Elastofibroma dorsi: management, outcome and review of the literature

Elastofibroma dorsi is an uncommon, benign, slow-growing soft-tissue tumour of uncertain aetiology. It classically presents as an ill-defined mass at the inferior pole of the scapula with symptoms which include swelling, discomfort, snapping, stiffness and occasionally pain.

We report the symptoms, function and outcome after treatment of 21 elastofibromas in 15 patients. All were diagnosed by MRI and early in the series four also underwent CT-guided biopsy to confirm the diagnosis. In all, 18 tumours were excised and three were observed. After excision, the mean visual analogue score for pain decreased from 4.6 (0 to 10) pre-operatively to 2.4 (0 to 8) post-operatively (p = 0.04). The mean shoulder function, at a mean follow-up of 4.2 years (3 months to 16 years), was 78.1% (30 to 100) using the Stanmore percentage of normal shoulder assessment scoring system. The mean range of forward flexion improved from 135° (70° to 180°) to 166° (100° to 180°) after excision (p = 0.005). In four patients a post-operative haematoma formed; one required evacuation. Three patients developed a post-operative seroma requiring needle aspiration and one developed a superficial infection which was treated with antibiotics.

Our findings support previous reports suggesting that a pre-operative tissue diagnosis is not necessary in most cases since the lesion can be confidently diagnosed by MRI, when interpreted in the light of appropriate clinical findings. Surgical excision in symptomatic patients, is helpful.

It has been suggested that elastofibroma is caused by a local tissue reaction and is not a true neoplastic process. A strong association has been noted between elastofibroma and repetitive use of the shoulder, which is supported by our findings.

Patients and Methods

Between 1996 and 2008, 15 patients (21 tumours) with a diagnosis of elastofibroma were identified from the unit’s database. Of the unilateral tumours, one lesion occurred on the left side and eight on the right. Bilateral lesions were found in six patients. The dominant arm was involved in 12 tumours. All the tumours were found at the inferior border of the scapula, mostly deep to serratus anterior. The male:female ratio was 8:7 and the mean age at presentation was 61 years (40 to 71). The symptoms and signs were variable at presentation, but included the presence of a mass, pain, stiffness, scapular snapping and impingement-like features.

There is little information on elastofibroma with most reports consisting of limited case reports and case series. We present a series of 21 tumours which were treated at our institution. The symptoms, function, causal relationships and pre- and post-operative range of movement were evaluated. We also describe the radiological and histological features and suggest optimum management.

Elastofibroma was first described by Jarvi and Saxen in 1961. It is a benign, slow-growing lesion classically presenting as an ill-defined mass at the inferior pole of the scapula, hence the term elastofibroma dorsi. The lesion is normally deep to serratus anterior and latissimus dorsi, and may be fixed to the periosteum of a rib. It is often bilateral.

There is a predilection for the lesion to occur in women over fifty years of age and it is extremely rare in children. The aetiology remains unclear and is a source of ongoing debate. Typically, symptoms may include the presence of a mass, pain, stiffness, scapular snapping and impingement-like features.

The Royal National Orthopaedic Hospital, Stanmore, Middlesex HA7 4LP, UK

Correspondence should be sent to Mr M. T. R. Parratt; e-mail: timothyparratt@doctors.net.uk


Received 10 June 2009; Accepted after revision 14 September 2009

From the Royal National Orthopaedic Hospital, Stanmore, England
In four lesions the diagnosis was established by combined MRI and CT-guided biopsy. The remainder were diagnosed by MRI alone. Excision was undertaken after appropriate patient counselling during which pre-operative consent was obtained, potential benefits and possible risks of surgery, and in those patients whose symptoms justified it, for example variables such as mass, pain and snapping were considered. A transverse or oblique incision at the inferior pole of the scapula was used. The aim of surgical management was marginal rather than wide excision and all operative samples were sent for histopathological evaluation.

Data regarding symptoms, occupation and family and employment history (including hobbies) were collected during outpatient attendances or from the clinical records. A visual analogue score (VAS) in which 0 represented no pain and 10 extremely severe pain was used to assess levels of pain, and function was assessed using the Stanmore percentage of normal shoulder assessment score. Four patients were also assessed pre- and post-operatively for the range of movement of the shoulder, particularly for forward flexion which was the movement deemed most likely to be affected by the tumour.

### Statistical analysis

Data were analysed using SPSS software (SPSS Inc., Chicago, Illinois). Mean values pre- and post-operatively were evaluated using the t-test. A p-value < 0.05 was considered statistically significant.

