

## Diagnostic features of Elastofibroma dorsi: about 4 cases

### Caractéristiques diagnostiques de l'Elastofibrome dorsal : 4 cas

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#### RÉSUMÉ

L'élastofibrome dorsal est une tumeur bénigne rare des tissus mous, lentement évolutive, siégeant préférentiellement au niveau de la région sous scapulaire, survenant chez le sujet âgé actif. Sa localisation profonde rend une éventuelle biopsie chirurgicale délicate. Le but de ce travail est de décrire les signes radiologiques permettant de confirmer

Le diagnostic. Les auteurs rapportent 4 cas d'élastofibrome dorsal dont le diagnostic a été évoqué sur des éléments cliniques et échographiques et confirmé par le scanner et l'imagerie à résonance magnétique, pouvant ainsi épargner le recours à la biopsie chirurgicale. Aucune biopsie n'a été effectuée et le contrôle à l'imagerie n'a pas montré d'évolutivité tumorale ainsi qu'une disparition des symptômes sous traitement antalgique.

#### Mots-clés

Tumeur, thorax, diagnostic, échographie, IRM, élastofibrome

#### SUMMARY

Elastofibromas are rare benign soft tissue tumors that are commonly located at the inferior pole of the scapula, typically in the right side, deep to the serratus anterior muscle. They occur mainly in elderly women and are usually asymptomatic. We present four cases of elastofibroma dorsi in which the clinical and ultrasound diagnosis was confirmed by CT and magnetic resonance imaging. The location of the lesions (bilateral in two cases) was typical, in the thoraco-scapular region. When the lesion show typical imaging features and produces no symptoms, as is usually the case; further investigations are probably unnecessary.

#### Key-words

Tumor, chest, diagnosis, echography, MRI, elastofibroma

Elastofibromadorsi is a slow growing soft-tissue tumor characterized by fibroblastic proliferation and accumulation of abnormal elastic fibers deep to the inferior angle of the scapula. The pathogenesis of these lesions remains unknown, however some authors described a relationship with repetitive trauma, which can lead to a degenerative reaction of the elastic fibers. This lesion is usually seen in female patients over the age of 50 years and commonly painless. Physicians should be aware of the clinical manifestations, and that diagnosis can be established through imaging findings (MRI).

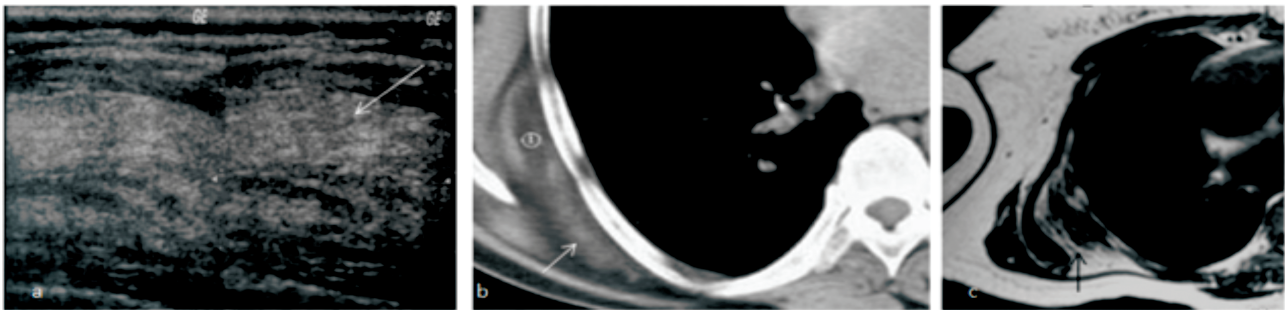
### METHODS

This study is a clinical assessment of four patients with a diagnosis of elastofibroma. Two cases were bilateral. The site of occurrence was in the typical infrascapular region in our series. There were two males and two females. The mean age at diagnosis was 52.2 years (range 43–60 years). Mass, discomfort and occasionally pain were the presenting symptoms. Average duration of symptoms was 20months (range 6-35months). Physical examination

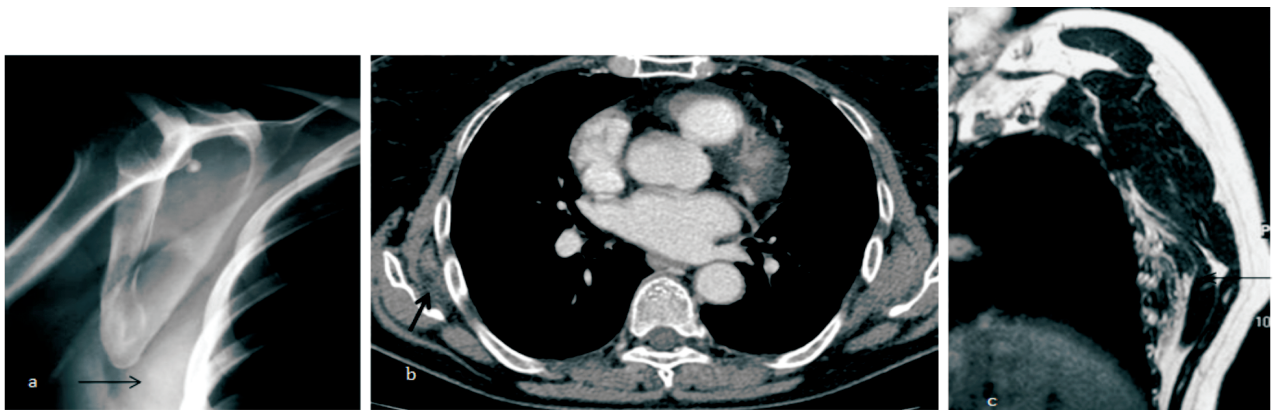
showed painful mass of the upper back with no inflammatory sign and a loss of range of motion of the right shoulder in two cases and swelling was not tender on palpation in the other cases. All the patients had their diagnosis based on clinical and imaging findings (US, CT, MRI). All patients were explored by echography, 3 of them by computed tomography (CT) and 4 by Magnetic Resonance Imaging (MRI). X-ray radiograph was performed for one patient.

