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CASE REPORT

Elastofibroma dorsi : A report on 6 cases

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Elastofibromas are slow growing fibroblastic proliferations, which occur mainly in elderly women. They are typically located in the right subscapular region and are usually asymptomatic.
The characteristic findings in ultrasonography, computed tomography and magnetic resonance imaging usually allow to make the diagnosis. We report six cases of elastofibroma dorsi, the clinical presentation and radiology findings are reviewed and compared with literature.

Keywords : elastofibroma ; soft tissue ; pseudotumour.

INTRODUCTION

Elastofibroma dorsi (ED) is a rare benign soft tissue pseudotumoral lesion, characterised by the proliferation of fibrous tissue with elastin in the infrascapular region ; it mostly affects elderly women. Recognition of the lesion is important as the differential diagnosis includes other benign and also malignant tumours.

Although ED is an uncommon lesion, the site and imaging findings are characteristic (*14*). We report six cases of ED and discuss the radiological aspects of the lesions.

MATERIELS AND METHODS

The study included six patients, 4 females and 2 males, aged 40 to 60 years, who were diagnosed as presenting an elastofibroma dorsi. All patients presented with soft tissue masses in the dorsal region and functional disturbance in two cases. On physical examination a non tender, firm mass was palpated. All patients were explored with sonography, five of them with computed tomography (CT) and three with Magnetic Resonance Imaging (MRI). Radiographs were performed in two patients.

RESULTS

In all cases, ultrasound examination showed an oval soft tissue mass. The echogenicity of these masses was slightly heterogeneous, with hyperechoic linear streaks along the great axis of the mass. Radiographs were interpreted as normal in one case and showed a soft tissue density in the

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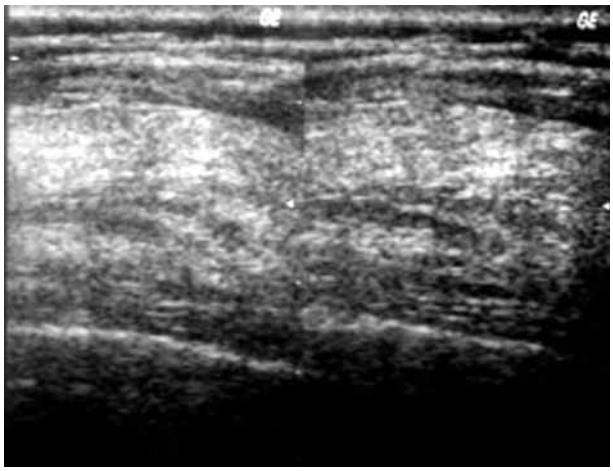

Fig. 1. — Ultrasound examination of the right scapular area shows an oval soft tissue mass located behind the ribs. Its echogenicity is slightly heterogeneous, with hyperechoic streaks along the great axis of the mass.

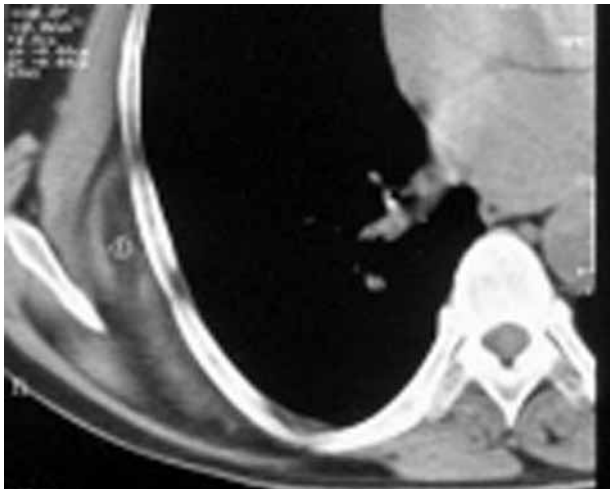

Fig. 2. — Computed tomography (CT) scan demonstrated a right subscapular ill-defined, non heterogeneous soft tissue mass bordered by the serratus anterior dorsi and ribs latissimus.

