on the contralateral side should be kept in mind especially in manual labourer patients.

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**REFERENCES**


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**Table 2:** Outcomes of selected series of elastofibroma dorsi

<table>
<thead>
<tr>
<th>Published series, first author, year</th>
<th>n</th>
<th>F:M</th>
<th>Heavy manual labour (%)</th>
<th>Symptomathic patients (%)</th>
<th>Tumour size (cm)</th>
<th>Resection (%)</th>
<th>Complication (%)</th>
<th>Recurrence (%)</th>
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</thead>
<tbody>
<tr>
<td>Nagamine (1982) [7]</td>
<td>170</td>
<td>158:12</td>
<td>94.7</td>
<td>35.2</td>
<td>2–14</td>
<td>19.4</td>
<td>Unspecified</td>
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<tr>
<td>Mortman (2007) [4]</td>
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<td>3:3</td>
<td>Unspecified</td>
<td>100</td>
<td>Unspecified</td>
<td>100</td>
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<tr>
<td>Koksel (2008) [16]</td>
<td>8</td>
<td>7:1</td>
<td>Unspecified</td>
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<td>5–12</td>
<td>100</td>
<td>25</td>
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<tr>
<td>Nishio (2012) [17]</td>
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<td>5:6</td>
<td>Unspecified</td>
<td>27.3</td>
<td>5–9</td>
<td>45.5</td>
<td>20</td>
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<tr>
<td>Marino (2013) [18]</td>
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<td>35.7</td>
<td>50</td>
<td>3.5–15.7</td>
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<td>71</td>
<td>48:23</td>
<td>59.2</td>
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<td>16</td>
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<td>3–15</td>
<td>100</td>
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