

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Surgery Quiz – Case 21

An otherwise-healthy 78-year-old retired farmer housewife presented to the emergency department complaining of swelling and snapping of the left scapula over the preceding four months. Physical examination revealed a firm, non-tender, relatively fixed and moderately increased in size during that period mass projecting prominently, on elevation of the left arm, below the tip of the left scapula at the level of the inferior angle (fig. 1). The patient had no history of trauma or malignancy. Chest computed tomography (CT) revealed the presence of a well-defined soft tissue mass with attenuation similar to that of the adjacent skeletal muscles located between the chest wall, the serratus anterior, and latissimus dorsi muscle (fig. 2). On magnetic resonance imaging (MRI), the lesion showed inhomogeneous structure due to the presence of fibroelastic and fatty components (hypo- and hyperintense, respectively, in T1- and T2-weighted images) (fig. 3).

What is your diagnosis?

- (a) Lipoma
- (b) Sarcoma
- (c) Elastofibroma
- (d) Peripheral nerve sheath tumor.

Comments

As primary chest wall tumors are rare, the resulting unfamiliarity



Figure 1



Figure 2

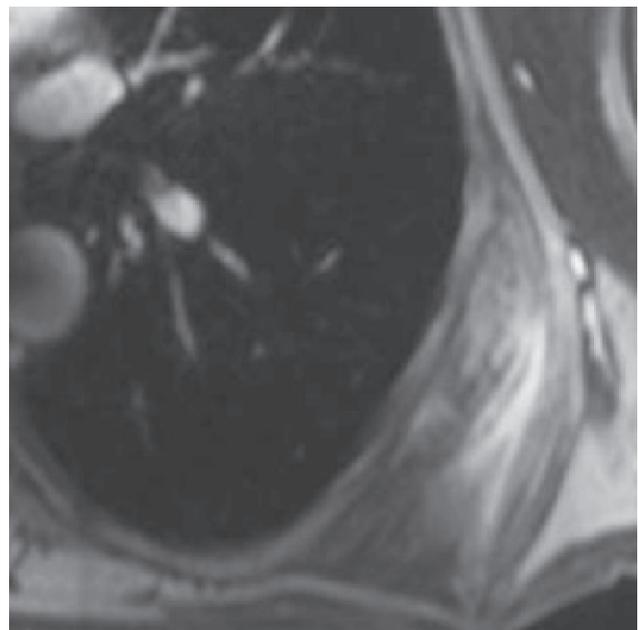


Figure 3

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2019, 36(5):715–716

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makes differential diagnosis difficult. Primary chest wall tumors may arise from bone structures or adjacent soft tissues. Soft tissue tumors can be divided into six categories with characteristic radiologic and pathologic features: (a) Adipocytic tumors (lipoma, liposarcoma); (b) vascular tumors (hemangioma, lymphangioma, angiosarcoma); (c) fibroblastic-myofibroblastic tumors (elastofibroma, fibromatosis); (d) fibrohistiocytic tumors (undifferentiated pleomorphic sarcoma); (e) peripheral nerve sheath tumors (schwannoma, neurofibroma, malignant peripheral nerve sheath tumors); and (f) cutaneous lesions (epidermal inclusion cyst, pilomatricoma, dermatofibrosarcoma protuberans).

Elastofibroma dorsi is a rare, benign, primary soft tissue chest wall tumor located at the inferior pole of the scapula deep to the serratus anterior muscle. On neutral arm position, the lesion appears as a projecting mass below the tip of the scapula at the level of the inferior angle. On arm elevation, the tumor can be easily palpated as firm, non-tender and relatively fixed mass. Patients may be asymptomatic or may present with swelling and snapping of the scapula associated with pain and restriction of arm movements. Elastofibroma usually presents in middle and old age patients, with an apparent peak in females beyond the 6th decade. Bilateral tumors occur in 10% of the patients, whereas multiple distinct tumors (as many as 17) in different locations have also been described in the literature. On chest MRI, elastofibroma presents as a well-defined soft tissue mass located between the chest wall, the serratus anterior and latissimus dorsi muscle. The tumor shows an inhomogeneous structure due to the presence of fibroelastic and fatty components which are depicted as hypo- and hyperintense, respectively, in T1- and T2-weighted images. Although excision is not necessary in asymptomatic patients as malignant transformation

has never been described, marginal resection is widely recommended in symptomatic patients.

In the present patient, the tumor had typical clinical and radiologic findings of elastofibroma dorsi. Final diagnosis was made by Tru-Cut biopsy; histology revealed that the tumor composed of fibrous, collagenous strands and plump and densely packed round-shaped elastic fibers; fibrocytic and fibroblastic cells were without atypia and mitotic activity, and the tumor was relatively hypocellular. The patient was referred for surgical excision to a tertiary thoracic surgery unit. In conclusion, diagnosis of elastofibroma in the present patient should have been straightforward as the tumor had typical clinical and imaging features; however, due to the rarity and clinicians' unfamiliarity of this entity, diagnosis required meticulous literature research, high clinical and imaging suspicion and biopsy confirmation before definitive differential diagnosis and management.

References

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