### RESULTS

Ultrasound examination showed an oval soft tissue mass only in two cases (Figure 1). The echogenicity of these masses was slightly heterogeneous, with hyperechoics linear streaks along the great axis of the mass. The X-ray radiography (Figure 2) showed soft tissue opacity in the periscapular region. CT revealed four masses with attenuation values similar to those of the skeletal muscle and lineal hypodense areas interspersed, suggesting fatty streaks within the mass (Figure 1,2). On MRI examination, the signal intensity was heterogeneous with

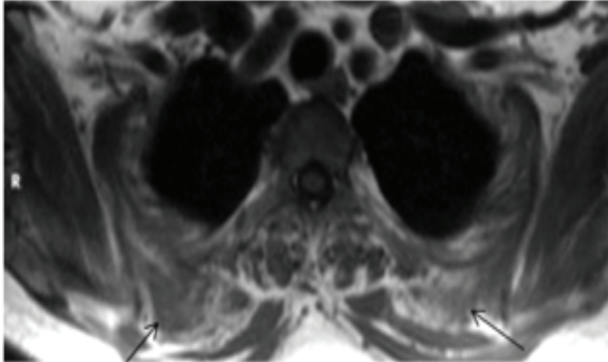


**Figure 1:** a : ultrasound examination of the right scapular area shows the oval soft tissue mass located behind the ribs. Its echogenicity is slightly heterogeneous, with hyperechoics streaks along the great axis of the mass. b: Axial unenhanced computed tomographic (CT) scan demonstrated a right sub scapular ill-defined, non heterogeneous soft tissue mass bordered by serratus anterior dorsi and ribs latissimus c : Axial MRI: unenhanced T1-weighted image demonstrates the presence of different tissue components within the lesion: alternated plan of fat-like and fibrous-like tissue getting the lesion a multi layered pattern.

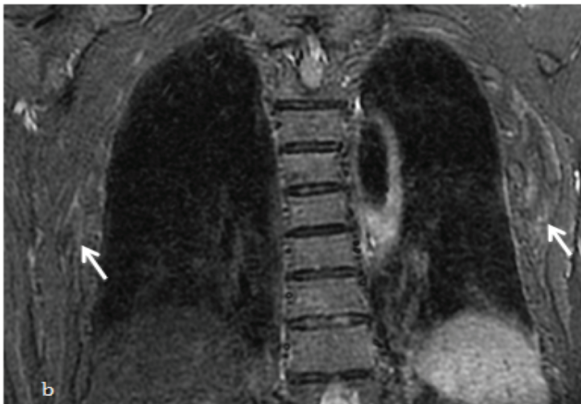
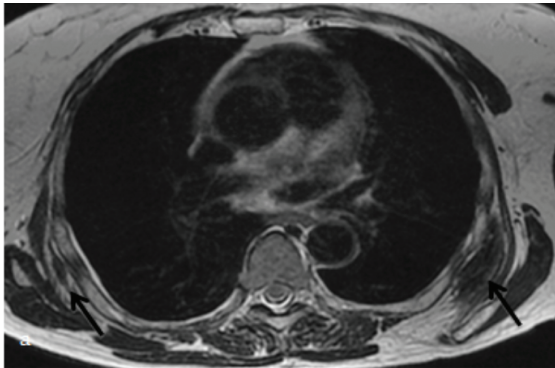


**Figure 2:** a : X-ray radiography shows a soft tissue opacity (arrow) b : Axial unenhanced computed tomographic (CT) scan: a right sub scapular ill-defined, heterogeneous soft tissue mass bordered by serratus anterior dorsi and ribs latissimus c : on MRI, sagittal T2 weighted image, the tumor shows heterogeneous signals of iso and hyperintensity

the majority of the mass being intermediate (similar to muscle) on T1-weighted images with linear strands of highly hyperintensity signal similar to subcutaneous fat (Figure 3, Figure 4). After injection of Gd-DTPA no significant enhancement of the signal intensity was detected.



**Figure 3 :** On MRI, axial unenhanced T1-weighted image of the thorax demonstrates bilateral sub scapular mass. The lesion shows typical MR signal intensity characteristics of elastofibroma : mass with signal intensity similar to that adjacent skeletal muscle with interspersed linear areas of the high signal intensity.