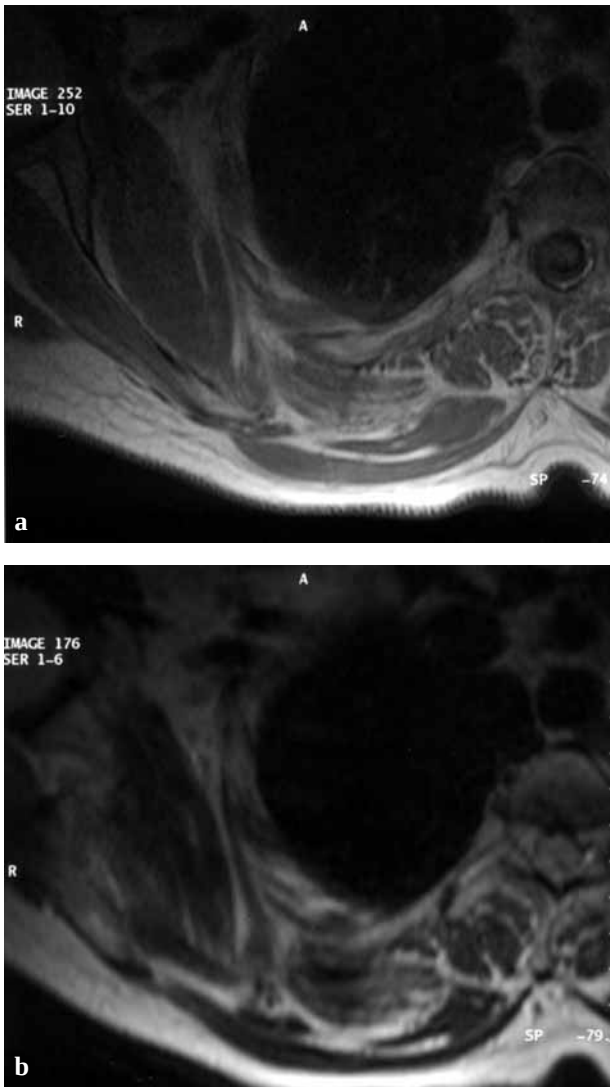

Fig. 3. — T1-weighted and T2-weighted images of the thorax showing a right subscapular mass. The lesion shows typical MR signal intensity characteristic of elastofibroma = mass with signal intensity similar to that of adjacent skeletal muscle, with interspersed linear areas of high signal intensity.

periscapular region in the other case. CT revealed a mass with attenuation values similar to those of the skeletal muscle and linear hypodense areas interspersed, suggesting fatty streaks within the mass. On MRI examination, the signal intensity was heterogeneous with the majority of the mass being intermediate (similar to muscle) on T1-weighted images with linear strands of highly hyperintense signal similar to subcutaneous fat. After injection of gadolinium (Gd-DTPA), no significant enhancement of the signal intensity was detected. Lesions were localised in the right subscapular region in four cases, with involvement of the contralateral side in two cases. In the two remaining cases, the lesion was located in the left subscapular region in one case and on the right side of the neck in the

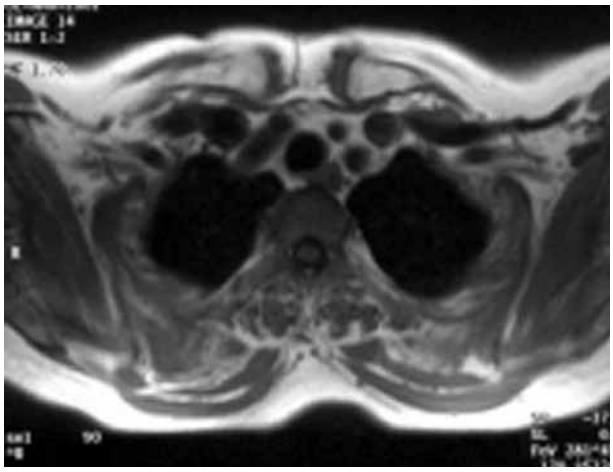

Fig. 4. — Axial MRI : T1-weighted image of the same patient demonstrating bilateral involvement of the subscapular soft tissue mass.