### Results

None of the patients had a family history of elastofibroma or related conditions. Nine were, or had been involved in manual labour or occupations involving heavy lifting. An additional two patients participated in tasks (hobbies) in which there was extensive use of the shoulder. Two patients had undergone a previous operation on the affected shoulder. One had arthroscopic repair of the rotator cuff and the other arthroscopic capsular release. One patient had sustained a previous gunshot wound to the affected shoulder.

All the patients had MRI using standard T1, T2, STIR and contrast-enhanced sequences. All the lesions had the characteristic appearances previously described in the literature, namely, a poorly circumscribed, unencapsulated mass with attenuation similar to that of skeletal muscle on both T1 and T2 sequences, containing linear streaks of tissue with the signal attenuation of fat (Fig. 1). The underlying bone was intact and there was no evidence of infiltration of the chest wall in any patient.

### Pathological findings

Macroscopically, all the excised tumours were firm, unencapsulated and had an indistinct border. The cut surfaces revealed tan-coloured fibrous tissue with yellow flecks distributed throughout. On review of the histopathological reports, marginal excision had been achieved in 11 tumours and an infiltrative margin (involving lesional tissue) was noted in the remaining seven. The mean length of all the excised tumours was 101 mm (50 to 190). The microscopic findings showed characteristic hypocellular lesions consisting of eosinophilic collagen and elastic fibres, which were branched and unbranched and had a central dense core with serrated margins. All the tumours contained areas of interspersed fat and there was no evidence of cellular atypia.

### Outcomes

In the operative group of 18 tumours, the mean VAS for pain was 4.6 (0 to 10) pre-operatively and 2.4 (0 to 8) (p = 0.04) post-operatively. The mean Stanmore score post-

<table>
<thead>
<tr>
<th>Case</th>
<th>Mass</th>
<th>Pain</th>
<th>Snapping</th>
<th>Catching</th>
<th>Locking</th>
<th>Stiffness</th>
<th>Employment history</th>
<th>Hobbies</th>
<th>Excision</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
<td>Plumber/builder</td>
<td>Sea fishing</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>2</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
<td>NA*</td>
<td>NA</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>3</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
<td>Plumber</td>
<td>Fishing</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>4</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Fireman</td>
<td>NA</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>5</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Y</td>
<td>Builder</td>
<td>NA</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>6</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>NA</td>
<td>NA</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>7</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>Nurse</td>
<td>Sailing</td>
<td>N</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Y</td>
<td>Gardener</td>
<td>Horse-riding</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>9</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Y</td>
<td>Musician</td>
<td>NA</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>10</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Nurse/foster parent</td>
<td>Gardening/</td>
<td>Y</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Badminton</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>Y</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>Gym instructor</td>
<td>NA</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>12</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>NA</td>
<td>NA</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>13</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Warehouse worker</td>
<td>Coarse fishing</td>
<td>N</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>General practitioner</td>
<td>Golf</td>
<td>Y</td>
<td>N</td>
</tr>
<tr>
<td>15</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>NA</td>
<td>NA</td>
<td>N</td>
<td></td>
</tr>
</tbody>
</table>

* NA, not applicable
operatively was 78.1% (30 to 100) at a mean of 4.2 years (3 months to 16 years). The mean forward flexion increased from 135° pre-operatively (70° to 180°) to 166° (100° to 180°) post-operatively (p = 0.005).

Complications included a local haematoma in four tumours which required surgical evacuation in one, a seroma in three tumours requiring needle aspiration in the outpatient clinic and one case of superficial wound infection treated with oral antibiotics. There was one local recurrence at eight years which was re-excised.

In the non-operative group of three tumours the mean VAS for pain at last follow-up at a mean of 5.75 years (3 to 8) was 2.7 (2 to 3). The mean forward flexion was 140° (90° to 170°) and the mean Stanmore score was 81.7% (65 to 100).

Discussion
Elastofibroma dorsi is an uncommon, benign connective-tissue tumour classically located at the inferior angle of the scapula. However, it can be found at other locations and has been described in the deltoid muscle, foot, greater trochanter, olecranon, greater omentum, cornea, stomach, ischial tuberosity, intraspinal space and the chest wall.8-16 In addition, one report describes a single patient with multiple elastofibromas who presented with 15 separate masses which included the classical infrascapular positions as well as in the buttocks and upper limbs.17

Elastofibromas are traditionally considered to be rare, but their exact prevalence is unknown. Incidental lesions have been found in 2% of adults over 60 years of age by CT.18 Jarvi and Lansimies19 showed, in a series of 235 post-mortems in patients older than 55 years, changes in the subscapular thoracic fascia similar to elastofibroma in 24.4% of women (29 of 119) and 11.2% in men (10 of 89). The discrepancy in the incidence is probably related to the great variability in the size of the tumour. Bilateral lesions are not uncommon and are thought to occur in up to 60% of patients.11 Our series was in keeping with this figure.

