

Elastofibroma dorsi: what do we need to know about it?

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Learning objectives

To illustrate the pathognomonic imaging findings (US, CT and MR) in a series of patients observed in our department between August 2006 and November 2012. To suggest a management algorithm for typical and atypical cases of disease.

Background

Epidemiology

Elastofibroma dorsi was first described by Jaervi and Saxen in 1961 as a benign slow-growing lesion of the soft tissue characterized by fibroblastic hyperplasia and fatty tissue [1]. Generally elastofibroma dorsi is incidentally observed in up to 2% of elderly females who undergo chest CT. Instead, in autopsy studies, it has been found in 11,2% of men and 24,4% of women. The reasons for the difference in prevalence between CT and autopsy studies are related to insufficient contrast resolution in CT of the small lesions found in autoptic studies. In addition, the mentioned difference could be attributed to radiologists' lack of confidence in reporting elastofibroma dorsi. Therefore the reported prevalence of 2% at CT may be more appropriate in radiology. It typically occurs in female with a ratio of 4:1 F:M after age of 50 years [2]. The lesion usually arises in the deep subscapular region (93%) between the thoracic wall and the lower third of the scapula under the serratus anterior and the latissimus dorsi muscles.

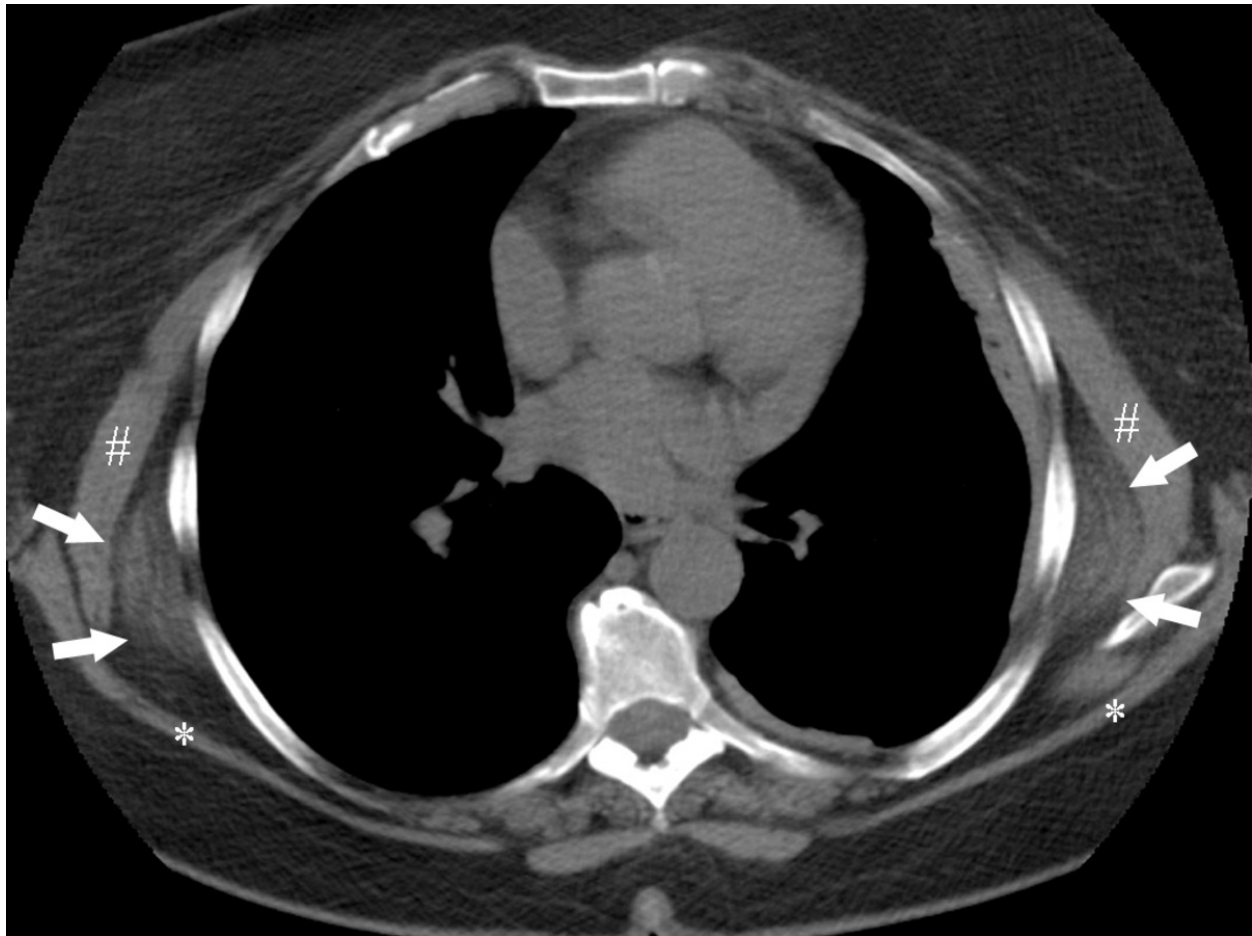


Fig. 1: A 60-years-old female with bilateral elastofibroma dorsi. Axial unenhanced MDCT scan shows subscapular semilunar masses (white arrows) with density similar to skeletal muscle, internal striations and scattered areas of decreased attenuation similar to fat signal. Masses are typically located adjacent thoracic wall, under the serratus anterior (white hash) and the latissimus dorsi (white asterisk).

References: Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele" - Catania/IT

In less than 1% of cases it can be found in extrascapular regions like deltoid, trochanter, ischium, olecranon and foot [3]. It occurs predominantly (60-70%) on the right side, but in 54% of cases, it is found bilaterally.

Etiopathogenesis

Elastofibroma dorsi has an unknown pathogenesis. At first, since frequently observed in heavy manual workers, it was supposed that friction of the scapula against the thoracic wall due to long-term manual jobs or repetitive trauma, could determine an excessive production of elastin and degenerated collagen. This hypothesis does not justify, however, the atypical site of the lesion. Another pathogenetic theory suggested reactive fibromatosis and elastotic degeneration as the pathogenetic sequence to a vascular insufficiency. Moreover, many authors have proved the lack of association

between trauma and the development of elastofibroma dorsi. Recently some authors proposed the hypothesis of a primary dysplasia of the elastic tissue favoured by an enzymatic defect which would also explain family predisposition of elastofibroma dorsi [4]. This theory was confirmed by several histological studies. A familial predisposition correlated with an underlying enzymatic defect may exist in 30%, but this has never been finally proved. Large case series from Japan strongly suggest that hereditary factors may be a predisposition for this lesion. The nature of the altered elastic fibres is still debated. They may be caused by abnormal elastogenesis or by degenerating as a secondary process, or even by a combination of both processes.

