Elastofibroma at the Scapular Region

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ABSTRACT: **Background:** Elastofibroma is a rare type of lesion consisting of elastic fibers within a stroma of collagen and fatty tissue. It is usually located on the lower scapular region attached firmly to the thoracic cage, often causing debilitating pain. Its clinical presentation mimics a soft tissue tumor.

Objectives: To evaluate the diagnosis and treatment results of elastofibroma.

Methods: Clinical and radiographic evaluations were performed in 11 patients with thoracic wall mass. In five of them a biopsy was taken before surgery. All patients were operated and the diagnosis of elastofibroma was confirmed by histology.

Results: Two patients had a postoperative seroma that resolved spontaneously within a few days. All patients resumed their preoperative activities, including sports.

Conclusions: Considering the slow-growing nature of this tumor and its typical presentation, we believe that when this diagnosis is suspected, investigation does not necessitate staging (as in sarcomas). Also, marginal surgical excision is sufficient. Observation is an acceptable alternative to surgery. *IMAJ* 2009;11:170–172

KEY WORDS: elastofibroma, scapular region

Elastofibroma, first described by Järvi and Saxen in 1961 [1], is a rare type of slow-growing fibroma characterized histologically by elastic fibers within a stroma of collagen and fatty tissue. It is a pseudotumor, and by definition is benign and lacks a capsule. Clinically, the lesion is most often located at the scapular region (under the rhomboid and serratus anterior muscles), firmly attached to the rib cage. For this reason it was originally named elastofibroma dorsi.

Demographically, elastofibroma tends to affect women more often than men, with a female to male ratio of 8:1 [2]. Elastofibroma typically occurs in female patients over the age of 55 [3]. It presents bilaterally in 10% of cases [4]. Although the most common location is the infrascapular region, less common sites include the greater trochanter, olecranon, ischial tuberosity, deltoid muscle, foot, tricuspid valve, inguinal region, orbits, stomach, greater omentum, axilla, and the intraspinal space [2,3,5,6].

The demographic data presented characterize the great majority of cases. However, rare presentations of the disease appearing during childhood [7,8] or as a multiple subcutaneous (rather than single) lesion in the same patient [9] have been described. Traditionally, elastofibroma is considered a rare lesion, but autopsies have shown this lesion to exist, most probably subclinically, in up to 24% of women and 11% of men over the age of 55 [1].

PATIENTS AND METHODS

Between January 2003 and March 2007, 11 patients (9 females and 2 males) between age 52 and 71 (mean age 63) were surgically treated by the orthopedic oncology service at our institution [Table 1]. All patients were followed in the outpatient clinic for at least 1 year, except for the last patient who was followed for 6 months.

Patients were followed every 2 weeks for the first month and subsequently every 3 months. All the patients were functionally active, with full range of activities of daily living. All patients at presentation complained of a bothersome mass in the scapular area. Only one had a history of previous shoulder trauma. Clinical and radiological evaluation was performed in all patients, five of whom underwent preoperative histological evaluation as well (fine needle aspiration or core needle biopsy) [Figures 1 and 2].

All patients were operated under general anesthesia in a lateral decubitus position enabling free movement of the affected arm [Figure 1]. The tumors were approached directly through a longitudinal incision. Tumors were excised marginally.

Macroscopically, the tumors seen in surgery were ill-defined from their surroundings, strongly adherent to the outer chest wall and ribs. In two cases, detachment of the rhomboids was necessary to enable resection. Pathological examination confirmed the diagnosis of elastofibroma: macroscopic by the typical appearance of a mixed yellowish fatty mass with white collagen fibers in it and microscopic by the hematoxilin & eosin stain [Figure 3].

All patients were discharged on the day following surgery with the exception of one patient who had a seroma at the surgical wound site and who was discharged 4 days following surgery and removal of a drain. In the two patients in whom detachment and reattachment of muscles was necessary, strenuous activity of the arm was allowed gradually only after 6 weeks.

