

## CASE REPORT

# Retrospective Analysis of 6 Cases of Elastofibroma Dorsi

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**Abstract.** Elastofibroma dorsi is a benign lesion of fibroelastic tissue. It often presents bilaterally, is frequently localized to the subscapular region, and is more common in middle-aged and elderly women. The pathogenesis is still unclear but it has been suggested to involve reactive, degenerative, or neoplastic processes. We present a retrospective study of the findings from 6 patients with elastofibroma dorsi. Elastofibroma should be included in the differential diagnosis of subcutaneous scapular tumors, as it is a benign lesion that only requires surgery when associated with symptoms or the lesion is extensive. Noninvasive techniques such as ultrasonography, computed tomography, and, above all, magnetic resonance imaging, may be sufficient to guide diagnosis. The results of histology and imaging are closely correlated, so the need for unnecessary biopsy and surgery may be avoided.

**Key words:** elastofibroma dorsi, elastofibroma, computed tomography, ultrasonography, elastic tissue disease.

## ELASTOFIBROMA DORSI: ESTUDIO RETROSPECTIVO DE SEIS CASOS

**Resumen.** El elastofibroma *dorsi* es una lesión benigna de tejido fibroelástico, a menudo bilateral, localizada frecuentemente en la región subescapular, más común en mujeres de edad media y ancianas. La patogénesis es desconocida, y se ha sugerido que se trate de un proceso reactivo, degenerativo o neoplásico. Presentamos un estudio retrospectivo de los hallazgos observados en seis pacientes con elastofibroma *dorsi*. El elastofibroma debe plantearse en el diagnóstico diferencial de tumoraciones subcutáneas de localización escapular, ya que es una lesión benigna que sólo requiere tratamiento quirúrgico cuando es sintomático o muy grande. Técnicas no invasivas, como la ecografía, tomografía computarizada y especialmente la resonancia magnética pueden ser suficientes para orientar el diagnóstico, con estrecha correlación entre el cuadro histológico y las imágenes radiológicas evitando biopsias y cirugías innecesarias.

**Palabras clave:** elastofibroma *dorsi*, elastofibroma, tomografía computarizada, ecografía, alteraciones del tejido elástico.

## Introduction

Elastofibroma dorsi is a rare benign process, characterized by a poorly defined mass of fibroelastic tissue and fat, commonly located in the subscapular region. It usually presents from the age of 50 years, predominantly among women. The condition was first described by Jarvi and Saxen<sup>1</sup> in 1961 as a slow growing tumor of the soft tissue. Recognition of elastofibroma is important in differential

diagnosis with subcutaneous scapular tumors, mainly in middle-aged and older patients. This type of lesion can adhere to the muscle, the periosteum of the ribs and the scapula, mimicking a malignant tumor.<sup>2</sup> There have been no reports of malignant transformation and no treatment is needed, except in symptomatic patients. Noninvasive methods, like computed tomography and magnetic resonance imaging, are very useful in diagnosis, with typical images being specific although not characteristic, meaning biopsy would only be necessary in atypical or dubious cases. Elastofibroma dorsi has received little attention in the dermatological literature,<sup>3-8</sup> and as far as we are aware no cases have been described in Spanish dermatological journals. The aim of the present study is to describe the clinicopathological and radiological characteristics of cases of elastofibroma dorsi treated in our hospital and to review recent theories on pathogenesis.

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**Table.** Clinical data and radiological examinations

	Age	Sex	Clinical Characteristics	Radiological Studies	Bilateral Location	Diameter (cm)	Treatment and Follow-up
1	57	Male	Left subscapular tumor	CT Ultrasound	No Left	L 9 × 4	No 2 mo
2	71	Female	Left subscapular tumor Pain in left shoulder	CT Ultrasound	Yes	R 6.3 × 8 × 3.5 L 7.8 × 8 × 5	No 10 mo
3	64	Male	Subcutaneous right subscapular tumor	CT	No Right	R 8 × 6 × 2.5	Resection 24 mo
4	50	Female	Elastic right subscapular subcutaneous tumor	CT	No Right	R 4 × 2	No 30 mo
5	44	Male	Right subscapular tumor observed following an injury	CT Ultrasound	Yes	D. 8.6 × 13 × 5 L 10 × 12 × 3.4	No 12 mo
6	53	Female	Right subscapular tumor	CT Ultrasound	Yes	R 6 × 7.8 × 2 L 2 × 3.5	No 12 mo

R: right; L: left; CT: CT

### Case Reports

We present a retrospective review of 6 patients diagnosed with elastofibroma dorsi between July 2003 and February 2007 in the radiology department of the Hospital Marqués de Valdecilla, Santander, Spain. The images were independently reevaluated by 4 radiologists (MAE, RL, JI, and EG). All patients had undergone computerized tomography and 4 had also been given an ultrasound examination of the soft tissue. The clinical data were taken from their medical histories, and from telephone conversations with the patients. A pathology study was available in 1 case. The clinical characteristics and results of exploratory radiological examinations are summarized in the Table.