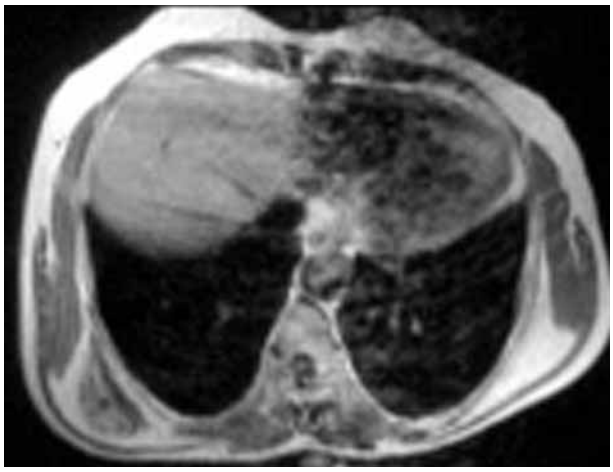

Fig. 5. — Axial MRI : T1-weighted image demonstrates the presence of different tissue components within the lesion : alternated plan of fat-like and fibrous-like tissue giving the lesion a multi layered pattern.

other. The diameter of the lesion varied between 4 and 7 cm. Surgical excision was performed in two cases and a biopsy of the lesion in one case. Histopathologic diagnosis was elastofibroma, confirmed with special staining techniques. For the other three cases, the aspect and the topography were considered typical of ED.

DISCUSSION

Elastofibroma dorsi (ED) first described by Jarvi and Saxen (*6*) in 1961 is a benign, slowly growing mesenchymal soft tissue lesion. It usually occurs in active individuals above the age of 50. Women are predominantly affected, with a ratio as high as 13/1 (*7*). There is a right side predominance but bilaterality has been reported in up to 66% of the cases (*5, 8*). Infraolecranon lesions concomitant with scapular lesion are common and were reported to be present in 16% of patients in a study of 170 ED's (*11*). Rare instances of this lesion have been reported in the regions of the greater trochanter, deltoid muscle, ischial tuberosity, breast, lateral chest wall and foot (*7*). Brander *et al* (*3*) reported that the prevalence of ED revealed by computed tomography was 2% in an asymptomatic elderly population. In autopsy, small subclinical elastofibroma was found in 24% of females and 11% of males over 55 years of age (*14*).

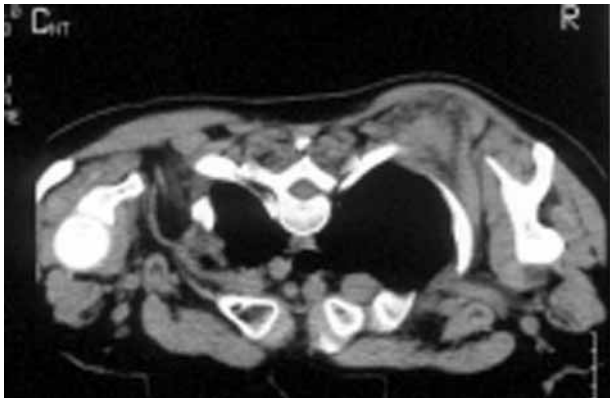

Fig. 6. — An enhanced axial CT in prone position showing an encapsulated soft tissue density largely surrounded by fat. The mass is bordered posteriorly by the trapezius muscle, anterolaterally by the serratus anterior muscle and medially by the ribs.

