1. Introduction

Primary tumors of the thoracic wall are a very heterogeneous group of neoplasms that develop in bones and soft tissue of the thoracic wall. They are infrequent, representing only 1–2% of all primary tumors. Within this group, the most common are the cartilaginous tumors (condroma and condrosarcoma) and then those derived from the soft tissue (fibromas, lipomas, neurogenic tumors) [1].

Elastofibromas (EF) are usually present in the dorsal subscapular region. They are benign neoplasms, characteristic of females and slow to develop. Their relationship with repetitive trauma, which can lead to a degenerative reaction of the elastic fibers, has been described [2]. When dealing with tumors of the thoracic wall, many diagnostic possibilities must be considered; thus, we present this series to illustrate the diagnosis and treatment of this rare tumor.

2. Patients and methods

This is a retrospective and descriptive study that looks at a series of eight cases admitted to our Thoracic Surgery Service with a diagnosis of EF. Pathological antecedents were evaluated as well as the clinical characteristics of the patients, possible factors related to the appearance of the neoplasm, diagnosis by image, cytology, histology, treatment, and short- and long-term outcome.

Diagnostic protocol and treatment followed was that which we normally carry out in patients with thoracic tumors: clinical history, physical examination with special attention to clinical signs of the tumor, basic analysis (routine serum biochemical investigations, coagulation and hemoanalysis), chest X-ray, computed tomography (CT) of the thorax and/or magnetic resonance imaging (MRI). In all cases an excisional biopsy of the lesion was done. In some cases where there were important doubts as to the diagnosis, a fine needle aspiration biopsy (FNAB) was done previously. In the case of bilateral tumors, once the diagnosis of EF was confirmed, surgical intervention of contralateral tumor was not done if there were no important clinical manifestations.

Surgical intervention was always performed by subscapular incision. A total excision of the tumor was carried out and perioperative biopsy done to confirm that it was benign, after which a conventional method was used to close the incision. Patients were discharged the day after the intervention. The follow-up was between six months and three years. If no relapse was detected the patient was discharged from the outpatient service after a period of no less than 2 years.

3. Results

Between 1998 and 2007 we attended eight patients whose histologic diagnosis was EF of the thoracic wall. Seven females and one male were treated between the ages of 44 and 62 years with an average age of 54.9 years (Table 1). In all cases the main sign was a subscapular tumor and five patients (62.5%) presented with pain. In four cases (50%) range of movement of the arm was limited. None of
Table 1
This table describes all cases included in the study

<table>
<thead>
<tr>
<th>Cases</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms/signs</th>
<th>CT</th>
<th>MRI</th>
<th>FNAB</th>
<th>Bilateral elastofibroma</th>
<th>Side resection</th>
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<td>1</td>
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<td>Male</td>
<td>Pain/functional limitation</td>
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<td>No</td>
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</tr>
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<td>Yes</td>
<td>No</td>
<td>Right</td>
</tr>
<tr>
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<td>Male</td>
<td>Pain</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes**</td>
<td>Left</td>
</tr>
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<td>4</td>
<td>59</td>
<td>Female</td>
<td>Pain</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes**</td>
<td>Left</td>
</tr>
<tr>
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<td>Female</td>
<td>Pain</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes**</td>
<td>Right</td>
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<tr>
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<td>No</td>
<td>Yes</td>
<td>Yes**</td>
<td>Right</td>
</tr>
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<td>8</td>
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<td>Yes</td>
<td>No</td>
<td>Yes*</td>
<td>Right</td>
</tr>
</tbody>
</table>

*Only radiological finding.
**Physical examination and radiological finding.

the patients had repetitive or previous traumas in the area of the tumor. In three cases (37.5%) another contralateral tumor was palpated in the same place. One patient had been operated on in another hospital on previous occasions with repeated bilateral relapses, presenting on this occasion with a right tumor.

An MRI was done on five patients (62.5%) and on another five CT was done, confirming in all cases the presence of a subscapular mass with intermediate density muscle-fat and compatible with a diagnosis of EF. In five cases (62.5%) a contralateral mass of similar characteristics was found (palpable in 3). On four occasions (50%) FNAB was done that was diagnosed EF in two and not conclusive, although a tumor was suspected in another two.

The definitive diagnosis, obtained by excision biopsy of the lesion, was always EF. The tumor was bilateral in six cases (75%) (clinically detected by radiology; n = 5 or by antecedents, n = 1). A resection was carried out on the right side on six occasions (75%) and on the left side in two, confirming the source of the tumor (Fig. 1). In no case was a bilateral resection done. None of the patients suffered postoperative complications and the clinical outcome was correct with no signs of recurrence in any of the cases. In the follow-up it was detected that in all cases there was an improvement in most of the symptoms presented previous to the intervention.

4. Discussion

EF is a rare tumor, described less than 50 years ago [3]. Most of the descriptions are of the thoracic region, and encompassing a large series of 170 patients studied [4].

It is much more frequent in women between the ages of 40 and 70 years and a bilateral affection is described in 10%, many times asynchronous [2]. A diagnosis is arrived at after the presence of symptoms, mainly pain or limitation of movement in the upper extremity [5–8]. These data were present in our series in which 87% were women and in 62.5% pain was the main symptom. Bilateral affection, however, was higher (75%) in our series. In our experience, surgical indication was established by the presence of symptoms in all cases and by the absence of a clear diagnosis in six of the eight patients. However, in those cases where the diagnosis of EF is clear with no important symptoms, it should be kept in mind that the transformation of EF to malignant is a fact that has not been described up to now [9].

In some previous works asymptomatic EF has been described, usually small tumors. About 50% of these cases were bilateral [10], a lower number than in our series. This may lead one to believe that in reality this tumor is much more frequent than described and that its benign character and few symptoms make it difficult to diagnose [11]. Its bilateral character, despite it being described before [12], has not been studied sufficiently up to the present.

The pathogenesis of EF is still not clear. Excessive movement or repetitive trauma of the area have been cited as possible inductors with a response in the form of degeneration of the elastic fibers [13]. None of the patients in our series presented this etiological factor. The theory of a neoplastic origin of the tumor is based on the existence of chromosomal changes discovered in some previous studies. It confirms its relation with other fibromatoses and including sarcomas [14].

From a radiological point of view, the typical image in the MRI presents high and low signal zones with cystic images in its interior [14] (Fig. 1). FNAB is the best method to correctly diagnose the tumor, although on occasions the sparse cellularity that it presents makes it difficult to interpret the cytology [15]. This occurred twice in our
series. FNAB as well as the characteristic radiographical image can give the wrong preoperative diagnosis [14]. The histology shows mature adiposities, fibroblasts and aggregations of petaloid globules with a matrix of collagen and signs of degeneration in the elastic fibers [7, 15].

Some authors have cited that surgical intervention with resection of the tumor is the standard treatment of EF. If the resection is incomplete, relapses occur in a described 7% [7] which presented in one of our patients who had previously undergone surgery in another hospital. Nevertheless, recent studies support a conservative treatment in the case of a diagnosis of EF by radiological image and FNAB [7]. The surgical indication can be reserved for cases with severe symptoms or when there is diagnostic doubt. In our experience, the EF excision has gone according to the clinical improvement in all the patients. We decided not to surgically treat any of the contralateral tumors we found and that did not present important symptoms.

We conclude that the rare aggression and slow growth of EF make clinical conservatism viable, reserving surgery for those cases in which the growth is rapid, if there is clinical doubt or if the patients experience severe symptoms.

References