Clinical presentation

Clinically elastofibroma dorsi has a typical presentation. The lesion in 81% of cases is a solid hard mass, usually over 5 cm, commonly located in the dorsal region. It is fixed to the deep underlying soft tissue and mobile to the superficial ones. Elastofibroma dorsi, due to its location, may be visible and palpable during adduction and forward flexion of the arm. Occasionally the tumour may be impalpable because firmly constricted between the scapula, the intrinsic and extrinsic muscles of the scapula, and the thoracic wall [4]. Literature reports that the majority of patients (50 % of cases) are asymptomatic or have mild discomfort only [3]. Symptoms of elastofibroma dorsi depend on site and size and consist in clicking, snapping and clunking during the anteposition of the scapula, restriction of shoulder movements and rarely shoulder pain. This clinical presentation may be misdiagnosed as a rotator cuff tear or subacromial bursitis. Macroscopically elastofibroma dorsi presents as a white-yellowish mass, 5-10 cm in size, with hard-elastic consistency; it is generally non-encapsulated and firmly adherent to scapula and ribs separated by connective tissue. Microscopically all the lesions show fibroelastic paucicellular tissue with copious collagen and elastic fibers alternated by marked eosinophilic substance and areas of mature fat cells mixed with fibroblasts in absence of mitotic abnormalities [5]. Considering the dramatic intensification of computed tomography (CT) and magnetic resonance (MR) examinations, a progressive increase in the incidental detection of scapulohumeral elastofibroma is likely to occur.

In fact, in our series of patients, elastofibroma dorsi was mainly observed as an occasional finding. Its site and clinical presentation could mimic other bone or soft tissue tumors like: lipoma, desmoids tumour, sarcoma or metastasis. Therefore there is a possibility that elastofibroma dorsi is misinterpreted, it is possible to misdiagnose it as suspected neoplasms by radiologists unaware of the features of this disease [6].

PREVALENCE	> 2%
GENDER	4:1
LOCATION	Typical: subscapular region among thoracic wall and the lower third of scapula. Atypical: deltoid, trochanter, ischium, olecranon, foot.
CLINICAL PRESENTATION	<u>Asymptomatic or mild discomfort (50%)</u> <u>Symptomatic</u> : clicking, snapping, clunking during scapula movements; restriction of movements, pain (rarely).

Table 1: Table summarizing main characteristics of elastofibroma dorsi.

References: Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele" - Catania/IT

Imaging findings OR Procedure details

Clinical findings associated with imaging lead to a correct diagnosis without the need of confirmation from biopsy. Radiological methods consist especially in ultrasound (US), CT and MR. US is the first examination performed because non-invasive, quick and inexpensive. The image is more readable by scanning the patients prone with arm abducted. US show an oval mass with ill-defined margins and 4 different patterns:

- Type I, the most frequent and typical (54% of cases), shows an inhomogeneous fasciculated pattern with fibrous and fat strands.
- Type II (22%) presents an aspecific inhomogeneous pattern;
- Type III (15%) reveals a homogeneous pattern prevalently hyperechoic
- Type IV (9%) has a homogeneous pattern prevalently hypoechoic.

Color-Doppler is necessary because all lesions show a lack of intrinsic vascularization. CT and MR usually are performed to confirm characteristic US findings and to study atypical lesions or those that may require surgery. They are able to visualize the semilunar or round-shaped lesion in the deep periscapular region, with well-defined margins, lack of parietal infiltration and of reactive perilesional edema. CT appearance of elastofibroma dorsi is diagnostic. The scan shows a poorly defined soft-tissue mass with density similar to that of the adjacent muscle with internal striations or scattered areas of fat attenuation (low-density) [3]. However, elastofibroma with homogeneous attenuation at CT without internal foci of low density have been reported. This appearance seems to be typical of smaller lesions in early stages of development. The application of intravenous contrast agent generally shows a mild enhancement, but also a marked enhancement that could mimic malignant lesions [7]. So the application of contrast agent is considered useless Fig. 2 on page 11.

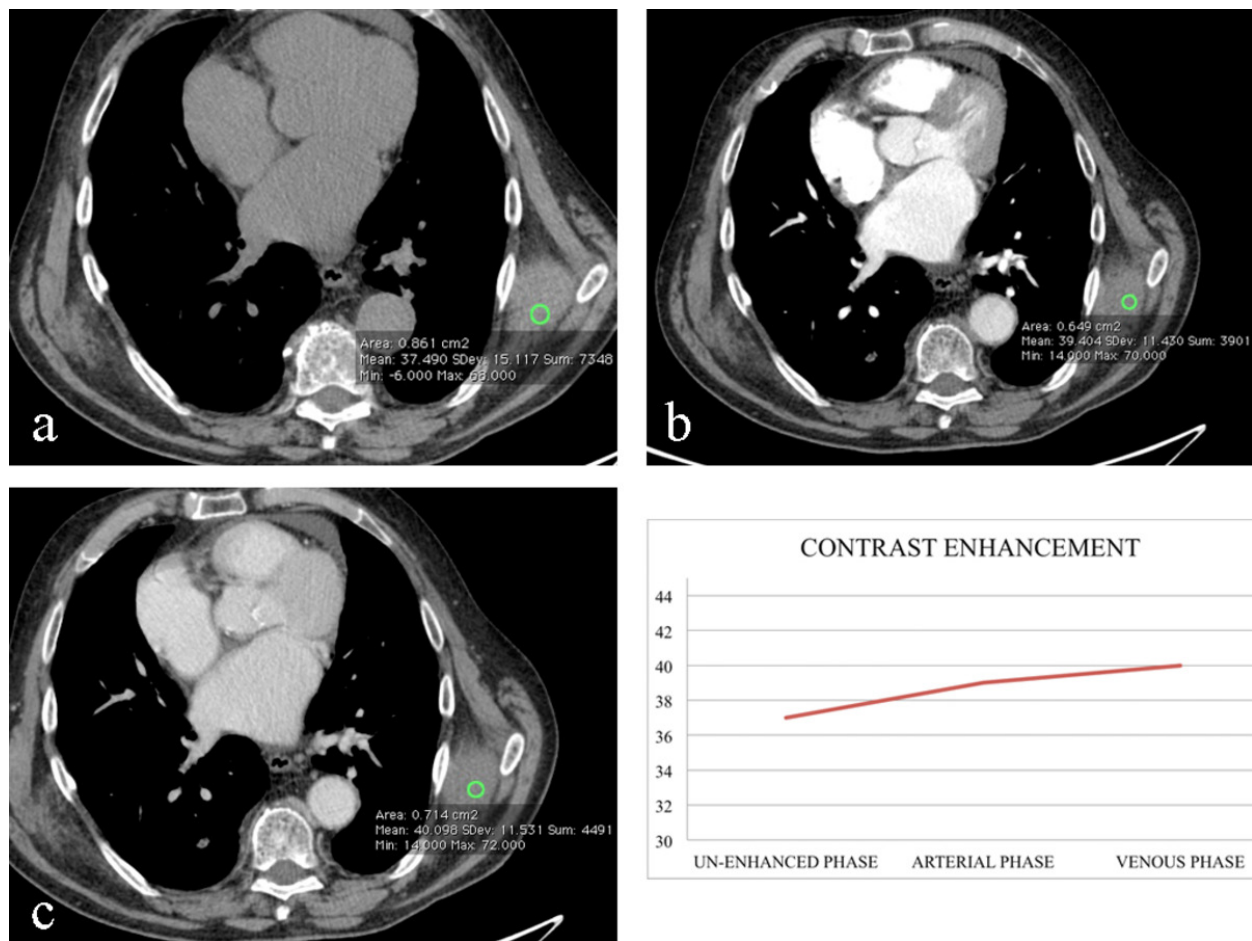


Fig. 2: A 81-years-old male with bilateral elastofibroma dorsi. Axial unenhanced (figure 2a), arterial phase (figure 2b) and venous phase (figure 2c) MDCT scans show a mild inhomogeneous contrast enhancement. The histogram shows a typical enhancement pattern in different phases.

