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Elastofibroma dorsi: three cases of personal experience

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SUMMARY: ***Elastofibroma dorsi:* three cases of personal experience.**

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Elastofibroma dorsi is a rare, slow growing, soft tissue lesion, which occurs mainly in elderly women, tipically located in the infrascapular region. This lesion is characterized by a poorly defined solid mass of fibroelastic and fatty tissue, enlarging into intermuscular spaces. Elastofibroma may simulate an aggressive behaviour, and differential diagnosis with malignant tumors is sometimes difficult. Surgical resection is generally recommended in symptomatic lesions or in the suspicion of malignancy.

We report our experience of three cases of elastofibroma, two of which typically located in the infrascapular region and one located in the supraclavear fossa, atypical site never reported so far.

RIASSUNTO: ***Elastofibroma dorsi:* tre casi dall'esperienza personale.**

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L'elastofibroma dorsi è un raro tumore dei tessuti molli, a lenta crescita, che si manifesta prevalentemente nel sesso femminile in età avanzata e tipicamente nella regione sottoscapolare. Tale lesione si presenta come una massa a margini non ben definiti, costituita da tessuto fibroelastico e adiposo, che si accresce negli spazi intermuscolari. L'elastofibroma può simulare una patologia tumorale maligna e spesso la diagnosi differenziale risulta difficoltosa. L'asportazione chirurgica è raccomandata nelle forme sintomatiche o nei casi in cui permanga il sospetto di neoplasia maligna.

Riportiamo la nostra esperienza di tre casi di elastofibroma dorsi, due dei quali tipicamente localizzati nella regione sottoscapolare ed uno nella fossa sovraclaveare, sete atipica mai riportata prima in letteratura.

KEY WORDS: *Elastofibroma dorsi* - Soft tissue tumor - Surgery.
Elastofibroma dorsi - Tumore dei tessuti molli - Chirurgia.

Introduction

Elastofibroma dorsi is a rare non-encapsulated benign soft tissue lesion (1). It is characteristically located in the distal infrascapular region, between the scapula and the thoracic wall. Even if rarely observed, the correct identification of this lesion is important since it may mimic other tumors of the thoracic wall that could require more aggressive treatment (2).

Although some authors report that ultrasound scan (US) and magnetic resonance imaging (MRI) can differentiate elastofibroma from the other soft tissue lesions (3, 4), others emphasize the importance of surgical resection and histological examination for a definite diagnosis (3, 5-8).

We report our experience on three cases of elastofibroma, two of which located in the typical infrascapular region and one located in the supraclavear fossa, atypical site never reported so far.

Clinical cases

Case 1

A 59 years-old woman was referred to our Division for a left-painful subscapular palpable mass dating several months. On physical examination, the lesion had a hard-elastic consistence, an irre-

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gular shape and it was not adherent to the overlying skin. The mass appeared more clearly when the shoulder was abducted forward. The clinical appearance resembled a lipoma.

The US was in agreement with the clinical evaluation describing an oval mass, partially covered by the scapula, of 2.5 cm in minor diameter compatible with a lipoma (Fig. 1). A MRI identified a non-capsular, circumscribed, heterogeneous soft tissue mass of 3 cm in diameter in the subscapular region, resembling the features of a fibrolipoma. Surgical resection was hence recommended; the lesion resulted as a fibroelastic mass, located beneath the latissimus dorsi muscle and firmly adherent to the chest wall. Macroscopic examination of the resected specimen showed a nodule of fibroadipose tissue of 8x7x2 cm (Fig. 2a). The final histopathological report diagnosed an *elastofibroma dorsi* (Fig. 2b).

The post-operative course was uneventful and no recurrence of disease has occurred at 12 months of follow-up.

Case 2

A 56 years-old man was referred to our Division for a painful, rapidly growing mass located in the right subscapular region. Clinical examination was indicative of a large lipoma.

