Our Dermatology Online

Case Report

Adventitious discovery of elastofibroma dorsi on skin biopsy

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ABSTRACT

Elastofibroma dorsi is a rare soft tissue pseudotumor, slow-growing, sitting in 99% of cases at the subscapular region and occurring in the elderly active people. Its pathogenesis is unclear. It is often asymptomatic. However, the diagnosis can be made on the typical topography of the mass and its characteristic appearance on CT and MRI. Thus, in the literature, most of the reported cases were radiologically discovered. An incidental histological discovery, like in our case is rare. We report the case of a 63 year-old man who had multiple nodular lesions, well circumscribed, firm, sometimes inflammatory, measuring between 3 and 5 cm, and located on the thighs and the paravertebral and scapular regions. The chest x-ray showed a right basal opacity suggesting a malignant processus. These nodules were biopsied in search of cutaneous metastasis of a probable pulmonary neoplasia. At histological examination, the diagnosis of elastofibroma was retained. Despite its rarity, the dorsal elastofibroma deserves to be known, thus avoiding excessive surgery. We propose to study its clinical, radiological and pathological features and its therapeutic modalities.

Key words: elastofibroma; dorsal; histology

INTRODUCTION

Elastofibroma (elastofibroma dorsi) is a relatively rare and slowly growing pseudotumor of the soft tissue. It is usually located at the inferior subscapular region, between the lower pole of the scapula and the chest wall. Other localizations are possible but remain rare. It is more frequent in old individuals with a predilection for women. Generally, elastofibromas are unilateral and asymptomatic. Multiple forms are rare. In most reported cases, this lesion was incidentally discovered by radiological examination. In our case, it was an incidental histological discovery.

CASE REPORT

A 63 year-old man consulted for fever, weight loss and impaired general condition. On physical examination, there were multiple nodular lesions, well circumscribed, firm, sometimes inflammatory, measuring between 3 and 5 cm and located on thighs, paravertebral and periscapular regions. The chest x-ray showed a right basal opacity suggesting a malignant processus. These nodules were biopsied in search of cutaneous metastasis of a probable pulmonary neoplasia. The periscapular nodules were biopsed, they corresponded to a very limited, non-encapsulated lesion, showing numerous eosinophilic elastic fibers which were fragmented, often globulous and scattered (Figs 1 - 3). The collagen fibers were thick. Rare fibroblasts and mononuclear inflammatory elements were found. The diagnosis of elastofibroma was retained.

The patient's informed consent was obtained.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

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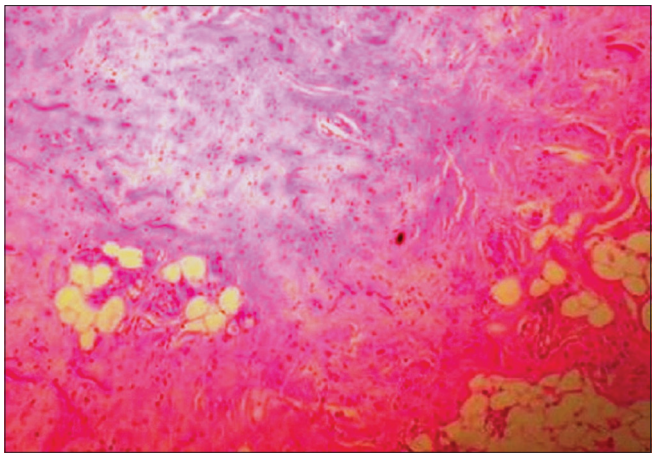

Figure 1: Fragmented and globulous elastic fibers within a hyalinized connective tissu. (HEx10)

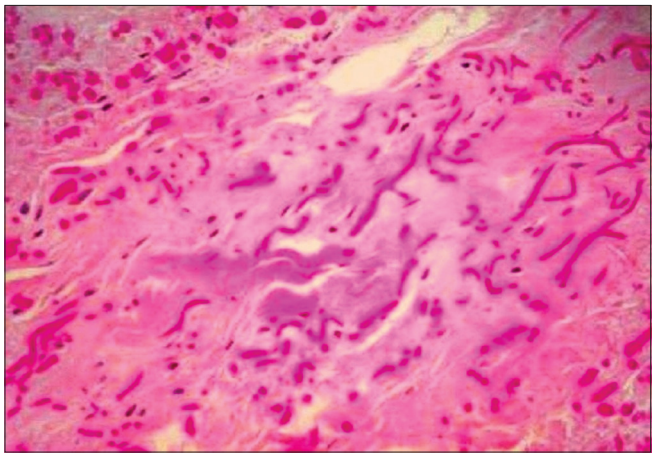

Figure 2: Fragmented, globulous and scattered elastic fibers intermixed with thick collagen fibers within a halinized stroma. (HE ×40)

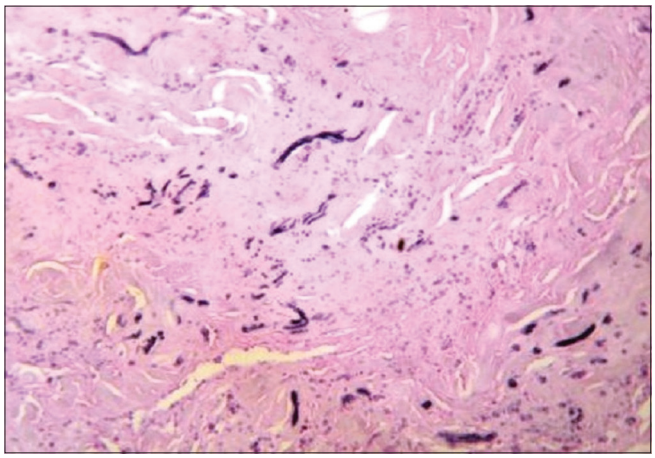

Figure 3: Orcein stain showing numerous fragmented and globular elasticfibers.