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Nevertheless, MR is more reliable than CT to demonstrate fatty areas and a clear cleavage from adjacent tissues. MR reports an alternating pattern of fibrous and adipose tissues. The fibrous tissue has signal of low intensity, relative to skeletal muscle, with both T1- and T2-weighted sequences and fatty tissue shows high signal intensity on T1 and intermediate on T2-weighted sequences. The presence of fatty tissue is confirmed with STIR (short T1 inversion recovery) sequences, specific to suppress the signal from fat. STIR sequences reveal the low signal intensity of fatty tissue suppressed with a slightly higher signal intensity of the fibrous tissue. In contrast-enhanced MR images there are both areas with and areas without enhancement. Therefore, on T1-weighted MR images, the majority of the mass is isointense relative to skeletal muscle with scattered foci of linear and curvilinear hyper-intense. The margins of this mass are relatively well defined and non-encapsulated [Fig. 3](#) on page 12.

At CT and MR it is possible to detect 3 patterns:

- [Type A](#) (84%) inhomogeneous fasciculated compatible with Types I, III and partially with Type II US pattern,
- [Type B](#) (8%) inhomogeneous aspecific compatible with Type II US pattern;
- [Type C](#) (8%) homogeneous isodense or isointense to the muscle compatible with Type IV US pattern [3].

Literature reports a small number of cases of hyper-metabolism in a subscapular mass with the classic CT or MR imaging findings of elastofibroma dorsi at positron emission tomography-CT (PET-CT) that should be kept in mind [8]. The pathognomonic fasciculated pattern of elastofibroma dorsi at CT and/or MR discriminates it from other tumours of the soft tissues like lipoma (tender mass, mobile on deep and superficial planes with well-defined margins and a homogeneous pattern); desmoid tumour (hard solid mass, fixed to the adjacent planes, with irregular and ill-defined margins and with marked and uniform enhancement) soft tissue sarcoma/metastasis (clearly inhomogeneous mass with areas of marked enhancement, sign of local infiltration and bone erosion) [3]. When atypical suspected radiological patterns are present at CT and/or MR, it is necessary to perform a biopsy in order to exclude malignant tumours. Some authors suggest that clinical data associated to typical imaging findings (particularly MR) are strongly diagnostic for elastofibroma dorsi [Fig. 4](#) on page 13 [Fig. 5](#) on page 14.

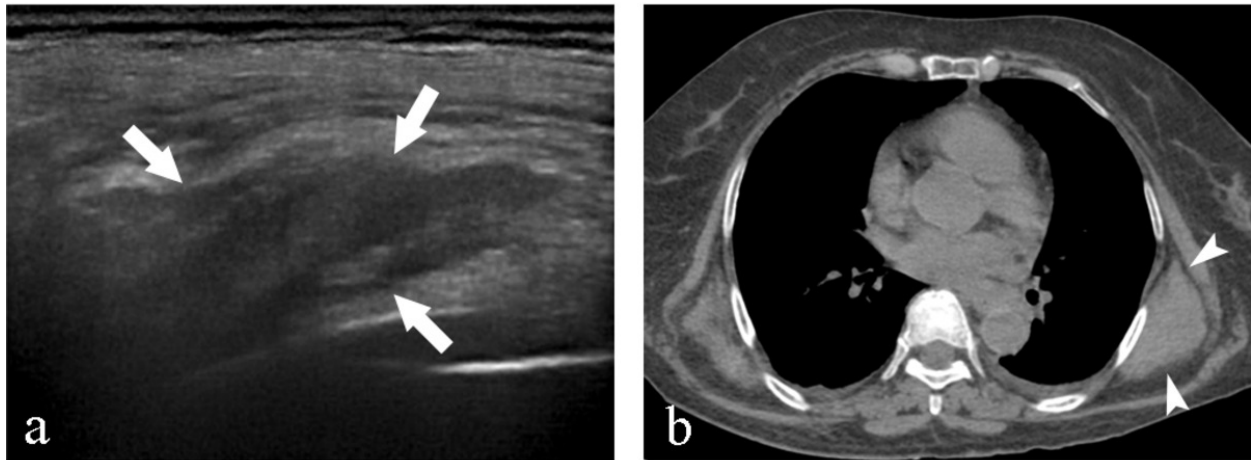


Fig. 4: A 74-years-old female with elastofibroma dorsi. Ultrasound image (figure 4a) shows a lenticular poorly circumscribed mass (white arrows) below the serratus anterior muscle with an inhomogenous pattern characterized by alternating of hypo- and hyperechoic areas. Axial unenhanced MDCT image (figure 4b) confirms the presence of this round-shaped mass (arrow heads) in the deep periscapular region with well-defined margins and lack of parietal infiltration (CT pattern type C). Lesion presents with density similar to adjacent muscles and with internal striations and scattered areas of low-density similar to adipose tissue.

References: Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele" - Catania/IT