An US demonstrated a well-defined, hyperechoic, oval-shaped mass of 5.5x4x2.5 cm, partially lying under the scapula.

The patient underwent surgery in view of a symptomatic lipoma. The lesion appeared as a bulky mass, extending under latissimus dorsi and tenaciously adherent to the surrounding tissue, mimicking a malignant soft tissue tumor. Macroscopic examination showed an egg-shaped lesion of 9x3x5 cm with fibrotic connective tissue and fatty cells at histopathology (Fig. 2c). Diagnosis of *elastofibroma dorsi* was made.

The post-operative course was uneventful and no recurrence has occurred at 18 months of follow-up.

Case 3

A 67 years-old woman was admitted at our Division presenting a palpable mass located in the right supraclavear region. She complained a 6-months history of a slowly growing mass associated to rising pain. Patient's clinical history included a lipomatosis of the dorsal region.

On physical examination, a solid-elastic mass not adherent to the overlying skin, clinically consistent with a lipoma, was appreciable.

Surgical excision was hence performed and the definitive histology described a 3x3x1 cm mass compatible with an elastofibroma (Fig. 2d).

The post-operative course was uneventful and no relapse has occurred after 28 months.

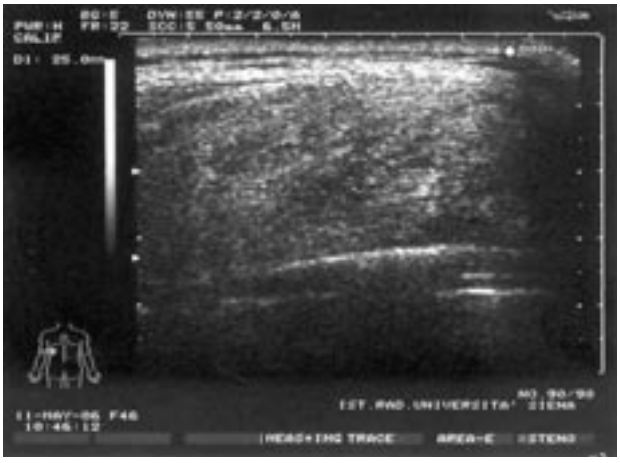

Fig. 1 - US: ovalar mass partially covered by the scapula.

Discussion

Elastofibroma dorsi, firstly described by Jarvi and Saxen in 1961 (1), is a benign, slow growing soft tissue lesion occurring most often in the infrascapular region (9). Although 99% of the cases are located between the distal part of the scapula and the thoracic wall (3), the lesion has been occasionally reported in other sites such as hand, foot, neck, mediastinum, stomach, greater omentum, cornea, orbit, ischial tuberosity, inguinal region, intraspinal and intra-articular spaces (5). Characteristically, it affects elderly women, usually aged more than 60 years, with a female to male ratio of 5-13 to 1 (3, 9-11). Observation of elastofibroma in young people is rare, although its occurrence has been sporadically described in children (4,11). Albeit not observed in our cases, bilateral location is a frequent event (10-66%) (3, 4, 12, 13).

The pathogenesis of this rare lesion remains undemonstrated, nevertheless it has been postulated the degeneration of connective tissue secondary to friction of the scapula against the rib cage caused by repetitive use of the shoulder girdle (3-5, 10, 11, 14). However, cases reported in literature frequently deny a history of intense physical activity or thoracic wall malformation. Moreover, the predominance of female gender and the report of uncommon localizations in a regions not submitted to mechanical stress contrast with this theory (14). Consistently, our three patients did not present a history of sport activity or intense physical work.

On gross appearance, *elastofibroma dorsi* is a non capsulated, solid mass, varying in diameter (1-13 cm). It is characteristically located at the angle of the scapula, deeply the rhomboid and latissimus dorsi muscles, adhering tenaciously to deeper planes but cleavable from the overlying muscle. In our third case the lesion was located in the right supraclavear fossa. It was not adherent to overlying skin or to other surrounding structures and it was easily enucleated under local anaesthesia. To our knowledge, this represents the first case located in this site and reported in literature.