DISCUSSION

Elastofibroma dorsi was first described by Jarvi and Saxen in 1961 [1]. This uncommon benign and mesenchymal connective tissue lesion usually occurs in elderly people but it has been described in children under 6 years of age [2]. Women are more affected with a sex ratio M/F over 1 / 13 [3]. In our case, the patient was a man. The mean age at onset is 70 years.Elastofibroma's site of predilection is the subscapular region (99%), deep to serratus anterior, often attached to the periosteum of the ribs. Rarely, it is found in other locations such as extremities, head, abdominal and thoracic cavities, spinal canal and even cornea [3]. This lesion is often unilateral but it can be bilateral in 10 at 66% of cases occuring in the subscapular region [4]. Multiple elastofibromas were rarely described; Satoko Shimuzu, et al.. reported 17 distinct elastofibromas in a single patient [5]. The pathogenesis of this lesion is unclear although it is thought that mechanical microtrauma by heavy manual labor causes the friction of the scapula against the ribs and so causes this fibro-reactive lesion and this would explain the right- sided predominance. Genetic factors may also be involved. In fact, 32% of reported cases had a family history of elastofibroma. Actually, there is evidence of cytogenetic and molecular genetic changes in elastofibroma. Aberrations of the short arm of chromosome 1 and translocation involving chromosome 8 and 12 have been described [6].

Clinically, elastofibroma is often asymptomatic like in our case. However, patients can present with swelling, discomfort, snapping, clicking or clunking of the scapula and occasionally moderate pain. Subclinical elastofibromas have been found at autopsy. On physical examination, it presents as a well circumscribed and non adherent to the overlying skin mass. Otherwise, the diagnosis of elastofibroma can be made by both, histological or radiological examination.

Ultrasound examination, in the typical location of the elastofibroma, shows an abnormal mass of tissue with an alternating pattern of hyperechogenic and hypoechogenic lines that are roughly parallel to the chest wall. Computed tomography usually shows a heterogeneous soft tissue mass with poorly defined margins. MRI is the technique of choice and it reveals characteristic findings. Elastofibromas appear as poorly circumscribed soft tissue lesions with alternating areas of fibrous and fatty tissues. On T1-weighted and T2-weighted sequences, fibrous tissue produces low-intensity signals identical to that produced by muscular tissue, while the fatty tissue is seen as a high-intensity signal on T1- weighted sequences and as an intermediate signal on T2- weighted sequences.

The need of biopsy is controversial. Hayes, et al. [7] recommended it to confirm the diagnosis. Massengell

and al thought that biopsy is not necessary for diagnosis if clinical findings were typical and the MRI pattern was characteristic [8]. As cases of coexisting sarcoma and elastofibroma were reported, Alberghini, et al.. suggested a possible link between the two pathologic states so they consider that histological evaluation is essential [9]. In the recent literature, authors reserved histological confirmation to difficult and atypical cases.

Elastofibroma have typical macroscopic and histological aspects.

Macroscopically, it is ill-defined, gray white, rough-textured, measuring 5 to 10 cm. Sectioning reveals cystic degeneration and fat islets [10,11].

Histologically, elastofibromas present as non-encapsulated lesions which blend with the surrounding fat and connective tissue. The diagnosis is based on the presence of the altered elastic fibers embedded in a collagenous matrix, riddled with various amounts of fat cells. These elastic fibers are often fragmented into discs or globules and larger than regular ones. The fibers, which account for almost 50% of the tissue, stain black with the Verhoeff elastic stain. Some fibers are branched while others show a serrated edge.

Elastofibroma is stained positively with vimentine and CD34 but not with SMA, desmin, p53 and S100 [12]. These features indicate the fibroblastic nature of this tumor-like lesion.

The differential diagnosis is made with the other soft tissue tumors of the scapular region like lipomas, desmoid tumors, neurofibroma, cicatricial fibroma and sarcomas. Unlike elastofibromas, these tumors usually show strong enhancement after gadolinium injection.

Several treatment options have been discussed. It depends on whether or not there are symptoms. In fact, asymptomatic patients are simply observed while severely symptomatic people should have marginal excision which decreases recurrence risk. In some studies, radiotherapy can give good results [13]. Kransdorf, et al.. reported a rate of recurrence of 7% and attributed it to incomplete resection [14]. No malignant transformation has been mentioned [15].

CONCLUSION

In conclusion, elastofibroma dorsi is an under diagnosed lesion which should be considered in the differential diagnosis of the soft tissue tumors of the scapular region. Its diagnosis is easy when the clinical presentation and the radiological characteristics are typical. Recently, authors recommend biopsies only for atypical cases. In our knowledge, this is the first case of elastofibroma whose diagnosis was made incidentally on histological examination.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article and any accompanying images.

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