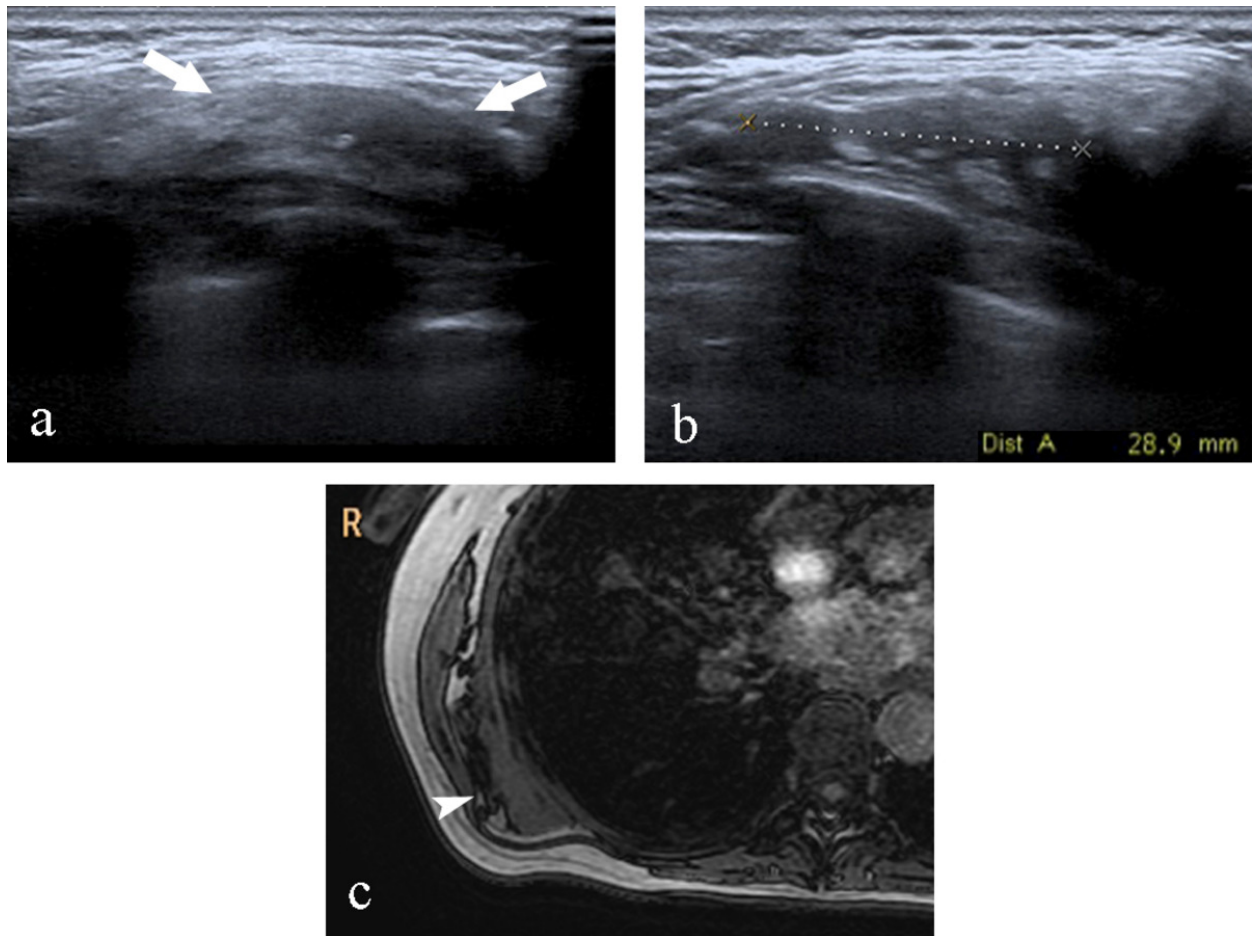


Fig. 5: 55 years-old female with elastofibroma dorsi located in the right side. Ultrasound images (figures 5a-b) show the semilunar mass (white arrows) closely adjacent to intercostal muscles with an inhomogenous pattern characterized by alternating of hypo- and hyperechoic areas (white arrows). Axial in-phase T1 weighted MR acquisition (figure 5c) confirms the presence of mass (arrow head) on the right side between chest wall and subscapular region. It presents with intermediate signal intensity similar to that of adjacent skeletal muscle combined with linear areas of increased signal intensity similar to adipose tissue.

References: Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele" - Catania/IT

Management

The management of this lesion is somewhat controversial.

In elderly asymptomatic patients a conservative treatment with a simple observation for 2-3 months is recommended, because the literature does not report malignant transformation in the natural history of elastofibroma dorsi. If follow-up shows a growth of the mass, biopsy is necessary.

On the other hand, surgery could be evaluated for symptomatic lesions or esthetical reasons.

In these cases marginal excision is suggested, although several reports show a high incidence of postoperative complications such as seroma or hematoma [Fig. 6](#) on page 15 [Fig. 7](#) on page 16.

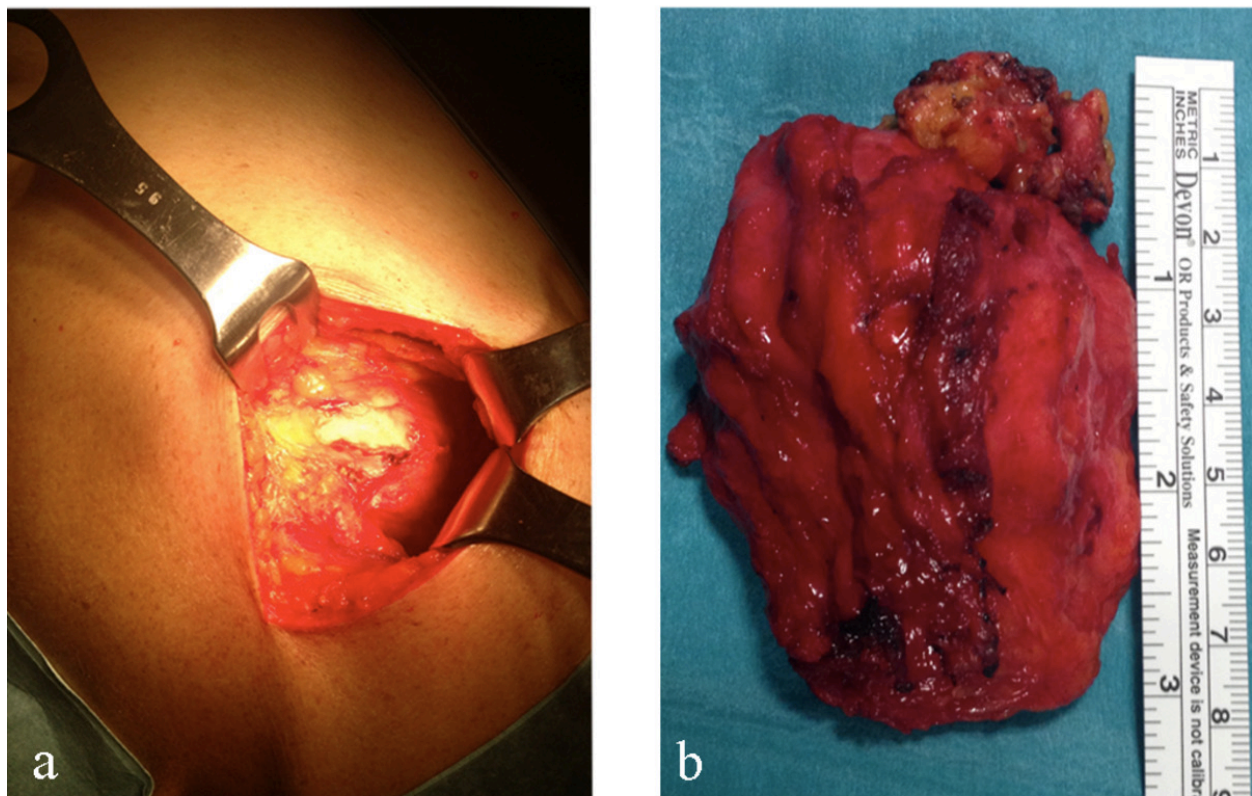


Fig. 7: Surgical approach of elastofibroma dorsi (figure 7a). Macroscopic appearance of elastofibroma dorsi (figure 7b) shows a no-encapsulated mass with fibrous and fatty areas. Courtesy of Professor Migliore Marcello, Department of Thoracic Surgery, University of Catania, Catania, Italy.