RESULTS

All patients were available for follow-up, which ranged from 6 to 72 months. Two patients had a postoperative seroma that resolved spontaneously within a few days. In two patients (# 1 and 3), the arm was held in a shoulder immobilizer for 3 weeks to allow solid reattachment of the rhomboid muscles. There

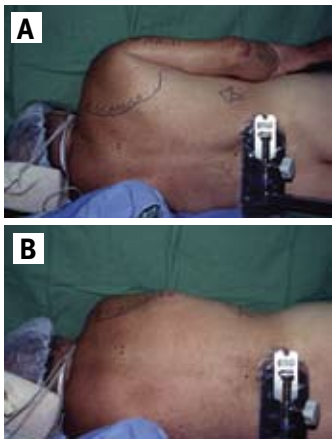

Figure 1. Clinical findings. **[A]** The mass disappears when the limb lies parallel to the body, but is visible **[B]** when the arm is adducted forward

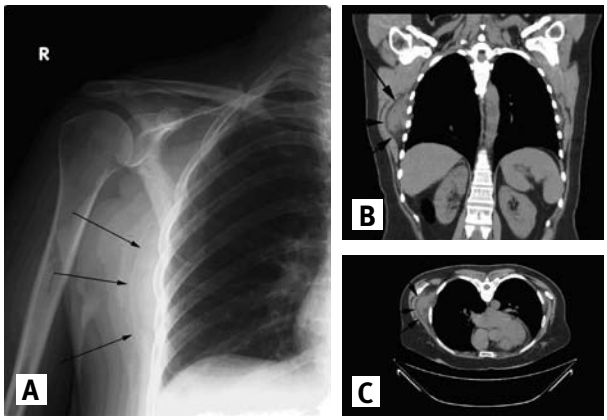

Figure 2. Radiographic findings. **[A]** X-ray, antero-posterior view. **[B]** CT findings, coronal view and **[C]** sagital view; black arrows indicate the mass

were no infections or postoperative "winging" of the scapula. All patients resumed their preoperative activities, including sports.

DISCUSSION

The scapula, a large, mostly flat membranous bone, is in close proximity to the proximal humerus, lung and ribs. It is a "favorable" location for various bone and soft tissue tumors. Ewing sarcoma, chondrosarcoma, as well as aneurysmal bone cysts and/or lipomas and myxomas are tumor types that may be found in this region.

Elastofibroma, a benign and relatively rare tumor, is included in the differential diagnosis with other soft tissue tumors, including sarcoma. The typical features of this entity will lead the clinician to suspect the diagnosis using the basic tools of history and physical examination. Careful preoperative imaging evaluation and, when necessary, FNA or CNB, will dictate the surgical procedure. This is even more important considering the otherwise poor prognostic features of the typical patient presenting with elastofibroma – namely, over age 55 years, proximal location in the limb, deeply seated location, and large diameter (> 5 cm) of the tumor. Proper preoperative diagnosis enables marginal resection of the tumor, in contrast to wide margins necessary for malignant tumor resection, which in this area is associated with devastating impairment to the shoulder girdle.

The etiology of the tumor is not fully understood. Three etiological theories are proposed as explanations for the pathogenesis of the condition. Historically, the lesion was first observed among heavy manual workers [1,2,]. The proposed explanation

FNA = fine-needle aspiration
CNB = core needle biopsy

Table 1. Patient and tumor data

					Presenting signs and symptoms			
Patient	Age (yrs)	Gender	Affected side	Previous trauma	Pain on movement	Click	Lump	Tumor size
1	52	F	R	+	+	+	+	5 x 3 x 2
2	57	F	R	-	+	-	+	6 x 4 x 3
3	63	F	R	-	-	+	+	7x 3 x 3
4	52	F	L	-	-	-	+	4 x 2 x 2
5	59	F	L	-	+	-	+	3 x 3 x 4
6	60	F	R	-	+	-	+	5 x 4 x 2
7	74	F	R	-	-	-	+	7.5 x 3 x 3
8	71	M	R	-	-	+	+	4.5 x 6 x 10.5
9	66	M	R	-	-	+	+	5 x 5 x 9
10	69	F	R	-	-	+	+	5 x 3 x 6
11	60	F	B	-	-	+	+	10 x 3 x 3.3