The most common presenting complaint was the presence of a subscapular tumor of firm or elastic consistency. Patients were referred to the dermatology or general surgery consultant with a clinical diagnosis of lipoma or soft tissue tumor (Figure 1). In 2 cases there was a history of regular manual labor. The lesions were of unknown cause, discovered by chance in examinations for other reasons, and, in 1 case, by the patient himself following an injury to the back. No family history of elastofibroma dorsi could be established and no other lesions suggestive of elastofibroma were discovered during examination.

The ultrasound revealed a mass not adhering to the scapula with a fibrillar and fasciculated appearance and hyperechoic striae parallel to the axis of the mass.

In the computerized tomography scan, the lesions were found on the inferior border of the scapula adjacent to the thoracic wall, with areas isodense with skeletal muscle and hypodense laminar areas with no remodeling of the adjacent bone (Figure 2). X-ray examination revealed bilateral involvement in 3 cases.



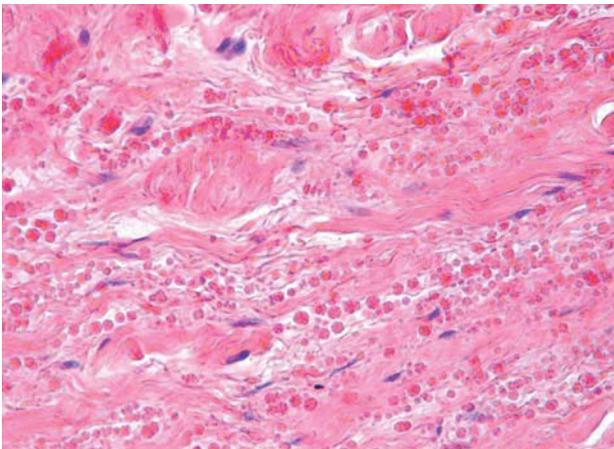
**Figure 1.** Right subscapular mass that becomes more evident when moving the arms forward.



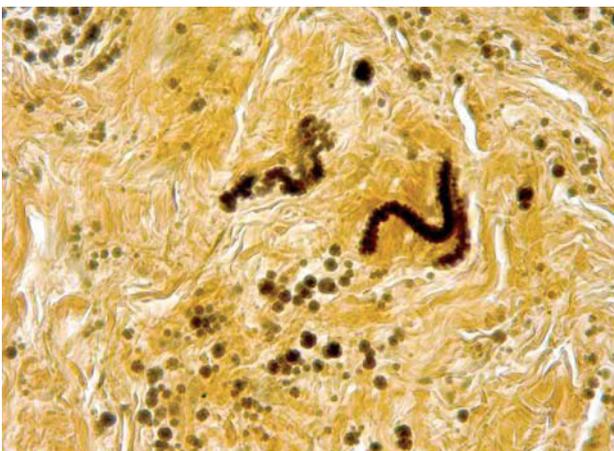
**Figure 2.** Computerized tomography axial section showing a bilateral subscapular tumor with areas of the same density as the muscle, and lower density areas corresponding to the characteristic fatty deposits of elastofibroma.



**Figure 3.** Elastofibroma. Poorly delimited tumor of a firm consistency, whitish-grey when cut with yellow zones and fibroadipose aspect.



**Figure 4.** Elastofibroma. Abundant thick fragmented elastic fibers and globules of elastin appearing refringent against a collagenous matrix (hematoxylin-eosin  $\times 200$ ).



**Figure 5.** Elastofibroma. Staining with the Verhoeff method ( $\times 200$ ) for elastic fibers reveals a jagged surface and a higher density core.

In 1 patient who underwent surgical intervention a poorly circumscribed whitish-yellow mass was obtained, measuring 8 cm in diameter (Figure 3). Histological studies showed the tumor to be composed of hypocellular connective tissue with broad collagen banding, isomorphic fibroblasts, and few adipocytes. Abundant elastic fibers and elastin globules of an acidophilic, appearing bright and refringent, were observed at the center of the lesion (Figure 4). Staining for elastin revealed numerous thick fragmented fibers and pinkish elastin globules (Figure 5). The fibers frequently presented a jagged or fragmented surface on a denser central core.

