ELASTOFIBROMA DORSI : CLINICAL AND PATHOLOGIC ASPECTS OF TWO CASES

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Elastofibroma dorsi is a benign entity that occurs most often in the subscapular area in elderly women. It has been a subject of controversy whether elastofibroma is a true neoplasm or a reactive fibrous lesion that produces abnormal elastic fibers. A biopsy should be performed to rule out sarcoma in all cases, but definitive treatment only requires simple surgical excision. We report two cases of elastofibroma and discuss the most relevant clinical and pathologic aspects.

Keywords : elastofibroma ; soft tissue tumors.
Mots-clés : élastofibrome ; tumeurs des tissus mous.

INTRODUCTION

Jarvi and Saxen (7) described in 1961 a benign chest wall tumor that was essentially formed by collagen and elastic tissues ; since all their five cases were located in the subscapular area, the entity was called elastofibroma dorsi. Whether elastofibroma is a true neoplasm or merely a reactive hyperplasic pseudotumor has been the subject of controversy. Nagamine *et al.* (11) in 1982 reported the largest series of elastofibromas in the literature (170 cases in Okinawa, Japan), which represented 65% of all cases reported until that time. We report two cases of elastofibroma dorsi, a lesion less frequent in Western countries, and review its most relevant clinical and pathologic aspects.

CASE REPORTS

Case 1 : A 45-year-old woman was admitted with a painless right subscapular mass, which had been increasing in size over the past two years. Physical examination showed a firm, well-circumscribed tumor ; when the patient was asked to abduct the arm, the mass became more evident. Chest wall xray was normal and ultrasonography showed a well-defined echogenic tumor, presumably a lipoma ; fine needle aspiration cytology confirmed the diagnosis. At operation, there was a fibrous mass that appeared to invade the surrounding tissues, including the periosteum of the scapula and the fifth to seventh ribs ; the mass was completely removed. There was no recurrence three years after surgery.

Case 2 : A 58-year-old woman developed over one year a painful, slowly-growing left subscapular mass. On physical examination, the tumor was not tender, had poorly-defined edges, and produced an annoying click with shoulder motion. At surgery, a white-colored fibrous mass was encountered below the scapula ; the tumor was widely adherent to the chest wall, but complete excision was achieved. Six years after the operation, the patient is free from recurrence.

Pathologic findings for both cases were similar. The tumors measured 6 and 12 centimeters respectively ; they were firm tan-yellow masses with ill-defined margins (fig. 1). On microscopic examination, both lesions consisted of a mixture of intertwining swollen eosinophilic collagen and elastic fibers, associated with fibroblasts and variously-sized aggregates of mature fat cells (fig. 2). On

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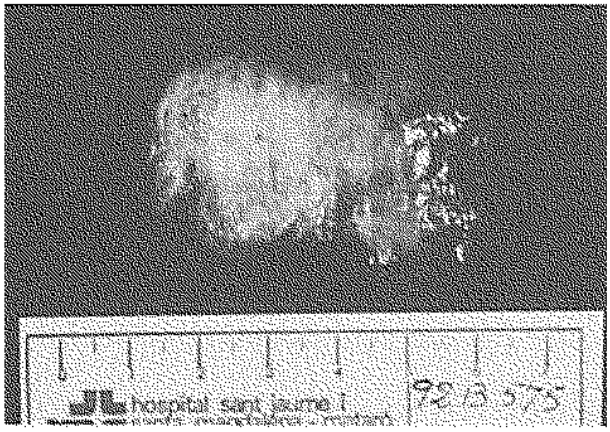

Fig. 1. — Gross appearance of one of the lesions. Note the irregular nonencapsulated margins of the mass.

Fig. 2. — Dense fibrous tissue interspersed with adipose tissue (Weigert's elastic stain × 160).

examination with the electron microscope, the elastic fibers appeared as a granular or fibrillary aggregation of electron-dense material along a central lucent core surrounded by an amorphous matrix containing scattered microfibrils, and the interspersed cells showed the characteristics of fibroblasts and myofibroblasts.

In both cases, the final pathologic diagnosis was elastofibroma dorsi.

DISCUSSION

Elastofibroma dorsi is usually found in an elderly population (2, 8, 11), and women are predominantly affected (2, 11). Jarvi *et al.* (6) detected an increased prevalence of elastofibroma dorsi in patients who perform manual labor, which may account for the right-sided preponderance (11). However, studies by Stemmerman and Stout (13), and Marin *et al.* (10) failed to find a correlation between muscular effort involving the shoulder girdle and elastofibroma formation ; besides, the occurrence within families and the multiplicity (11) suggest a possible nontraumatic cause. Most cases of elastofibroma reported up to now involved the subscapular region, although other sites have also been reported (10, 11).

Most patients affected by elastofibroma dorsi are asymptomatic because of the obscure location of the mass on the back and of its slow growth (2, 10, 11). Occasional complaints include stiff shoulders, local pain with arm movements, and an annoying click with shoulder motion (10, 11, 12). On physical examination, the soft tissue mass in the infrascapular area may be overlooked unless the patient is asked to elevate his arm laterally or anteriorly, allowing the scapula to slide over the lump (3). Chest radiographs may show an elevation of the scapula from the chest wall (3, 10), and both CT-scan and NMR may show specific appearances that differ from most other soft-tissue tumors, reflecting entrapped fat within a predominantly fibrous mass (9). In spite of this, incisional biopsy or excision is required to rule out fibromatosis, fibrolipoma or sarcoma.