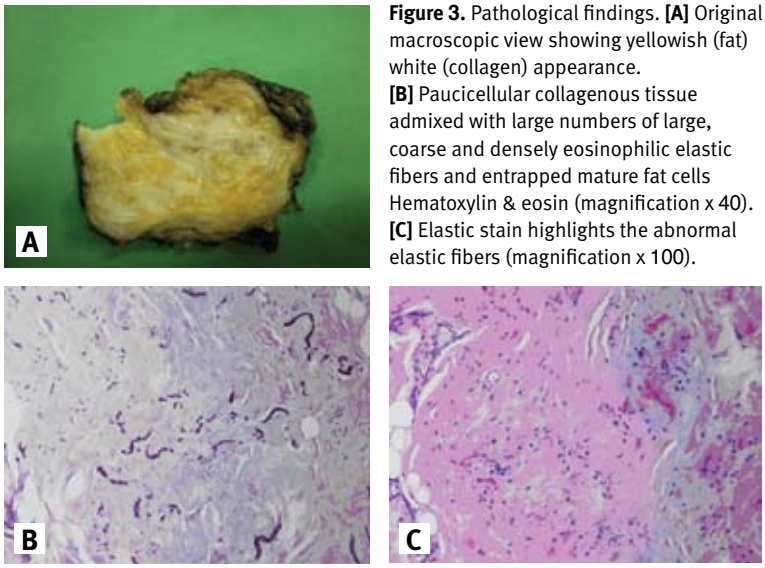

Figure 3. Pathological findings. **[A]** Original macroscopic view showing yellowish (fat) white (collagen) appearance. **[B]** Paucicellular collagenous tissue admixed with large numbers of large, coarse and densely eosinophilic elastic fibers and entrapped mature fat cells Hematoxylin & eosin (magnification x 40). **[C]** Elastic stain highlights the abnormal elastic fibers (magnification x 100).

was friction of the scapula against the thorax and an association between repetitive trauma and production of excessive amounts of elastic matrix by fibroblasts, which were observed in several studies [10,11]. Since elastofibroma as diagnosed today tends to affect women over the age of 55, another theory proposed reactive fibromatosis and degeneration secondary to vascular insufficiency and elastotic degeneration as the pathogenetic sequence [3,6]. Furthermore, a lack of association between trauma and tumor growth was reported by several authors [7,12].

The possibility of an underlying enzymatic defect was proposed by Fukuda et al. [13]. This observation is supported by the finding of an elastofibroma in the stomach of an individual with a subscapular elastofibroma and the few published cases of multiple familial occurrence [14-16].

Elastofibroma has a relatively consistent, typical clinical presentation. Its location is mostly in the proximal part of the extremity, with a tendency to be deeply seated, closely adherent to the underlying soft tissue. The size of the lesion is usually > 5 cm at presentation. The usual complaint is of discomfort (due to the size of the lesion) rather than pain. In the case of a sub-scapular location, which is the most common, the physician may palpate a mass, sometimes tender, under the scapula. Due to the location, the lesion may only be visible and palpable during adduction and forward flexion of the arm. A painful "click" as the scapula moves over the tumor is also not rare [17,18]. Therefore, care must be taken to perform palpation in a posture that exposes the tumor. At times, the lesion may be impalpable, since the tumor is often compressed between the scapula, the intrinsic and extrinsic muscles of the scapula, and the chest wall.

In general, once the diagnosis is suspected the clinician may choose one of two subsequent investigations: imaging of the lesion, or pathological sampling of the lesion in the office, in order to support the diagnosis. Various imaging modalities have been used in the evaluation of elastofibroma, including chest radiographs, ultrasound, computed tomography and magnetic resonance imaging. Plain films usually appear normal, but may occasionally show elevation of the scapula and a soft tissue mass without calcification in the sub-scapular and periscapular regions [Figure 1]. Bone erosion and invasion are rare, having been reported in only one case of an intra-articular elastofibroma.

Ultrasound images of elastofibroma have a typical appearance. Given the dynamic nature of this modality, the image will be more readily obtained by scanning the patients prone, with their arms abducted. The lesion appears well defined, with a multi-layered pattern and linear or curvilinear hypoechoic strands of fat in a background of echogenic fibroelastic material. Elastofibromas have characteristic imaging findings on MRI and CT that allow a definitive diagnosis in most cases [Figure 2]. On imaging, semilunar mass of soft tissue density is similar to the adjacent musculature and may contain linear areas of low density secondary to fat.

The series of patients presented here represent the typical course of elastofibroma in the scapular region. Clinical signs, as described, are typical: suggestive location, firm attachment to the surrounding soft tissue, and a "click" when the scapula moves over the tumor and exposes it to palpation. Proper histological evaluation and an attentive clinical evaluation will lead to the correct diagnosis and treatment.

We believe that history and physical examination, which are suggestive of elastofibroma, along with supporting radiological or histological evidence, are sufficient in order to excise the mass. The cumbersome task of staging necessary in the case of suspected sarcoma is avoided, along with the unnecessary expenses associated with costly imaging modalities. Furthermore, proper clinical diagnosis spares patients the fear of a malignant soft tissue tumor.

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