



Elastofibroma: management and surgical outcome



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Elastofibroma: management and surgical outcome

Elastofibroma is a benign slow-growing neoplasm of soft tissues, originally defined Elastofibroma dorsi because of the typical localization in the connective tissue placed between the bottom corner of the scapula and the chest wall.

From 1990 to 2013 at our center, 115 patients underwent elastofibroma surgical removal, including 2 bilateral and one relapsed. For all the patients ultrasound preoperative examination was requested, sometimes with a diagnosis of lipoma or fibrolipoma because of the rarity of this type of lesion and therefore the lack of experience of the radiologists.

In all 115 patients the lesion was detected and removed, only in 7 cases it was necessary, intraoperatively, to mobilize the upper limb and shoulder in order to better visualize the lesion.

On the operating table the lesion was situated below the muscle planes and looks like a solid mass, oval, with a pole firmly attached to the periosteum of the ribs and intercostal ligaments, with net margins and a diameter of 5-10 cm. The sides not attached to the chest wall were in continuity with the adipose tissue. The cutting surface was pink-grayish, with fibrous appearance. Foci of cystic degeneration interspersed with islands of fat that are vaguely reminiscent of the fibrolipoma could be found.

The treatment of choice is the surgical excision and subsequent histological examination solve the diagnostic dilemma. If the removal is radical, it is definitive because the lesion has no tendency to relapse.

KEY-WORDS: Elastofibroma, Elastofibroma dors, Elastic fibers, Surgical excision.

Introduction

The elastofibroma is a benign neoplasm of soft tissues described for the first time in 1961 by Jarvi and Saxen¹. It is a slow-growing lesion, fibro-elastic consistency, not encapsulated, with ill-defined margins.

It was originally defined *Elastofibroma dorsi* because of the typical localization in the connective tissue placed between the bottom corner of the scapula and the chest wall².

This name, which is still used by many authors, does not include all the different locations of this neoplasm. In the literature, in fact, other locations have been described :in the region of the greater trochanter³, in the deltoid⁴, at the level of the ischial tuberosity⁵, in the mammary region⁶ and in the oral region⁷.

It should however be noted that the site most affected, in all the case series in literature, is between the scapula and the chest wall.

The exact incidence of elastofibroma is not known. According to Enzinger and Weiss⁶ have been reported

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only 100 cases since 1961, the year in which it was first described this disease. Also according to these Authors, the true incidence was underestimated due to many cases never published and to the difficult clinical diagnosis of the lesion.

Elastofibroma is found in individuals over the age of 55 years, especially females, and is bilateral in a percentage in the literature ranging between 10 and 25% of cases^{6,8-11}.

From the clinical point of view the lesion, easily passes unnoticed due to poor symptoms.

The mass is deeply situated and, due to the slow growth, for a long time may not be visible. In fact, the most frequent location (below the lower apex of the scapula and below rhomboid and latissimus muscles) and adhesions to the chest wall (with the periosteum of the ribs and intercostal ligaments of the seventh and eighth ribs) make difficult, if not impossible, to highlight it when the lesion is small.

When the size increases the lesion begins to come out of hiding only in the movements of the shoulder, and remained permanently outside when the size will increase again.

Often the patient becomes aware of its presence by the feeling of a "shot" in the movement of abduction of the



Fig. 2: Macroscopic appearance of the surgical specimen.



Fig. 3: Surgical excision in a 59 year old woman.



Fig. 1: Surgical excision in a 65 year old man. The neoplasm was located below the lower apex of the scapula and below rhomboid and latissimus muscles.

scapula, when the mass literally "exits" by the scapular angle and can be appreciated clinically.

Clinically elastofibroma is characterized by hard consistence, fixed on the deep layers (chest wall) and free on the superficial plans, with poor and non-specific symptoms, only rarely can cause soreness accentuated by the movement.

With ultrasound it is often confused with lipomatous lesions, although the typical location must always make a diagnosis of elastofibroma. The operating table and histological examination confirms the diagnosis.

Material and Methods

From 1990 to 2013, 115 patients underwent elastofibroma surgical removal, including 2 bilateral and one relapsed.

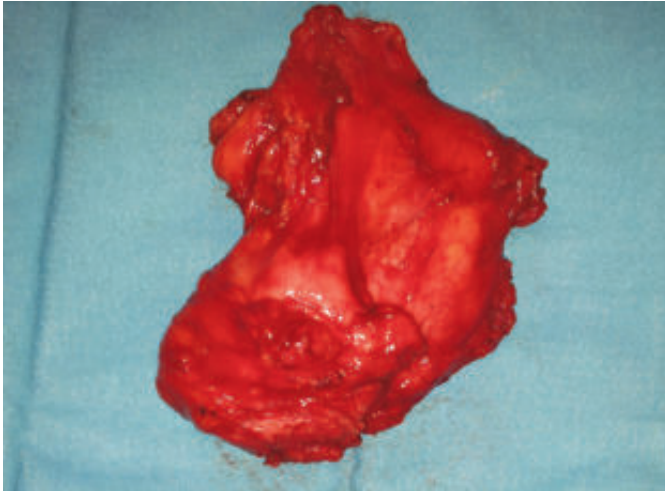


Fig. 4: Macroscopic appearance of the surgical specimen; note the pink-grayish color of elastofibroma.