Elastofibroma may be misdiagnosed clinically as a tear of the rotator cuff or subacromial bursitis20 All the patients in our study had evidence of a mass plus at least one other symptom (Table I). The natural history of the lesion is variable, with combinations of symptoms reported. The indication for excision depends on the severity of symptoms and patient preference, with those patients with few symptoms possibly warranting simple observation. There has been no reported case of malignant transformation.

The pathogenesis of elastofibroma is unknown, but the histopathology suggests that the elastic fibres result from the degeneration of collagen fibres.11,21,22 Furthermore, in situ hybridisation and immunoelectron-microscopic studies have suggested that the elastic fibrillogenesis is an abnormal, rather than a degenerative process.23,24 A genetic predisposition has been proposed since the chromosomal regions Xq12-q22 and 19 have been identified as potentially containing genes involved in the development of some tumours.25 In the largest reported series of 170 patients with the lesion, a familial predisposition was suggested with 32% having a positive family history for elastofibroma.11 This concept is not in keeping with our findings. Others have suggested that the lesion is a reactive condition19,26 and does not represent a neoplastic process. Hypertrophy, secondary degeneration and necrosis of elastic tissue were found in a post-mortem study, potentially caused by direct mechanical stress.19 It was concluded that a failing resistance of the vascular system secondary to friction against the
scapula and stretching movements of the upper limb could have played a role. Another post-mortem study found pre-elastofibromatous changes on histology in 81% of the lesions studied. Elastofibroma has been reported to have a higher prevalence in manual labourers. It has been reported in the foot of an intensive dancer and in the pitching arm of a baseball pitcher, reinforcing the concept that the condition arises due to some degree of attrition. In addition, osteoarthritis of the shoulder has been cited as a potential cause. Yamamoto et al described a case of subscapular elastofibroma which occurred after ipsilateral osteoarthritis of the glenohumeral joint and suggested that increased scapulothoracic movement was a contributing factor to the formation of the lesion. In our series the high incidence in patients whose occupations or hobbies involved heavy lifting, manual labour or other intensive use of the shoulder reinforces the concept that a reactive process is involved.

Imaging modalities for diagnosis include Doppler ultrasonography, CT and MRI. In a retrospective evaluation in three cases of elastofibroma dorsi using ultrasonography, Bianchi et al concluded that the lesion could be accurately diagnosed if the mass was in the classical subscapular region and had the sonographic appearance of a well-defined multi-layered pattern. This concept has been reinforced by other studies, particularly when used to supplement CT and MRI. Battaglia et al studied 43 patients with proven elastofibroma using ultrasonography followed by CT and MRI. It was shown that, if a typical fasciculated pattern was evident on ultrasonography, elastofibroma could be diagnosed. This was considered to be pathognomonic if a history of a slow-growing lesion, deep to the periscapular region, was observed in women aged between 50 and 60 years. Furthermore, CT and MRI were advocated only when a fasciculated pattern was not observed, the lesion was in an atypical location or the patients were candidates for surgery. Nevertheless, in clinical practice, CT and MRI are more commonly used as a first-line investigation of the tumour. A poorly-defined, heterogenous soft-tissue mass, with tissue attenuation similar to that of skeletal muscle interlaced with strands of fat is seen. It has been reported that a diagnosis can be made on the radiological appearance alone. The radiological differential diagnoses are reported to include fibromatosis (in predominantly fibrous lesions) and atypical lipoma or liposarcoma in which lesions have a considerable fat content but in the typical subscapular location, especially if bilateral, accurate diagnosis can be obtained without the need for biopsy. Elastofibromas are often bilateral which argues against a malignant aetiology. Careful examination should be undertaken to detect a co-existing tumour. We prefer to use MRI to diagnose an elastofibroma. Although four of our earlier patients had a CT-guided biopsy, this is no longer our standard practice since current evidence suggests that the lesion can be confidently diagnosed by radiological investigation alone if typical clinical features are present. No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References