References: Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele" - Catania/IT

These concepts regarding management of elastofibroma dorsi have been schematized by some authors in a simple algorithm; this algorithm could help the clinician to make quick and safe decisions [9].

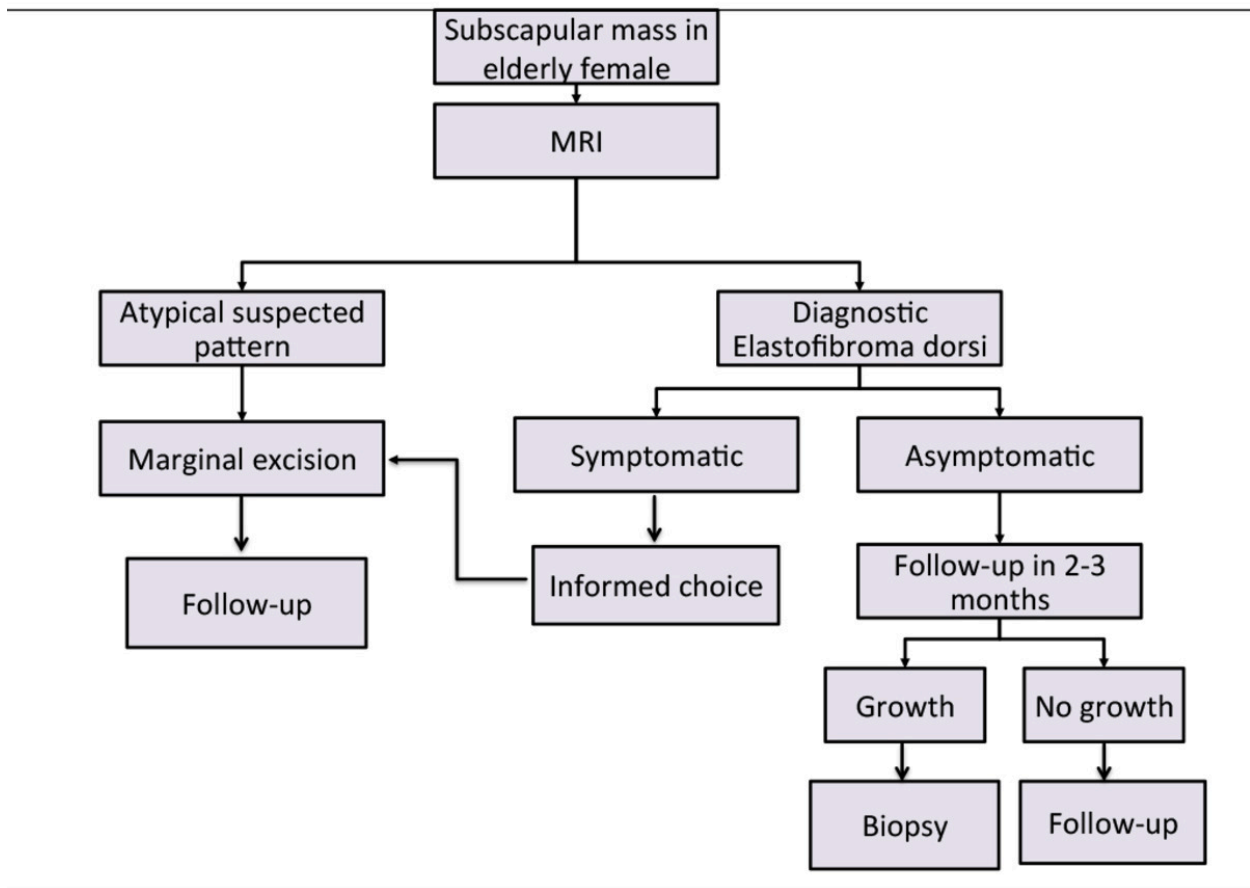


Table 2: Table summarizing management of elastofibroma dorsi.

References: Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele" - Catania/IT

Images for this section:

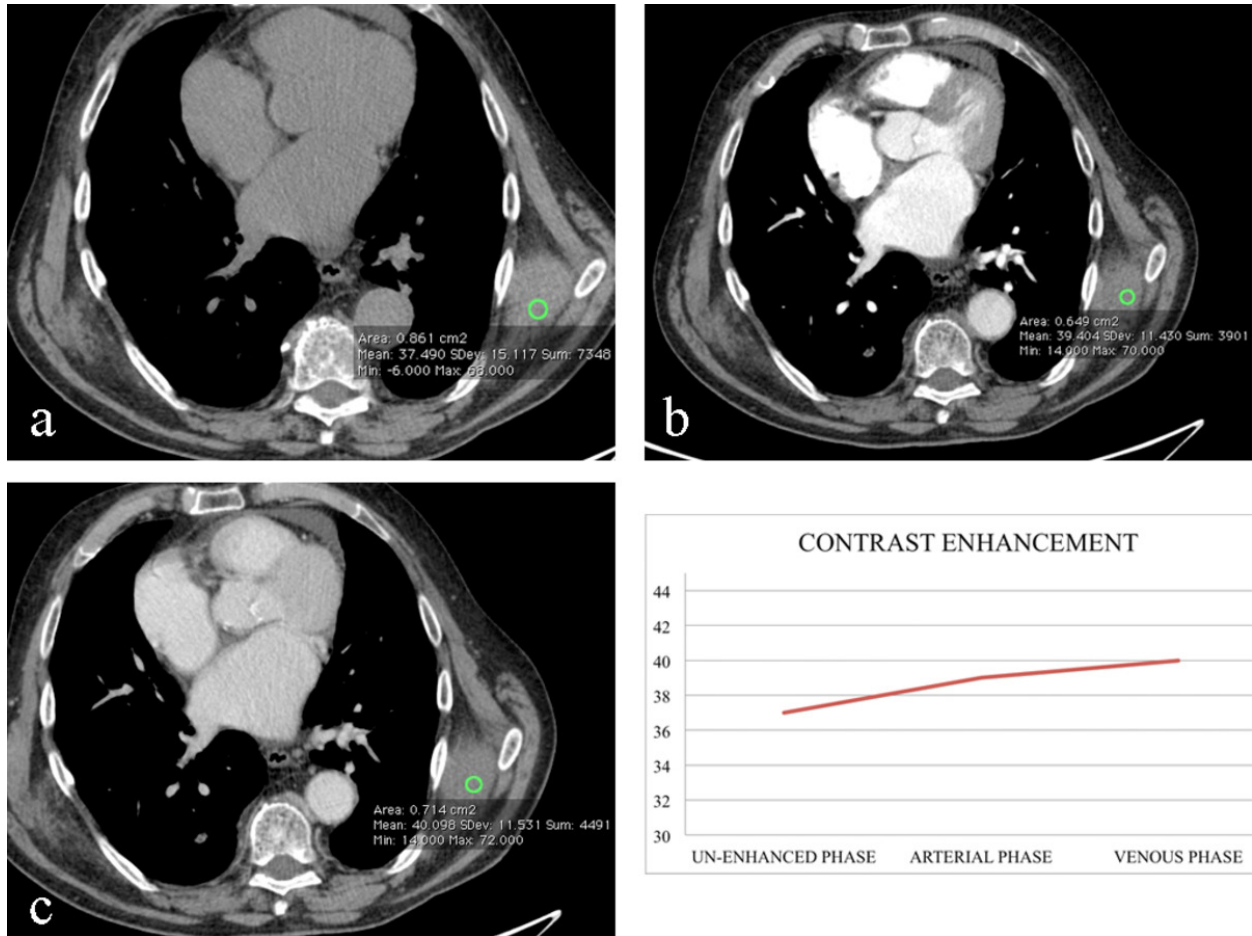


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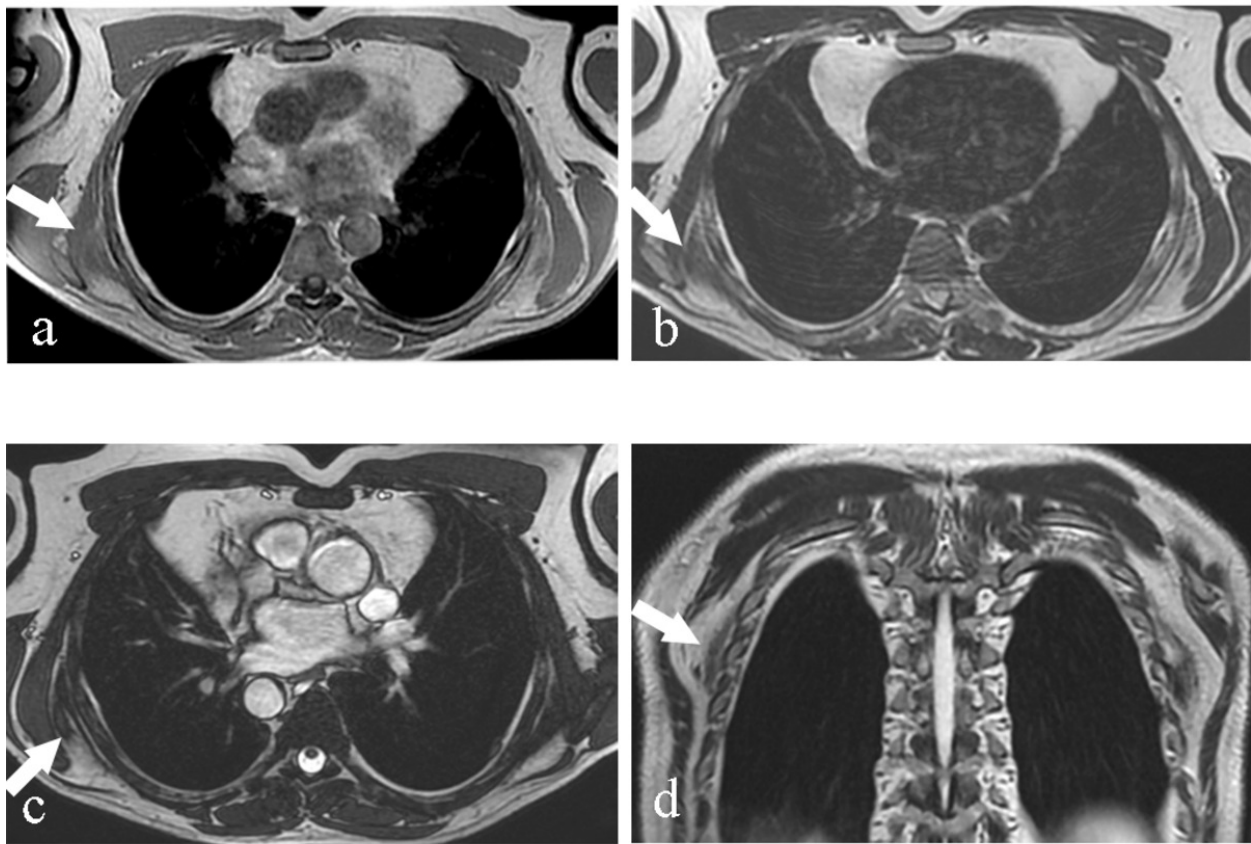


Fig. 3: A 65 years old male with bilateral elastofibroma dorsi, predominant in the right side. Axial in-phase T1-weighted (figure 3a), Fast Recovery Fast Spin Echo (FRFSE) T2-weighted (figure 3b), Steady State Free Precession (SSFP) T2-weighted (figure 3c) and coronal Fast Recovery Fast Spin Echo (figure 3d) MR acquisitions show a well-defined heterogeneous mass between chest wall and subscapular region. The lesion (white arrows) appears hypo-isointense, relative to adjacent skeletal muscle, with interspersed fatty foci or linear areas of hyperintensity.

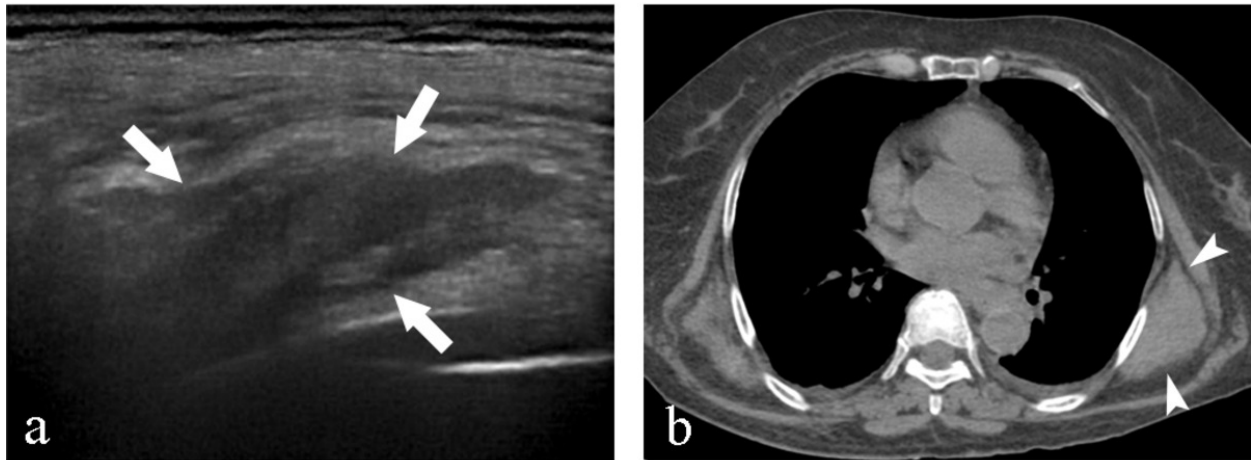


Fig. 4: A 74-years-old female with elastofibroma dorsi. Ultrasound image (figure 4a) shows a lenticular poorly circumscribed mass (white arrows) below the serratus anterior muscle with an inhomogenous pattern characterized by alternating of hypo- and hyperechoic areas. Axial unenhanced MDCT image (figure 4b) confirms the presence of this round-shaped mass (arrow heads) in the deep periscapular region with well-defined margins and lack of parietal infiltration (CT pattern type C). Lesion presents with density similar to adjacent muscles and with internal striations and scattered areas of low-density similar to adipose tissue.