The cause and pathogenesis of ED are unclear, but it is suspected that mechanical friction between the chest wall and the tip of the scapula may lead to reactive hyperplasia of elastic fibres with consequently increased production of fibrous tissue (*9, 13*). However, the detection of new cases in different sites strengthened the hypothesis of an underlying

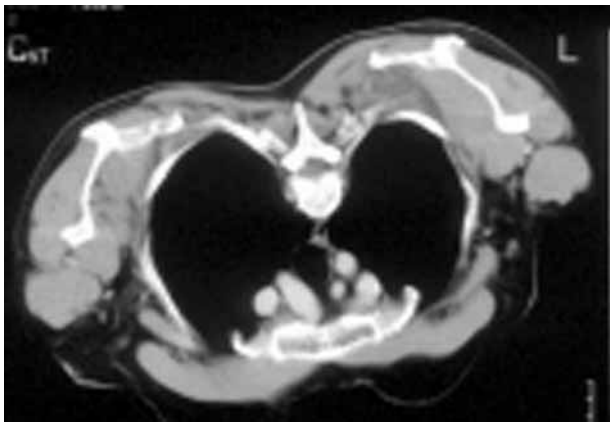

Fig. 7. — CT scan after contrast injection demonstrates a left subscapular ill-defined soft tissue mass with a predominance of fat within the lesion. No contrast enhancement was detected.

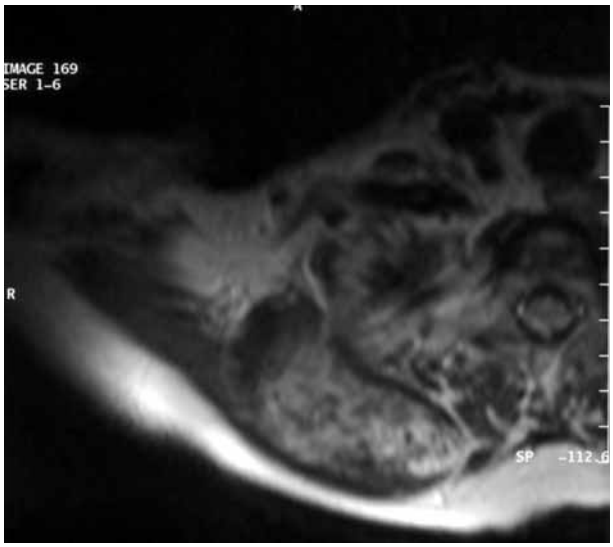

Fig. 8. — Axial T1-weighted magnetic resonance (MRI) image of the neck showing a soft tissue mass anterior to the trapezius muscle. This mass shows a signal intensity pattern similar to that of the subscapular lesions.

genetic disposition, inherent enzymatic defect or systemic involvement (*7*). Vanni *et al* (*16*) recently reported the cytogenetic findings in three elastofibromas, surgically excised from two patients, in which they described a picture of "chromosome instability".

The tumour remains asymptomatic in more than 50% of cases. Twenty five percent of patients may report simple discomfort, sometimes with a "clicking" or "catching" sensation associated with mobilisation of the arm. Pain is present in less than 10% of cases. Physical examination may reveal a rubbery asymptomatic mass located in the scapular region and barely noticeable when the arm lies against the chest (*4*). The size of the mass varies from a few cm to 15-20 cm and can remain stable or grow slowly (*14*).

Imaging examinations are usually performed in patients with chest wall masses to establish a diagnosis and to evaluate their anatomic extension prior to surgery. Chest radiographs may be normal or may show a soft tissue mass overlying the ribs, elevating the lower end of the scapula (*2, 13*). Ultrasonography (US) has become the most frequently used imaging technique for soft tissue masses. This evolution is mainly due to the improved resolution of present day ultrasound equipment and to increased experience of the examiners. The diagnostic value of ultrasonography for soft tissue masses and its correlation with the results of more invasive and costly imaging techniques (CT, MRI) have been established by many studies.