**Figure 4 :** On MRI, axial T2 weighted image, the tumor shows heterogeneous signals of iso and hyperintensity; (b), on coronal short time inversion recovery (STIR) T2WI (b'), the high signals can be suppressed by STIR sequence, suggesting the existence of fat tissue

## DISCUSSION

Elastofibroma dorsi is a rare benign soft-tissue pseudotumor characterized by accumulation of collagenized tissue with elastic fibers. It was first reported by Jarvi and Saxen in 1961 [1]. Elastofibromadorsi has typically been found in elderly women and almost always arises from the tissues between the lower angle of the scapula and the thoracic wall, mostly on the right side, deep to the rhomboid and latissimusdorsi muscles, with attachment to the periosteum and ligaments in the region of the sixth, seventh and eighth ribs [2]. In most cases, the lesion is unilateral; however, bilateral involvement is present in approximately 10% and in this case, the tumors have asynchronous development. Elastofibroma also has been found in extrascapular sites, including the small bowel, tracheobronchial tree, colon, deltoid region, olecranon, great trochanter, and foot [3]. The pathogenesis of these lesions remains unknown. Some authors suggest elastofibroma is a reactive process initiated by mechanical friction of the scapula on the rib cage because many patients have a history of intensive and repetitive manual labor. Other authors suggest that degenerative changes in collagen due to vascular insufficiency are the etiologic factors responsible for tumor formation. It appears as a firm mass located below the inferior angle of the scapula, attached to the deep plane of the ribs but not to the overlying skin. Symptomatic lesions generally have a diameter greater than 5 cm. The most commonly reported clinical symptoms are stiffness (approximately 25% of patients) and pain (10% of patients) [4].

Conventional radiography reveals no significant findings. US performed with high resolution equipment allows precise estimates of the size of the elastofibroma Dorsi, and usually reveals the typical striped appearance caused by the presence of alternating hypoechoic (adipose tissue) and hyperechoic (fibrosis) bands. [5,6]. The patient should be examined in the position that maximizes the visibility of the mass (in most cases with the back curved and the shoulders rolled forward). Color Doppler studies may reveal a few small flow signals within the mass [7]. Computed tomography and MRI provide more precise data on the size of large elastofibroma Dorsi. They are also better for visualizing the peculiar internal structure of the mass. On CT, elastofibroma is typically a poorly defined, inhomogeneous soft-tissue mass with attenuation similar to that of skeletal muscle, containing linear streaks of fat attenuation as seen in the two first cases.

On MR imaging, the appearance is similar-soft tissue with signal intensity similar to that of skeletal muscle on both T1- and T2-weighted images, interlaced with streaks of tissue with the signal intensity of fat. This appearance corresponds to areas of dense fibrous connective tissue

interlaced with areas of fat. Heterogeneous enhancement following gadolinium administration can be seen. Deformation of adjacent bony structures has also been reported [8]. One study categorized ED into 3 subgroups based on MRI findings. Type A is inhomogeneous, fasciculated type as case n°3; type B is inhomogeneous, non-specific type; and type C is isointense type with homogeneous muscle as cases n° 2 and 4. Average positive predictive value and sensitivity were found to be 93.3% and 100%, respectively [9].

Biopsy was performed in 2 cases of Nagano study [10] because, Preoperative MRI revealed an unencapsulated and lenticular-shaped mass of low intensity in T1-weighted images and iso- or low-intensity in T2-weighted images, reflecting the fibrous tissue content of the mass, which is in contrast to the characteristic high-intensity strands produced by fatty tissue on T1-weighted images and intermediate-intensity strands on T2-weighted images. Recently, reports about ED detection at positron emission tomography were published in the literature. Mild or moderate fluorodeoxy- glucose uptake was frequently observed and should not be interpreted as a malignant finding [11].

Histologically, macroscopic findings point out soft tissue typically characterized by an irregular, poorly defined fibroelastotic mass with an elastic consistence. The cut

surface show strands of white and yellow tissue caused by the entrapment of fatty remnants. The tumours were not encapsulated. In microscopic findings, elastofibroma is composed of fibrous, collagenous strands and sometimes elongated mostly round-shaped elastic fibres which were densely packed [12].

The typical location of the tumor, long-term symptoms, and typical radiological findings, particularly MRI, are generally sufficient for ED diagnosis. Biopsy is generally not necessary with these findings. There is no evidence of malignant transformation of ED. Alberghini reported a case having high spindle cell sarcoma together with ED. Therefore, lipoma, hemangioma, and soft tissue sarcoma should be kept in mind for differential diagnosis in cases where there is rapid growth, significant contrast involvement, and atypical location [13].

## CONCLUSIONS

When dealing with tumors of the chest wall, elastofibromas can usually be diagnosed on the basis of their imaging characteristics. Thus, we report this case to heighten radiologist awareness of the imaging features of elastofibromadorsi, so that we decrease misdiagnosis of this soft-tissue tumor as malignant entity and avoid unnecessary biopsies and surgical removal.

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