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Clinical Note

Elastofibroma Dorsi: An Uncommon and Under-Diagnosed Tumour

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ABSTRACT

Elastofibroma dorsi is a relatively rare soft-tissue tumor located at the infra-scapular level and/or subscapular regions. It usually occurs between the fourth and seventh decade of life, and is more common in females. We reviewed sixteen elastofibromas diagnosed in 12 patients (7 females, 58.3%). Four patients had bilateral elastofibromas. The most common symptom was pain. Presumptive diagnosis was made by physical examination. Chest ultrasound, computed tomography and/or magnetic resonance imaging were performed to confirm the diagnosis. Surgery was performed under general anesthesia. No major complications were observed. Elastofibromas are tumors of the chest wall with an uncertain impact. Surgical resection is indicated only in symptomatic patients.

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Fibroelastoma dorsi: un tumor infrecuente e infradiagnosticado

RESUMEN

El elastofibroma dorsi es un tumor poco frecuente de los tejidos blandos localizados a nivel infraescapular y/o subescapular. Su incidencia es variable, produciéndose entre la cuarta y la séptima década de la vida, y es más común en las mujeres. Se han revisado 16 casos de elastofibroma diagnosticados en 12 pacientes (7 mujeres, 58,3%), de las cuales 4 pacientes presentaron fibroelastomas bilaterales. El síntoma más frecuente fue el dolor. El diagnóstico se realizó mediante exploración física, tomografía computarizada y/o resonancia magnética nuclear para confirmar el diagnóstico en casos dudosos. La cirugía de exéresis se realizó bajo anestesia general sin observarse complicaciones mayores. El elastofibroma es un tumor de la pared torácica infrecuente y/o infradiagnosticado con un impacto incierto que requiere su exéresis solo en pacientes sintomáticos.

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Introduction

Elastofibroma dorsi is a relatively rare soft-tissue tumor. It is a hypocellular, non-encapsulated lesion with variable content of collagen, fat and elastic fibers. It was described for the first time by Jarvi and Saxen in 1961,¹ its incidence is variable and infrequent although its true rate of incidence may be greater than that described in the literature. The differential diagnosis should be done with subcutaneous tumors or lesions such as lipomas, fibrolipomas, cystic formations or more aggressive tumors. The objective of this review was to analyze our series of patients diagnosed with elastofibroma dorsi who were surgically treated in our center.

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Clinical Observation

In our hospital, we carried out a systematic retrospective review of 12 patients who were surgically treated between December 2004 and December 2009 by the Thoracic Surgery Department as well as the Orthopedic Surgery and Traumatology Department. All the patients were informed of the intervention and gave their consent before the intervention. A total of 16 elastofibromas were intervened in 12 patients, 7 of whom were women (58.3%) and 5 men (41.6%). Mean age was 53 (range: 44-74 years). Four (33.3%) patients presented bilateral lesion.

Six patients (37.5%) presented pain, while the rest only referred periscapular tumor. The diagnosis was made by physical examination, ultrasound study and computed tomography (CT) in all patients (fig. 1). Magnetic resonance imaging (MRI) confirmed the presumed diagnosis in 9 patients (75%). Most (n: 9; 56.25%) of

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Figure 1. Subscapular soft-tissue tumor at the level of the scapulothoracic joint compatible with elastofibroma.

the encapsulated lesions were located in the right subscapular region.

Surgical treatment was indicated when there were symptoms and/or the clinical radiological diagnosis was not conclusive for elastofibroma. In all cases, surgery was performed under general anesthesia with the patient in lateral decubitus position to allow for free movement of the upper ipsilateral limb. A longitudinal incision was made over the lesion; the tumor was resected along with the peritumoral peripheral fat, leaving macroscopic free margins. The tumors were located between the superficial and deep muscle planes of the torso, related with the rhomboid and latissimus dorsi muscles, at the infra-scapular and/or subscapular level. The maximum mean diameter of the lesions was 71 mm (range: 4-150 mm). No mortality was registered and only one patient presented seroma (with no infection of the wound). The patients were discharged between the 3rd and 7th day post-op, with a mean hospital stay of 4.2 days. The anatomopathologic study confirmed a hypocellular mass with irregular collagen tissue associated with elastic fibers compatible with elastofibroma.

Discussion

Elastofibroma dorsi is an uncommon soft-tissue tumor that mainly presents in middle-aged women. In the majority of the series reported, the location is predominantly on the right side; however, between 10-66% of the cases are bilateral or synchronic in other places. In our series, 4 patients had bilateral tumors.

Although it is considered a rare tumor, it is probably an underdiagnosed pathology.¹⁻⁴ The symptoms depend on the size and location of the lesion; it may present with pain or "cracking" at the scapular level, although the majority of cases are asymptomatic and/or the diagnosis is incidental during thoracotomy or post-mortem studies. Giebel et al,⁵ in a series of 100 autopsies, found 13 patients (10 men and 3 women) with elastofibroma dorsi.

Although the etiopathogeny is unknown, degeneration of the collagen fibers has been suggested as a possible cause. This degeneration can occur as the result of microtraumas on the scapulothoracic joint, which induces hyperproliferation of the elastic fibers. This lesion is considered reactive and not a true neoplasm.^{6,7} The vascular insufficiency and family predisposition have also been

postulated as possible etiological causes. In our series, we have not found family histories, although perhaps the size of the sample affects this.^{3,8} Histologically, the mass is hypocellular and contains a mix of collagen, elastic fibers and fat.

Diagnosis is based fundamentally on symptoms. On physical exploration, the lesion is usually well-circumscribed without being firmly adhered to the skin covering it, although with difficult delimitation regarding neighboring structures. This is important during exeresis in order to achieve some free margins. Normally, the tumor moves, becoming palpable and/or more painful with arm movements. Its most frequent location is anterior to the scapula over the rib plane between the sixth and eighth ribs, in a deep location with regards to the muscles of said region, mainly the serratus anterior muscle, the latissimus dorsi and the levator scapula muscle. Ultrasound, computed tomography and nuclear magnetic resonance are the most frequently used complementary examinations to confirm the diagnosis.

In our series, thoracic ultrasound and computed tomography were carried out in all patients showing evidence of homogenous attenuation lesions, similar to the adjacent neighboring structures. 9,10 When the clinical, ultrasound and CT data were sufficient to allow for a presumption diagnosis, no magnetic resonance was ordered, although MRI is the non-invasive diagnostic technique with greatest sensitivity and specificity. 10-12 Positron emission tomography (PET)-CT and/or biopsy are necessary if the lesion is not characteristic or the previously-described complementary examinations indicate signs of malignancy, with the aim of making the differential diagnosis with mesenchymal-type neoplasms, such as liposarcoma, fibrosarcoma, malignant fibrous histiocytoma or metastasis.

Although the definitive diagnosis can be done only with complete exeresis of the tumor and the malignancy cannot be ruled out without exeresis, we recommend resection in patients with symptomatic lesions or when the clinical-radiological findings are insufficient to confirm the diagnosis of elastofibroma. The post-op is short with limited morbidity and mortality, although there is a possibility of hematoma of the surgical bed, seroma of the wound and recurrence if the margins are not wide enough, which in some cases is difficult to determine.

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