US is useful in delineating the anatomy and location of ED ; colour flow Doppler imaging provides information about blood flow. The sonographic appearance of ED consists of arrays of interspersed linear or curvilinear hypoechoic strands against anechogenic background (*1*). In our sixth patient, the US pattern was highly suggestive of ED, with a mobile lesion with streaky echostructure.

Computed tomography usually shows a non encapsulated heterogeneous soft tissue mass with poorly defined margin and a lenticular form with its long axis in craniocaudal orientation. Elastofibroma appears as a heterogeneous soft tissue attenuation mostly similar to the skeletal muscles, with linear interlaced low density streaks suggesting mature fat (*2, 14*).

The diagnosis of the contralateral tumour is often missed, because CT lacks sufficient contrast resolution to differentiate streaks of abnormal ED unless the latter reaches a sufficient size to be detected (*7*).

MRI is the best non invasive technique and the most useful for diagnosis. Elastofibromas appear as poorly circumscribed semilunar soft tissue lesions without discriminating the border between lesion and normal thoracic muscle bundles. The fibrous and fatty composition is usually well reflected on T1- and T2-weighted images. A predominance of areas of intermediate signal on T1-weighted images and relatively low-signal intensity (slightly higher than muscles) on T2-weighted images due to dense fibrous connective tissue can be seen mixed with linear strands which have a signal intensity similar to that of subcutaneous fat, due to the adipose interspersed elements (*1, 4, 14, 17*). Yu *et al* (*10*) speculated that T1 and T2-weighted spin echo MRI showing periscapular soft tissue neoplasm demonstrating a pattern of linear alternating regions of high and intermediate signal intensity might be sufficient to allow the diagnosis of elastofibroma (*17*). The MRI signal intensity pattern of the subscapular lesions in our cases was typical for elastofibroma and classical by location. Moreover in our third case, MRI was more sensitive in revealing the bilateral involvement of the tumour. The cervical lesion also showed a similar signal intensity pattern.

However, atypical ED can be seen with a homogeneous soft tissue appearance due to lesions with less amount of fatty tissue. Usually faint enhancement is seen with elastofibromas, although marked enhancement, mimicking malignant tumour, has been occasionally reported (*13*). In our cases, ED showed no contrast enhancement after Gd-DTPA injection.

Although these lesions can usually be diagnosed on the basis of their imaging characteristics, differential diagnosis with other soft tissue masses should be established. The peculiar feature of ED on MRI imaging is the relative hypo intensity on T2-weighted images. Thus, the differential diagnosis should be established with soft tissue lesions containing haemosiderin, such as giant cell tumours of tendon sheaths or with relatively acellular masses with large amounts of collagen such as musculoaponevrotic fibromatosis or desmoids tumours. A definitive diagnosis requires a biopsy showing the distinctive feature of elastofibromas : elastic fibers in a collagenised fibrous tissue with entrapped adipose tissue. Fine-needle aspiration represents the simplest and a quickest method of obtaining a definitive diagnosis in order to avoid an unnecessary radical and wide excision (*10, 12*).

In cases where the patient is asymptomatic, excision is unnecessary. Malignant transformation is unknown. Surgical excision is recommended when ED causes functional disability, compression symptoms, pain, disfiguring, swelling in the chest wall or when the tumour dimensions exceed 5 cm in diameter (*7, 8, 15*). Marginal excision carries a very low risk of recurrence and only one case with local recurrence has been reported in the literature (*11*). In our series, surgical excision was performed in only two cases. We have not observed any local recurrence during two years of follow-up.

CONCLUSION

Although elastofibroma is uncommon, it should be considered in the differential diagnosis of a soft tissue mass in the subscapular region. Its typical appearance on ultrasonography, CT and MRI as a soft tissue mass similar to adjacent muscles, interspersed with linear strands of fat in a characteristic location usually allows to make the correct diagnosis, avoiding an unduly aggressive procedure. No treatment is necessary for this degenerative pseudotumour in asymptomatic patients.

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