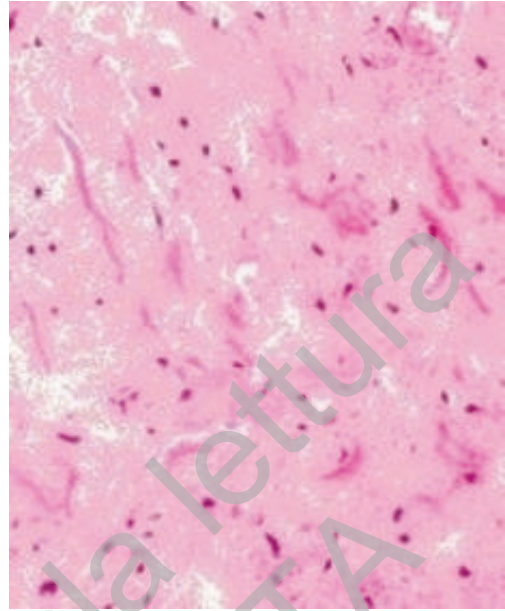


Fig. 7: Numerose fibre collagene eosinofile, allungate, con minore quantità di fibre elastiche (x250).



Fig. 5: Surgical excision in a 45 year old man. The lesion was firmly attached to the periosteum of the ribs and intercostal ligaments.

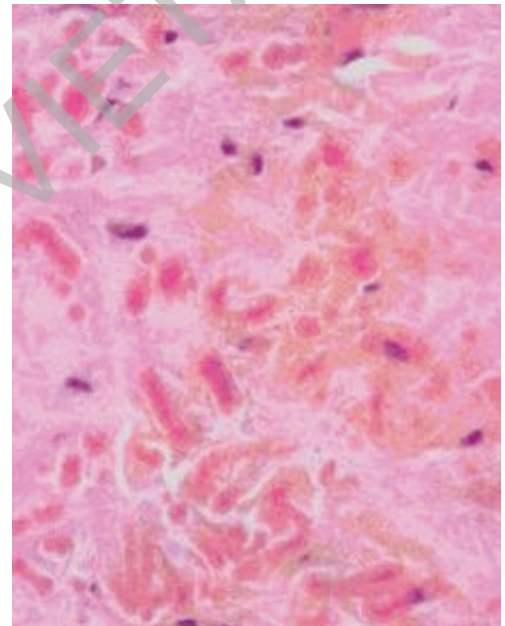


Fig. 8: Fibre elastiche di forma variabile, irregolare, talora serpiginose (X250)



Fig. 6: Macroscopic appearance of the side attached to the periosteum.

All the lesions were located under the shoulder blade, on left (50) or right (67) side, almost all (102) permanently protruding outside of the angle of the scapula, a few ¹³ not completely visible.

The removal of the latter was obviously more laborious and the only recurrence in our case series belonged to this group.

For all lesions ultrasound preoperative examination was requested, sometimes with a diagnosis of lipoma or fibrolipoma.

This is because of the rarity of this type of injury and therefore the lack of experience of the radiologists. In fact, when the examination was performed in major centers, the diagnosis has always been of elastofibroma.

Only two cases were referred to us having made the MRI, which had not been of any help in the performance of surgery.

Results

In all 115 patients the lesion was detected and removed, only in 7 cases it was necessary, intraoperatively, to mobilize the upper limb and shoulder in order to better visualize the lesion.

In all patients was positioned a suction drain, removed in the first day (79 patients), the second (26 patients) or in the third postoperative day (4 patients).

In cases of greater extension and/or greater adherence to the costal plane the drainage was kept in place for 5 - 7 days (5 patients).

Finally, 4 patients were subjected to repeated suction for the post-operative seroma formation.

Discussion

The elastofibroma's etiology is unknown, although since the first description of the tumor ¹, trauma due to friction between the scapula and the chest wall has been held responsible for an excessive deposition of abnormal collagen and elastic fibers.

In a histological study of autopsy material in 1975 ¹², the same authors showed aspects similar to elastofibroma in the tissues of the scapular region in 16% of cases examined in a series of 234 total cases.