## Discussion

Elastofibroma is a benign lesion of the connective tissue also known as elastofibroma dorsi due to the fact that 99% of cases are found in the scapular region, close to the inferior angle of the scapula, between the serratus, rhomboid major, and latissimus dorsi muscles. It has occasionally been described in other locations like the deltoids, the greater trochanter, olecranon, ischiatic tuberosity, thoracic wall, foot, tricuspid valve, arm pit, and eye socket. Histological changes similar to elastofibroma have been seen in the gastric mucosa of Japanese women with elastofibroma dorsi.<sup>9</sup> These are generally unilateral lesions with variable bilateral involvement. Rare cases of multiple<sup>6-10</sup> or ulcerated<sup>3</sup> lesions have been reported. Many cases of elastofibroma dorsi are asymptomatic and are quite often discovered incidentally in examinations for other causes and in autopsies. In fact, in post mortem examinations these were found in 24% of women and 11% of men aged over 55 years old, suggesting that the condition is far more common than was thought and that most cases are clinically silent.<sup>11</sup>

Clinical differential diagnosis is required with other tumors of the soft tissue—like lipoma, sarcoma, and hemangioma. In radiological examinations, it must also be distinguished from other lesions that give a similar intensity of signal to skeletal muscle, and even hypocellular lesions with abundant collagen—like extra-abdominal desmoid tumor, cicatricial fibroma, or fibrous histiocytoma. The usual presenting complaint is an asymptomatic mass of elastic tissue in the subscapular region, covered with normal skin, which is frequently misdiagnosed as lipoma. Sometimes the patient notices changes in the tumor with arm movements or a snapping of the scapula, both of which can guide diagnosis. Difficulty in moving the shoulder or the appearance of pain is uncommon.

The etiology is unknown and it has been suggested this may be a reactive, degenerative, or neoplastic process. Hereditary and constitutional influences are clear, with descriptions of familial cases and a greater frequency amongst Japanese patients. Nagamine et al<sup>10</sup> published the largest

series with 170 patients from the Okinawa region of southern Japan.

In the histological study a stroma of rudimentary connective tissue can be observed, similar to a keloid, with a component of abnormal elastic fibers mixed with areas of fat. These elastic fibers typically display scalloped edges and are fragmented in discs or globules, mimicking immature elastic fibers in the ultrastructural studies. These fibers are characteristic of elastofibroma, and their origin has been attributed to either the degeneration of collagen fibers, degenerative changes in the elastic fibers, or the synthesis of abnormal elastic fibers. Although the observation of clonal abnormalities suggests a neoplastic rather than reactive origin<sup>12,13</sup> the preferred location in areas of microtrauma due to friction between the scapula and the thoracic wall implies a reactive mechanism. The greater incidence of elastofibroma dorsi on the right side of manual workers would support this hypothesis.

Ultrastructural studies and fluorescent in situ hybridization have led to proposals that elastofibroma dorsi originates in periosteal cells that initiate abnormal elastogenesis where chronic irritation occurs.<sup>14</sup> Immunohistological studies produce staining patterns of the cellular component of elastofibroma that prove positive for vimentin and CD34 and negative for smooth muscle cell actin and desmin, suggesting a fibroblastic origin.<sup>13,15</sup>

This characteristic histological structure correlates with some defined radiological patterns. In ultrasound examinations, elastofibroma dorsi appears to have a similar fibrillar pattern to striated muscle, although less well organized and defined. Regular alternation of hyperechoic and hypoechoic striates is seen, with hyperechoic stromal areas similar to a keloid and hypoechoic fatty areas.<sup>7</sup> However, in some cases ultrasound can fail to identify the extent of the mass due to difficulties in distinguishing elastofibroma dorsi from surrounding soft tissue, rendering complementary examinations necessary. Computerized tomography and, above all, magnetic resonance imaging are the most used noninvasive diagnostic techniques. Elastofibroma dorsi appears as a nonencapsulated mass with a similar density to skeletal muscle, mixed with hypodense bands corresponding to mature adipose tissue.<sup>16</sup> Small lesions can be difficult to view, but they stand out in magnetic resonance imaging when gadolinium is administered.

Treatment is surgical and is only indicated when comprehensive symptoms, pain, asymmetry of the thoracic wall, or restricted movement are present. Surgery may also be requested by the patient—as was the case with 1 of our treatment group. Recurrence is rare.

In summary, we believe that elastofibroma dorsi is rarely recognized in clinical examination and that dermatologists should consider the entity in differential diagnosis with tumors of the soft tissue of the scapular area. For older

patients with lesions in the classic location, radiological, computerized tomography, or magnetic resonance imaging findings are very suggestive of elastofibroma dorsi, thereby avoiding more aggressive forms of diagnosis or unnecessary interventions.

### Conflicts of Interest

The authors declare no conflicts of interest.

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