Histological diagnosis is based on the finding of a connective stroma, constituted by poorly defined fibrolipomatous tissue, enclosing an abnormal elastic component represented by altered elastic fibers embedded in a collagenous matrix, riddled with various amounts of fat cells (2). Himmunohistochemically, it stains for vimentin but not for α-smooth muscle actin (SMA), S-100, desmin, or p53 (1).

97

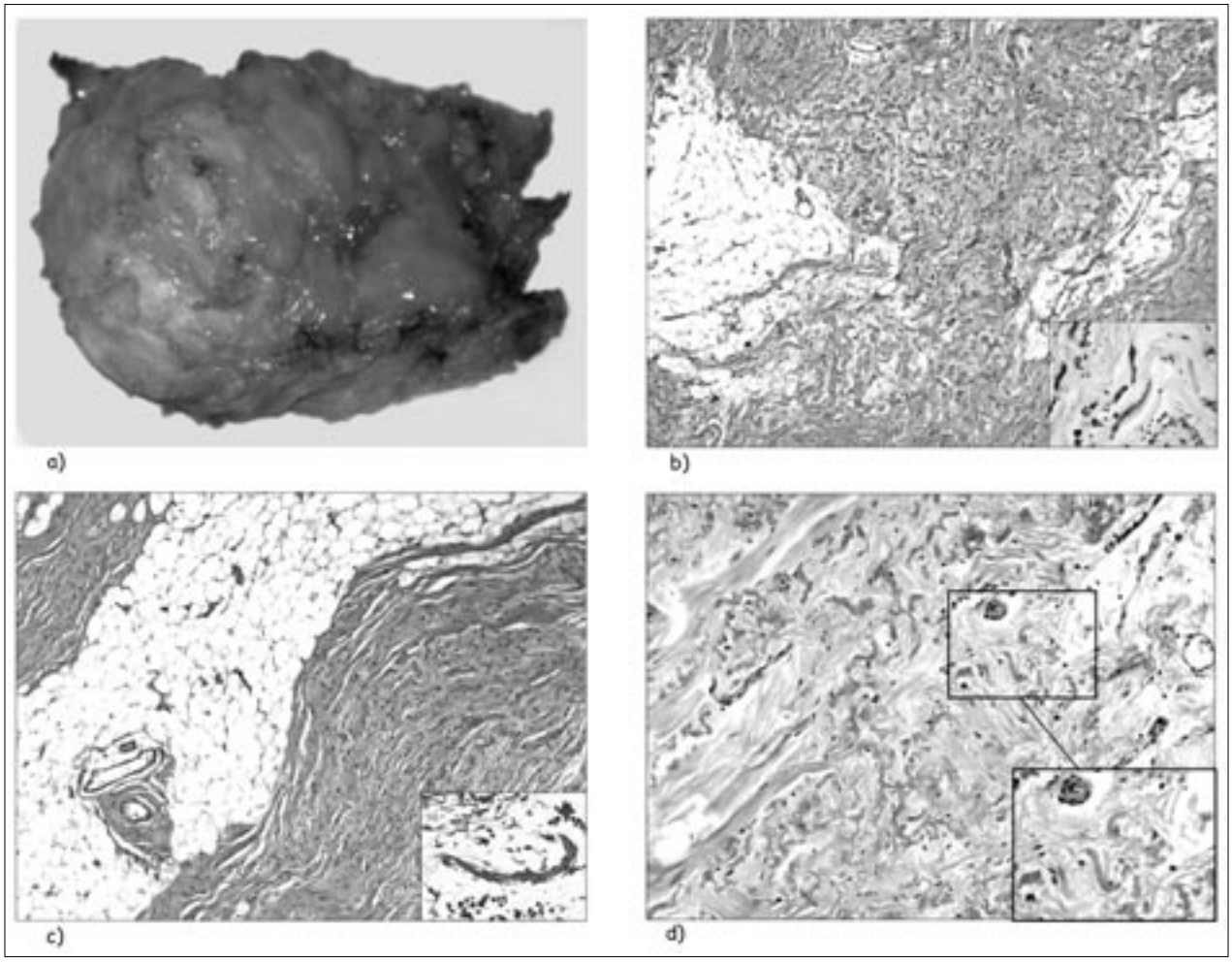

Fig. 2 - a) Specimen 8x7x2 cm. b), c), d) At low magnification, elastofibroma appears as an irregularly shaped lesion made of a paucicellular collagenous tissue containing numerous eosinophilic elastin bands admixed with mature fat. Elastic stains reveal the large fibers with a dense core and serrated or beaded margins [b: Hematoxylin-Eosin, original magnification (OM) 50x – Inset: Pinkus' Acid Orcein-Giemsa, OM 400x; c: Hematoxylin-Eosin, OM 50x – Inset: Weigert OM 200x; d: Hematoxylin-Eosin, OM 100x – Inset 400x].

Clinical examination usually reveals a slow growing, well circumscribed mass, non adherent to overlying skin (2, 6, 7, 10, 14, 15). The scapula may completely overlie the lesion thereby masking it until the patient abduct forward his arm. This allows the scapula to move up and let the mass to emerge. The differential diagnosis of such a soft tissue lesion includes lipoma, liposarcoma, sarcoma, fibromatosis, desmoid, neurofibroma, histiocytoma, hemangioma and metastasis.

In *elastofibroma dorsi*, imaging may give useful indications but it is often not conclusive. US shows characteristic findings evidencing a solid mass with regularly alternating fasciculated structures appearing as hyper- and hypoechoic striae (elastic and fat tissue, respectively). Overall, due to its muscle-like appearance, the ultrasound findings are generally insufficient to make diagnosis (2, 5, 14). CT confirms the similarity of the mass to the morphology of the muscular tissue and usually shows a heterogeneous soft tissue mass with poorly defined margins (7). MRI