Elastofibroma of scapula: a case report and literature review

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Case Report

Abstract: Elastofibromas are benign soft tissue tumors. They occur mostly in the infrascapular region between the thoracic wall, the serratus anterior and the latissimus dorsi muscle, with a prevalence of up to 24% in the elderly. The etiology of this lesion remains uncertain and is a source of ongoing debate. We herein report a 54-year-old female patient, manual worker who presented with bilateral subscapular slow growing tumors for 7 months. Physical examination showed two masses of 4 cm in diameter on the right, and 8 cm on the left, both located inferior to the inferior margin of the scapula. They were more or less firm and mobile on palpation. Computed tomography scan showed large fusiform subscapular soft tissue heterogeneous solid masses with linear areas of low density secondary to fat. The lesions measured 5 cm × 4 cm on the right side and 8 cm × 8 cm on the left. Resection of the largest left tumor was achieved. Macroscopic and histological findings were consistent with elastofibroma.

Key Words: Elastofibroma—surgery—thoracic wall—pathology—subscapular mass

Introduction

Elastofibromas are slow-growing soft tissue benign tumor originating from mesenchymal tissue. They typically occur in the subscapular or infrascapular region at the inferior pole of the scapula and the serratus anterior muscle over the ribs of the thoracic cage (1-3). Although it is unilateral in most cases, bilateral elastofibromas may be more common than previously recognized (3).

Theses tumors are infrequent, representing 1-2% of all primary tumors of the chest wall (4). We report a new case of bilateral subscapular elastofibroma.

Case report

A 54-year-old female patient, manual worker, with no past medical history, presented with bilateral subscapular slow growing tumors for 7 months. The predominant symptoms were swelling, discomfort and occasionally pain and involved the left tumor.

Physical examination showed two masses of 4 cm of diameter on the right and 8 cm on the left, located inferior to the margo inferior of the scapula. They were more or less firm and mobile on palpation. Computed tomography scan showed large fusiform subscapular soft tissue heterogeneous solid masses with linear areas of low density secondary to fat. The lesions measured 5 cm × 4 cm on the right side and 8 cm × 8 cm on the left and showed well defined margins from surrounding muscle planes. The latter were free from any detectable abnormalities.

Resection of the symptomatic largest left tumor has been achieved.

Macroscopic findings showed a poorly defined fibroelastotic mass with a slightly rubbery, elastic consistence measuring 9 cm × 8 cm. The cut surface showed strands of grey-white and yellow tissue (Figure 1). Histologically, the tumor was poorly circumscribed, and was composed of hypocellular fibrous collagenous strands admixed with large numbers of coarse, densely eosinophilic elastic fibers and entrapped mature fat cells. The elastic

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structures sometimes formed discs or globules (Figure 2). These findings were consistent with elastofibroma.

The patient is well nine months after with no evidence of local recurrence.

**Discussion**

Elastofibromas were first described in 1959 by Jarvi and Saxen at the 12th Congress of Scandinavian Pathologists who subsequently published their work in 1961 (5). Since, less than 400 cases were reported in the literature (6-8).

The WHO soft tissue tumor classification 2002, defined elastofibroma as a benign fibroblast/myofibroblast tumor (3). However, most scholars do not accept it as a real tumor, but as hyperplasia, transformation and tumor like growth of elastic tissues, which are derived from fibroblasts due to chronic trauma or frequent mechanical friction (9).

The pathogenesis of elastofibroma is unclear. Three etiological theories have been proposed as explanations for the pathogenesis of the condition. In fact, the lesion was first observed among heavy manual workers. The proposed explanation was friction of the scapula against the thorax (10,11). This repetitive trauma causes overproduction of collagenous connective tissue, with degeneration of collagen fibers, associated with production of excessive amounts of elastic matrix alternating with deposition of hyperplastic fat. Moreover, 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.