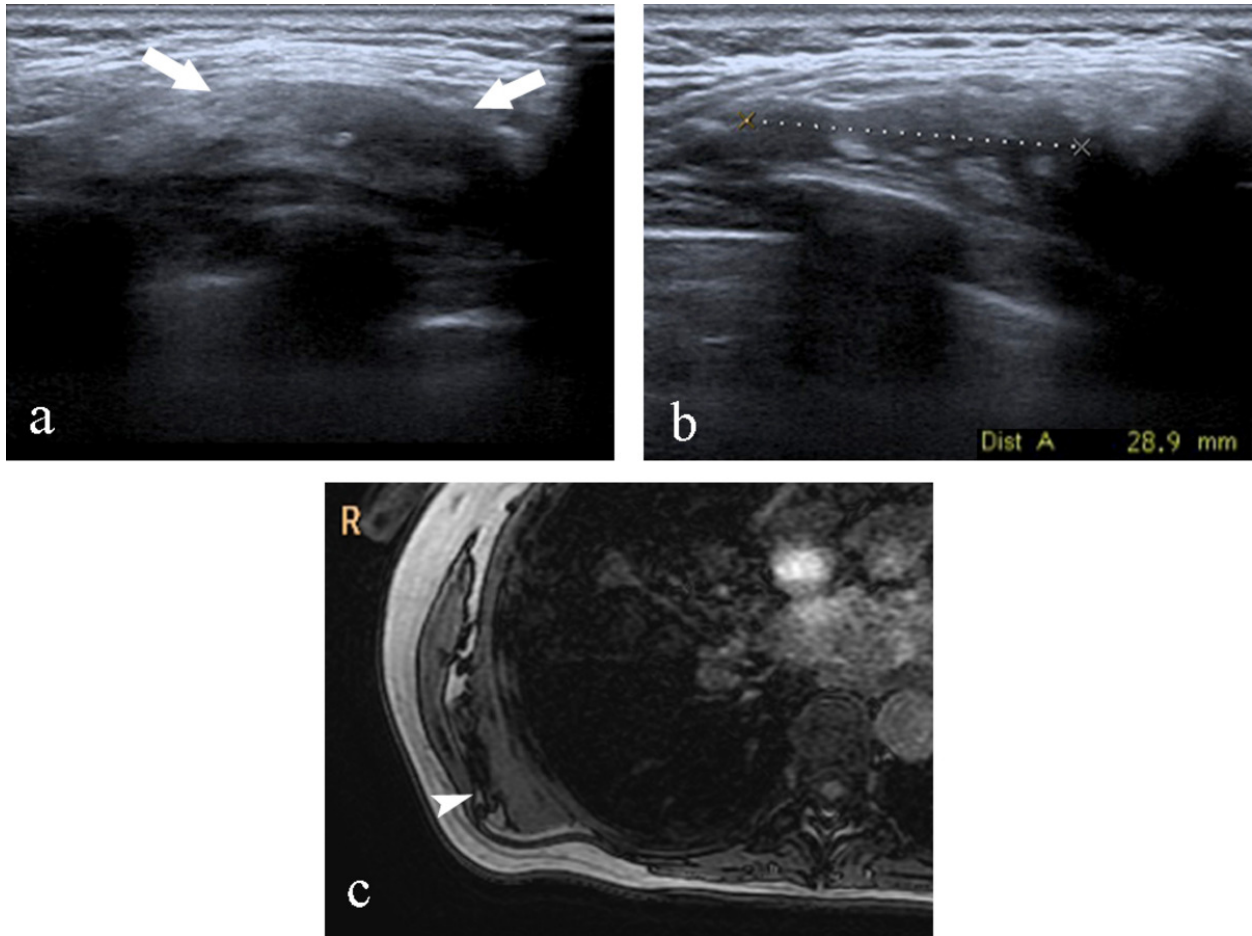


Fig. 5: 55 years-old female with elastofibroma dorsi located in the right side. Ultrasound images (figures 5a-b) show the semilunar mass (white arrows) closely adjacent to intercostal muscles with an inhomogenous pattern characterized by alternating of hypo- and hyperechoic areas (white arrows). Axial in-phase T1 weighted MR acquisition (figure 5c) confirms the presence of mass (arrow head) on the right side between chest wall and subscapular region. It presents with intermediate signal intensity similar to that of adjacent skeletal muscle combined with linear areas of increased signal intensity similar to adipose tissue.