Definitive diagnosis of elastofibroma dorsi depends on its characteristic histologic features as described earlier, and is based on the presence of fibers of elastinophilic material with degenerative appearance (4, 11) ; ultrastructural studies show a predominance of collagen fibers and dense granular bodies probably composed of elastin (4, 6). Elastinophilic fibers of elastofibroma are morphologically unique and diagnostic : recent studies (6) suggest that they are derived from an abnormal process of elastogenesis of elastic fibers, and exclude elastotic degeneration of collagen fibers (1, 13) or degenerative changes of the elastic fibers themselves (8).

The tumoral or reactive origin of elastofibroma remains unclear (2, 10), but it is undoubtedly a benign lesion. Therefore, asymptomatic patients may need no other treatment than biopsy to confirm elastofibroma histologically (11). Definitive treatment of elastofibroma is simple surgical excision, with an intact pseudocapsule and surrounding normal tissue ; as other authors have done before (10), we recommend excision under general anesthesia because dissection may be difficult owing to fixation of the tumor to the periosteum of the ribs and scapula. Recurrence of the tumor after complete excision has been reported in only one case (11).

REFERENCES

1. Barr J. R. Elastofibroma. Am. J. Pathol., 1966, 45, 679-683.
2. Bennet K. G., Organ C. H., Cook S., Pitha J. Bilateral elastofibroma dorsi. Surgery, 1988, 103, 605-607.
3. Berthoty D. P., Shulman H. S., Miller H. A. Elastofibroma : Chest wall pseudotumor. Radiology, 1986, 160, 341-342.
4. Dixon A. Y., Lee S. H. An ultrastructural study of elastofibromas. Human Pathol., 1980, 11, 257-262.
5. Fukuda Y., Miyake H., Masuda Y., Masugy Y. Histogenesis of unique elastinophilic fibers of elastofibroma : ultrastructural and immunohistochemical studies. Human Pathol., 1987, 19, 424-429.
6. Jarvi O. H., Saxen A. E. Subclinical elastofibromas in the scapular region in an autopsy series. Acta Pathol. Microbiol. Scand., 1975, 83, 87-108.
7. Jarvi O. H., Saxen A. E. Elastofibroma dorsi. Acta Pathol. Microbiol. Scand., 1961, 51, 83-84.
8. Jarvi O. H., Saxen A. E., Hopsu-Havu V. K. Elastofibroma : A degenerative pseudotumor. Cancer, 1969, 23, 42-69.
9. Kransdorf M. J., Meis J. M., Montgomery E. Elastofibroma : MR and CT appearence with radiologic-pathologic correlation. Am. J. Roentgenol., 1992, 159, 57-579.
10. Martin M. L., Perzin K. H., Markovitz A. M. Elastofibroma dorsi : benign chest wall tumor. J. Thor. Cardiovasc. Surg., 1989, 98, 234-238.
11. Nagamine N., Nohara Y., Ito E. Elastofibroma in Okinawa : A clinicopathologic study of 170 cases. Cancer, 1982, 50, 1974-1805.
12. Stemmerman G. N., Stout A. P. Elastofibroma dorsi. Am. J. Clin. Pathol., 1962, 37, 499-506.

SAMENVATTING

L. A. HIDALGO GRAU, J. ARDEVOL, T. SOLER, J. AULEDA, M. UBACH. Elastofibroma dorsi : Klinische en pathologische aspecten bij twee gevallen.

Elastofibroma dorsi is een benigne aandoening die meestal in de subscapulaire streek bij bejaarde vrouwen voorkomt. De aard van het gezwel blijft omstreden : echte neoplasie of reactioneel fibreus letsel dat aanleiding geeft tot abnormale elastische vezels. In elk geval moet er een biopt gebeuren om een sarcoom uit te sluiten. De definitieve behandeling bestaat uit een volledige en eenvoudige resectie. De auteurs rapporteren twee gevallen van elastofibroma dorsi en bespreken de voornaamste klinische en pathologische aspecten.

RÉSUMÉ

L. A. HIDALGO GRAU, J. ARDEVOL, T. SOLER, J. AULEDA, M. UBACH. Elastofibroma dorsi : aspects cliniques et anatomo-pathologiques. A propos de deux cas.

L'elastofibroma dorsi est une entité bénigne qui survient le plus souvent dans la région sous-scapulaire chez la femme âgée. Sa nature reste controversée : véritable néoplasie ou lésion fibreuse réactionelle qui produit des fibres élastiques anormales. Une biopsie doit être pratiquée dans tous les cas pour éliminer un sarcome, mais le traitement définitif requiert seulement une résection chirurgicale simple. Nous rapportons deux cas d'elastofibroma dorsi et discutons les principaux aspects cliniques et anatomo-pathologiques.