Finally, familial predisposition with an underlying enzymatic defect has been proposed by Fukuda and al. as possible etiologic factors (1,6,10). Nagamine et al. have noted that 32% of their cases occurred within a single family, theses were detected in Okinawa Prefecture in Japan (6).

Elastofibroma is much more frequent in women between the ages of 40 and 70 years, and bilateral involvement occurs in 10% of cases, many times asynchronous (3,4).

These lesions tend to be slow growing and asymptomatic in more than 50% of the cases (1,3,10,12). When symptoms are present, they are typical, consisting of local scapular swelling and a clunking sensation during abduction and adduction of the shoulder, with pain of moderate or, rarely, severe degree (8,13). Subscapular elastofibromas are found more frequently on the right side (60%), but in 66% of cases, they are bilateral (5,6). The size of the lesion is usually >5 cm at presentation (10).

In general, once the diagnosis is suspected radiological investigation is performed. CT, MRI or sonographic images can demonstrate the characteristic streaky collagen or elastic fibers of elastofibroma dorsi in the fatty background (9). In fact, ultrasound examination shows typical images with an abnormal well defined mass of tissue in the typical location of the elastofibroma, with an alternating pattern of hyperechogenic and hypoechochogenic striae of different thickness similar to that of muscle tissue, roughly parallel to the chest wall (6,13,14). Elastofibromas have
characteristic imaging findings on MRI and CT that allow definitive diagnosis. In most cases CT often shows poorer differentiation of tumor edges from surrounding muscle planes compared with ultrasonography and MRI. It shows semilunar mass of soft tissue with density similar to the adjacent musculature containing linear areas of low density secondary to fat. CT also allows eliminating any other bone or adjacent soft tissue abnormalities (9). On MRI, probably the most reliable non-invasive technique in diagnosis, the lesions mostly show a signal intensity, comparable to that of muscle, margins are well defined and signal intensity is mostly low. Interspersed adipose strands cause heterogeneous structure with longitudinal areas of higher signal intensity (1).

Biopsy is unnecessary when the radiological findings are sufficiently typical (13). Sometimes, despite the clinical and radiological data, the diagnosis may be difficult. In this case, lesion biopsy can be performed allowing histological diagnosis. Fine needle aspiration (FNA) is a commonly used technique to definitively diagnose the condition. However, the collagenous nature of the mass often results in a paucicellular FNA smear, resulting in a false-negative report (15). Hayes et al. suggested a core biopsy only in case of suspicion of a soft tissue sarcoma (9,13).

Macroscopically, elastofibroma is usually ill defined and rubbery, and exhibits grey-white fibrous tissue with interposing small areas of yellow fat. The mass varies from 2 cm up to 15 cm in diameter (3).

Histologically, the diagnosis is composed of mixture of paucicellular collagenous tissue and large number of elastic fibers, associated with small amount of mucoid stroma and entrapped mature fat cells. The elastic fibers are often large, coarse, deeply eosinophilic and fragmented into small, linearly arranged globules or serrated disks simulating beads on the strings (1,3).

In incidental diagnosis of asymptomatic lesions, there is no need for excision, as malignant transformation has never been described. Only in cases of discomfort, snapping or blocking scapula and pain, marginal resection is widely recommended according to the psychological and physical strain of the patient (1,4).

Surgical excision of tumor has good therapeutic outcome, with extremely low recurrence rate after marginal excision (9,11).

The most frequent complication observed postoperatively is hematoma due to the fact that the periscapular region is highly vascular. The prognosis after surgical excision is good, with an extremely low recurrence rate (8,11,16,17).

Conclusions

Elastofibroma of scapula is a rare, ill-defined, pseudotumoral lesion of the soft tissues that most frequently affects older females. Its characteristic location and its specific aspect in imaging studies most often provides the diagnosis following an incidental discovery. Nevertheless, pathologic confirmation is necessary to formally rule out a malignant tumor diagnosis.

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References
