

## Elastofibroma dorsi: report of a case and review of the literature

### Elastofibroma dorsi: olgu sunumu ve literatürün gözden geçirilmesi

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### Summary

Elastofibroma is a tumorlike process that was first reported by Jarvi and Saxen in 1961. Typically this benign lesion involves almost exclusively the subscapular area of elderly individual and manifests as a slowly growing, solid, ill-defined mass of fibroelastic tissue. We report the case of a 73-year-old woman who was found to have a mass in the left scapula. Surgical excision of the mass revealed a well-delimited lesion 10 cm in largest diameter. Histologically, the mass was composed of mature fat alternating with sclerotic connective tissue, which also contained extensive eosinophilic deposits and abundant collagen. The elastic nature of these deposits was confirmed by elastic staining. Elastofibroma should be considered in the differential diagnosis of subscapular complaints of elderly patients. Cytogenetic and molecular studies are needed to distinguish elastofibroma whether a true neoplasm or a reactive lesion formed by repetitive minor trauma.

**Key Words :** *Elastofibroma, pseudotumoral lesion, subscapular*

### Özet

*Elastofibroma ilk kez 1961 yılında Jarvi ve Saxen tarafından rapor edilmiş tümör benzeri bir süreçtir. Tipik olarak yaşlılarda özellikle subskapuler bölgede görülen bu benign lezyon yavaş büyüyen, solid ve iyi sınırlı olmayan fibroelastik dokudan oluşmaktadır. Bu raporda sol skapulasında kitle saptanan 73 yaşında bir kadın hastayı sunuyoruz. Kitlenin cerrahi eksizyonu 10 cm çapında iyi sınırlı bir kitle göstermiştir. Histolojik olarak kitle matür yağ, sklerotik konnektif doku ve aynı zamanda yaygın eozinofilik depozitler ve bol kollajenden oluşmaktadır. Bu depozitlerin elastik boyaması ile gösterilmiştir. Elastofibroma, subskapuler şikâyetleri olan yaşlı hastalarda düşünülmelidir. Ayrıca gerçek bir neoplazi ya da minör travmaya bağlı reaktif bir lezyon ayrımında sitogenetik ve moleküler çalışmalar gerekmektedir.*

**Anahtar Sözcükler :** *Elastofibroma, psödötümöral lezyon, subskapular*

### Introduction

Elastofibroma dorsi is a slowly growing ill-defined mass of fibroelastic tissue that are generally considered to be reactive rather than neoplastic (1,2, Table-1). It occurs almost exclusively in the elderly and is thought to result from mechanical friction.

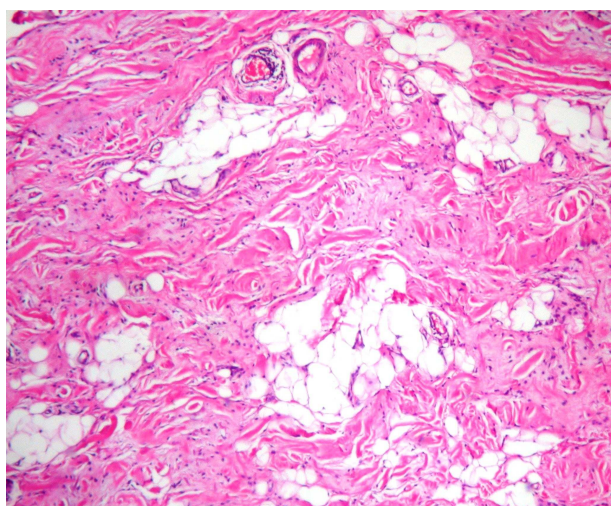
Whether elastofibromas are true neoplasms or merely reactive pseudotumors has been the subject of the controversy. It is important to recognize this benign lesion to avoid an unnecessary radical operation.

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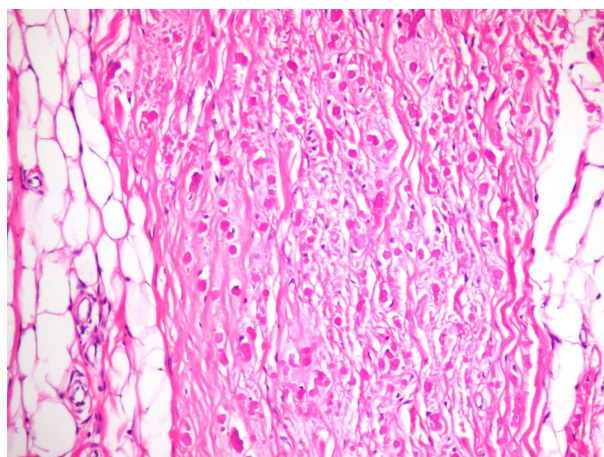
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**Table 1.** Various proposed pathogenetic origins of elastofibroma in the literature