For other authors ¹³⁻¹⁵, the typical elastic fibers of elastofibroma could represent the degeneration of collagen deposited as a result of chronic trauma.

Furthermore, the concept of the responsive mechanism to chronic trauma would be confirmed by the slow progression of the lesion, by the possibility of bilateral finding and the rare location in other sites of rubbing, as for example close to the greater trochanter, the ischial tuberosity and the deltoid region.

Finally, the low incidence of the tumor might involve other factors such as a wild sort of genetic predisposition or an enzyme defect in the metabolism of connective tissue ^{6,8,10}.

Although, in reality, it has never been proven nor an increase in the family, nor a family history of other diseases of the elastic system such as *pseudoxanthoma elasticum*. Ultrastructural studies ^{6,16} have demonstrated the presence of dense granular bodies, not surrounded by membrane, in the cytoplasm of fibroblasts and myofibroblasts, that could represent the precursor of extracellular elastin. It would therefore demonstrated the intracellular

formation of the characteristic elastic fibers of the lesion, in contrast to the theories which consider that the elastofibroma is due to the degeneration of elastic fibers or collagen already formed.

The symptoms are nonspecific and the differential diagnosis involves lipoma, fibroma, and fibrolipoma and the malignant neoplasms of soft tissue.

Among the diagnostic tests become important: the ultrasound, CT and MRI, although the confirmation is purely histological.

The execution of an ultrasound may be useful in cases where the tumor can be a big lipoma deeply located, but also an ultrasound examination positive for lipoma does not exclude an elastofibroma.

In fact, this lesion may present a more or less abundant adipose tissue, intra or especially peritumoral that can determine diagnostic confusion.

A further examination that can direct the pre-operative diagnosis is needle biopsy, although the deep position of the mass, its hard consistency and the fibrous nature of the same make difficult its execution.

It should be highlighted that, if not using a specific staining for elastin (Weigert, Verhoeff, Gomori), it could be difficult to recognize the elastic nature of the fibers. For cases where the lesion is still hidden below the scapula, the resonance is of greater utility, because it allows a good highlight also below the bone surface.

The treatment of choice is the surgical excision and subsequent examination solve the diagnostic dilemma. If the removal is radical, it is definitive because the lesion has no tendency to relapse.

On the operating table the lesion is situated below the muscle planes and looks like a solid mass, oval, with a pole firmly attached to the periosteum of the ribs and intercostal ligaments, with net margins and a diameter of 5-10 cm.

The sides not attached to the chest wall are in continuity with the adipose tissue. The cutting surface is pink-gray, with fibrous appearance. Foci of cystic degeneration interspersed with islands of fat, that are vaguely reminiscent of the fibrolipoma, can be found.

Histological examination shows a dense network of collagen and elastic fibers, fibroblasts and some aggregates of fat cells. The elastin eosinophilic fibers appear swollen and fragmented and are typically presented as numerous blood cells with a dense central core and indented margins, which correspond to the elastic fibers in section.

In conclusion, the surgical therapy allows radical excision of elastofibroma and is essential for the definitive histological diagnosis.

Riassunto

L'elastofibroma è una neoplasia benigna a lenta crescita benigna dei tessuti molli, originariamente definito elastofibroma dorsi a causa della localizzazione tipica nel

tessuto connettivo interposto tra l'angolo inferiore della scapola e la parete toracica.

Dal 1990 al 2013 presso il nostro centro, 115 pazienti sono stati sottoposti a rimozione chirurgica di elastofibroma, di cui 2 con localizzazione bilaterale ed uno recidivo. Per tutti i pazienti era stata richiesta un'ecografia pre-operatoria, a volte recante però una diagnosi di lipoma o fibrolipoma a causa della mancanza di esperienza di alcuni radiologi per la rarità della neoformazione. In tutti i 115 pazienti la lesione è stata individuata ed asportata, solo in 7 casi è stato necessario, durante l'intervento, mobilitare l'arto superiore e la spalla in modo da poter meglio visualizzare la lesione. Al tavolo operatorio la lesione è situata al di sotto dei piani muscolari e si presenta come una massa solida, di forma ovale, con un polo ben adeso al periostio delle costole e dei legamenti intercostali, con margini netti e un diametro di circa 5-10 cm.

I lati non aderenti alla parete toracica, invece, sono in continuità con il tessuto adiposo. La superficie di taglio è roseo-grigiastro, con aspetto fibroso. Possono inoltre essere riscontrati focolai di degenerazione cistica intervallati da isole di grasso che ricordano vagamente il fibrolipoma.

Il trattamento di scelta è dunque l'asportazione chirurgica e il successivo esame istologico consente di risolverne il problema diagnostico.

Infine se la rimozione è radicale, risulta essere definitiva perché la lesione non possiede alcuna tendenza a recidivare.

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