is the best non-invasive technique for diagnosis (3, 7, 14). At MRI, elastofibroma appears as a poorly circumscribed soft tissue mass with signal intensity similar to skeletal muscle, but with low signal intensity on T1-and T2-weighted images corresponding to the predominantly fibrous tissue of the lesion. Areas of increased signal intensity, when present, can correspond to adipose tissue (3, 4, 14). In our experience, US (cases 1 and 2) as well as MRI (case 1) missed both the diagnosis and the extension of the lesions. The error in defining the nature of the lesions is probably due to the rarity of elastofibroma, while the inaccuracy in defining their dimension is certainly related to the difficulty in delineating the margins from the surrounding muscle planes.

Regarding surgical resection, it is not mandatory in case of incidental diagnosis of asymptomatic lesions as malignant transformation has never been reported so far (3, 7, 8, 14, 16). Conversely, resection is recommended in symptomatic lesion and in the suspicion of malignant tumor. Practically, a definite diagnosis is often difficult prior to surgical excision both with imaging and fine needle aspiration cytology due to the scarceness of cellular elements (8, 11, 14). Regarding fine needle aspiration cytology, it must be emphasized that its use is no more recommended in the suspicion of soft tissue tumors (11, 14).

In conclusion, *elastofibroma dorsi* is a rare lesion of the chest which should be kept in mind in the differential diagnosis with benign or malignant lesions of the thoracic wall. The typical localization may suggest its diagnosis, but clinical and instrumental findings should be confirmed by histological examination to exclude a malignancy. Our cases confirm the difficulty in the diagnosis when a misleading instrumental diagnosis contrasted with an intraoperative aspect suspect for malignancy, which may result in an inappropriate aggressive surgical approach.

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