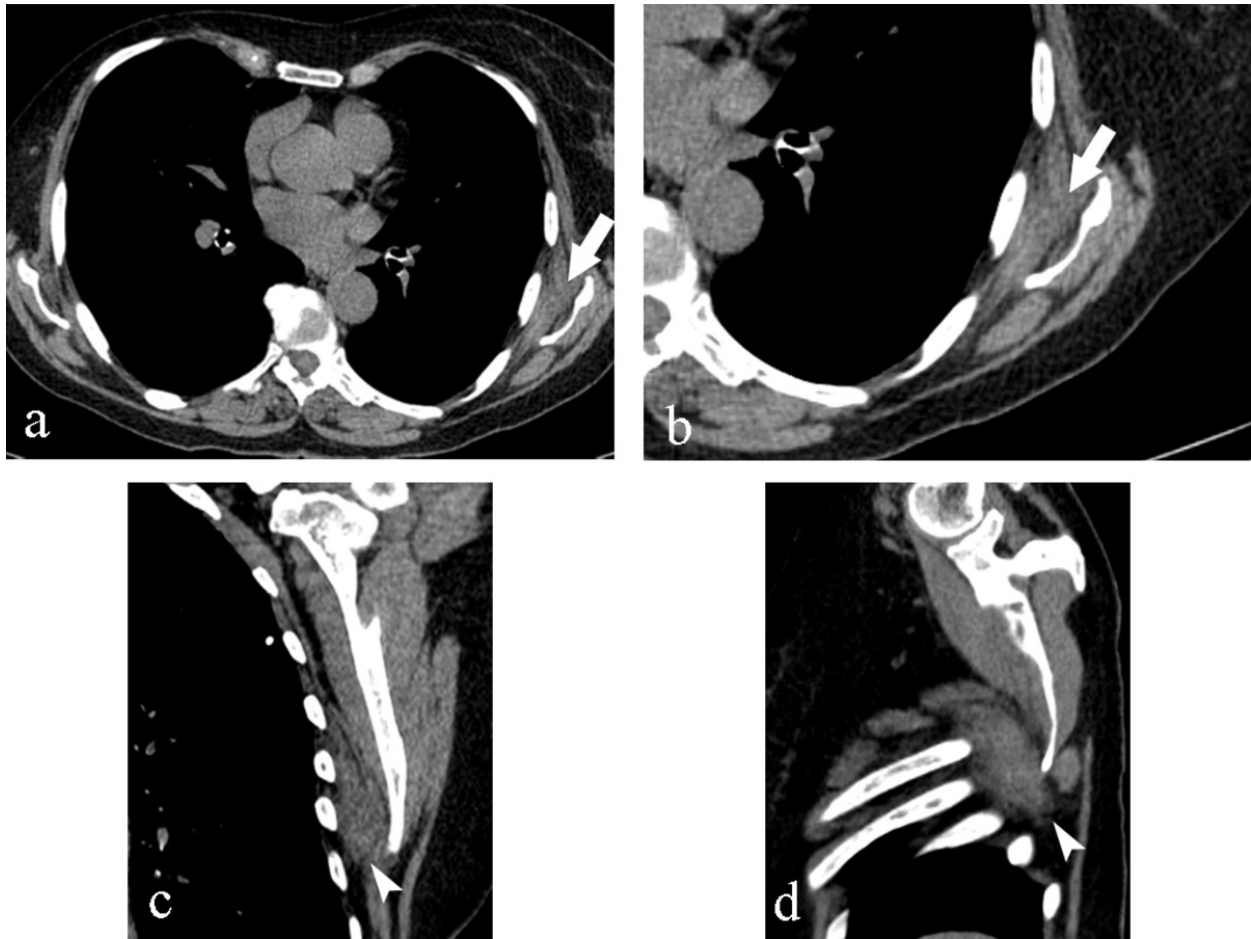


Fig. 6: A 74-years-old-woman with elastofibroma dorsi in the left side. She was an industrial worker; she complains restriction of scapula movements and clunking during ante-position of the scapula. Axial unenhanced MDCT scan shows a mass (white arrow) in the deep periscapular region firmly attached to the rib cage and intercostal muscle (figure 6a-b). Coronal (figure 6c) and oblique multiplanar reformattation (figure 6d) MDCT images show correct location of lesion (arrow head), between thoracic wall and the lower third of the scapula.



Fig. 7: Surgical approach of elastofibroma dorsi (figure 7a). Macroscopic appearance of elastofibroma dorsi (figure 7b) shows a no-encapsulated mass with fibrous and fatty areas. Courtesy of Professor Migliore Marcello, Department of Thoracic Surgery, University of Catania, Catania, Italy.

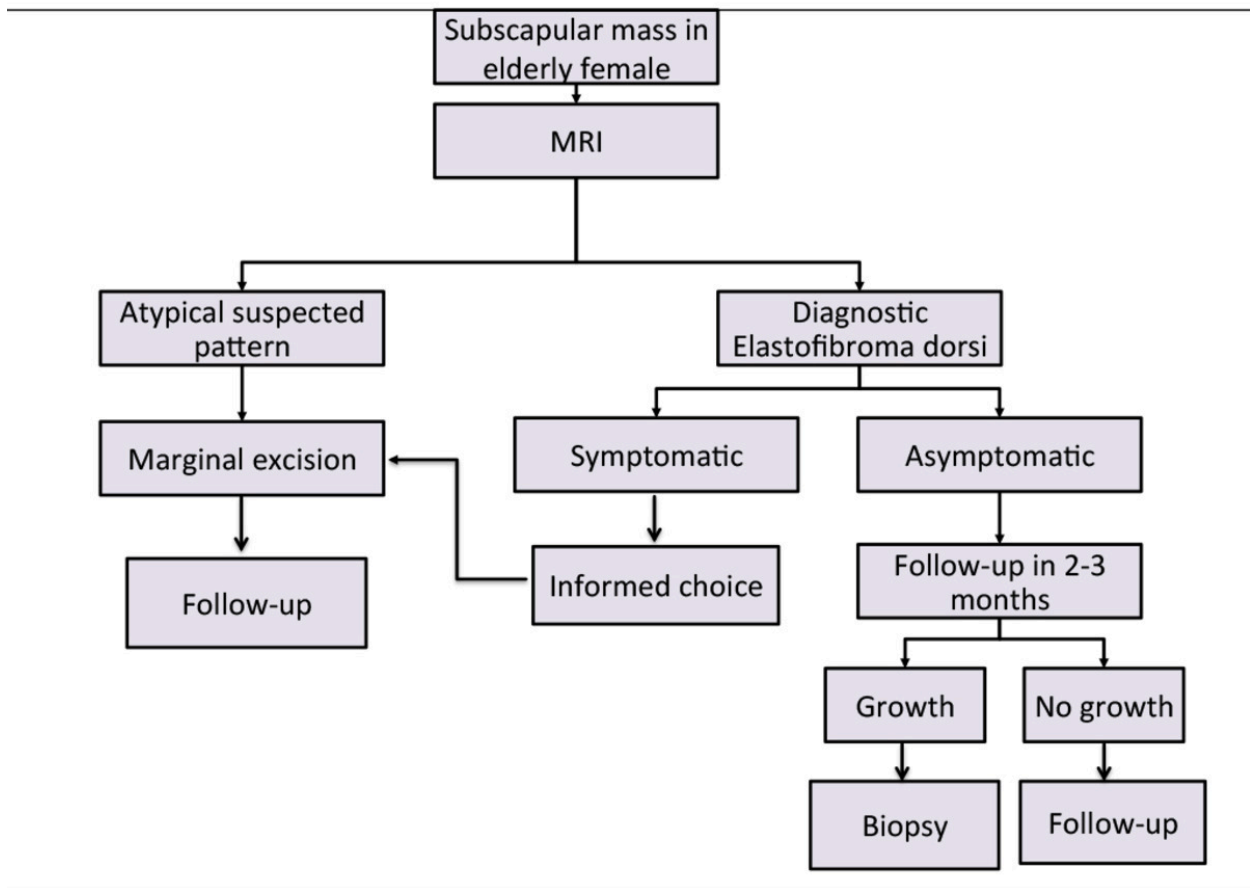


Table 2: Table summarizing management of elastofibroma dorsi.

Conclusion

Diagnosis of elastofibroma dorsi can be performed by means of its characteristic clinical data and radiological findings; biopsy is reserved only in cases of atypical patterns or typical ones with secondary growth, in order to differentiate them from malignant tumours.

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