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 supraclavicular 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-
Elastofibroma dorsi: three cases of personal experience

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 supraclavicular 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). Immunohistochemically, it stains for vimentin but not for α-smooth muscle actin (SMA), S-100, desmin, or p53 (1).
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 abducts 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.

References