Elastofibroma Dorsi: A Case Report and Literature Review
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Introduction
Elastofibroma Dorsi (EFD) is a rare benign soft tissue tumor, characterized by a poorly defined solid mass, of non-capsulated fibroelastic, and fatty tissues. It’s usually located in the infrascapular region, of elderly women1,2. This anatomical location, and distinctive clinical symptoms, distinguishes them from other malignant soft tissue tumors3. We report herein a typical case of elastofibroma dorsi, and a brief review of the literature.

Case report
A 58-year old housewife was admitted to our surgical department, because of a painless lump and prominence of her right scapular for several months. No fever, weight loss or other symptoms. A part from hypertension, she had no significant medical history. Clinical examination revealed a mass which was rubbery in consistency, with regular margin, and adherent to the chest wall, but not to the skin. There were no other masses present anywhere else in her body.

The differential diagnosis included lipoma and primary soft tissue sarcoma. All routine haematological parameters and plain radiography were normal.

Fine needle aspiration cytology of the mass showed scanty connective cells, with no malignant cells.

At the surgical exploration of the right infrascapular region, a solid non- capsulated but well demarcated mass attached to the chest wall, was found and removed. The patient developed seroma collection postoperatively, which was aspirated twice.

Histopathological features
The specimen was a clearly demarcated, and partially encapsulated, firm mass measuring 10 cm in its greatest dimensions.

Sections showed a lesion consisting of mature fat, collagenous tissues, and foci of myxoid connective tissue, the cells of the lesion have regular nuclei without mitotic activity. There are numerous elastic fibers, and acidophilic globules in the lesion. Histological picture consistant with elastofibroma dorsi.

Discussion
Elastofibroma dorsi was first described by Jarvi in 1961, in autopsy examinations. Since then there has been sporadic case reports in the world literature4.

It occurs in patients over 50 year with a mean age of 60 years3. It is more commonly encountered in females5,6, the reason for this preponderance remains obscure6. It had never been recorded in young adult, teenagers or children1.

The presentation of our case is consistent with the typical location of the tumor at inferior pole of the scapula and the chest wall7. This has been related to natural occurrence of fibro elastic tissue in the region1,5,7,8.

Though not common, elastofibroma can occur in other parts of the body, including ischeal tuberosity, deltoid muscle, orbit, greater omentum2,3,5, oral mucosa5,7, breast, foot7, and stomach8,10.

The lesion was thought to be due to friction of the inferior angle of the scapula against the thoracic wall5,6 or by repeated minor trauma2. However, genetic predisposition or enzymatic defect has been high lighted as possible causes as well5,7.

This fibroelastic tissue usually function as shock absorber to cushion impacts transmitted from the upper limb though the glenohumeral joint to the shoulder girdele.

Typical to our case EFD is usually unilateral4. However, bilateral occurrence was described3 and in some reports.

It was seen in 10% of 2, 5,7,11. Generally elastofibroma is a slow growing tumour, without subjective symptoms, but depending on its size, it can cause periscapular pain, tension, reduced range of movement1,2, obvious swelling making the scapula prominent and causing clunk of scapula on adduction1,3,7,11, while in 50% it is asymtomatic7,11.

The differential diagnosis includes, frequently observed subcutaneous neoplasm as lipoma, fibrolipoma, or more frequently aggressive tumor as liposarcoma, fibrosarcoma, malignant, fibrous histocyteoma1,3, haemangioma or desmoid’s tumor2,3,4.
Only four single case reports of the cytological finding were reported in English language literature, and were suggestive when correlating the clinical and radiological findings. The diagnosis is only confirmed by histological examination of core biopsy.

On ultrasonography the mass had a striated appearance with multiple linear septae of alternating high and low signal intensity due to interlacing of linear streaks of fat, similar to morphology of striated muscle.

CT scan and MRI are not helpful in differentiating it from soft tissue sarcoma.

Surgical excision is the standard treatment, but should be reserved for patient with distressing or compressing symptoms, functional disability, asymmetrical counter or when the size is greater than five centimeters.

The procedure requires general anesthesia as the tumour has poorly defined margins and fixed to the muscle, periosteum of rib or scapula. Recurrence is unlikely. Only one case of recurrence was reported after surgical excision.

In conclusion, the tumour is very rare, the typical anatomical location, coupled with symptoms, strongly suggest the diagnosis, which can be confirmed by core biopsy, and surgery is reserved for patient with distressing symptoms and worry.

References

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