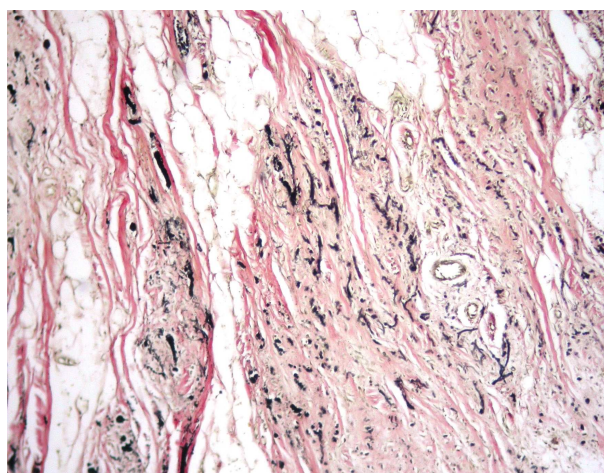
1961	Jarvi et al	Abnormal elastogenesis
1962	Stemmerman et al	Elastic degeneration of collagen fibers
1969	Jarvi et al	Degeneration of elastic fibers
1969	Winkelman et al	Elastodysplasia, overproduction of elastic matrix
1986	Nakamura et al	New production of elastin by fibroblasts
1987	Fukuda et al	Abnormal elastogenesis, enzymatic defect
1995	De Nictolis et al	Tumoral elastogenesis
2001	Batstone et al	Abnormal elastogenesis secondary to mechanical friction



**Fig 1.** Elastofibroma composed of a mixture of collagenized fibers and elastic tissue with intermixed mature fat (H&E; original magnification x40, x100 respectively).



**Fig 2.** Elastofibroma composed of a mixture of collagenized fibers and elastic tissue with intermixed mature fat (H&E; original magnification x40, x100 respectively).



**Fig 3.** Branched and unbranched elastic fibers and numerous globe like, eosinophilic deposits (Verhoeff-van Gieson stain; original magnification x100).

### Case

A 73-year-old woman was presented with a painless swelling that she had noticed near the lower end of her left scapula for about 6 months. Shoulder function was normal and painless. She had no family history of similar tumors. A chest radiograph and routine laboratory investigations were normal. She underwent a surgery under general anesthetic.

On macroscopic examination, the mass was unencapsulated and measured 10 cm in greatest diameter and had a smooth, bosselated surface. On section, it was solid, firm showing streaks of white tissue irregularly alternating with areas of yellow tissue.

Histologically, the mass was consisted of a mixture of intertwining swollen, eosinophilic collagen and elastic fibers associated with degenerated beaded appearance or fragmented into small globules and variously sized aggregates of mature fat cells (Fig-1-2). Elastic stain (Verhoeff) revealed deeply staining, branched and unbranched fibers and numerous elongated or globe like, eosinophilic deposits (Fig-3).

The patient had an uneventful recovery and had no evidence of disease during the follow up for 5 years.

## Discussion

Elastofibroma dorsi is a benign, fibroproliferative soft tissue tumor composed of excessive collagen and abnormal elastic fibers. It is most commonly found located in the periscapular region of elderly women and may be related to the natural occurrence of fibroelastic tissue in this region (3).

Although elastofibromas usually occur in patients aged over 50 years old, recently a number of cases of young patients have been documented. Jarvi reported that elastofibromas are not very rare in an autopsy study (4). Because of their obscure location on the back and common asymptomatic clinical presentation, the exact incidence of this lesion is unknown.

Elastofibroma may arises bilaterally or unilaterally and the majority of patients are asymptomatic, but some have shoulder stiffness or pain. The bilaterality will virtually exclude the diagnosis of malignancy (3, 5). The lesions have a characteristic location, however some unusual locations have been reported including stomach, axillary region, inguinal region, intraspinal space, and orbital area (1,6).

Histologically, elastofibroma is characterized by numerous and diffusely scattered thick, deeply eosinophilic, homogenous elastic fibers, which can be linear or fragmented into globules, in a background of eosinophilic, collagenized fibrous tissue.

Real neoplastic nature of elastofibroma is unknown and no malignant changes have been reported in the literature. The lesion has been historically considered a reactive process because of its prevalence in patients who perform hard manual work, its slow growth with no tendency to recur, and its occasional occurrence on both sides of the body (1). It is accepted that the excessive scapulothoracic motion was important in formation of the lesion. Many case studies revealed that elastofibroma is not a true neoplasm but may be a reactive lesion and appears to be the result of excessive formation of colla-

gen and abnormal elastic fibers secondary to repeated injury (4, 7). Direct mechanical stress on elastic tissue may be an important cause of hypertrophy and secondary degeneration of elastic fibres, and also of diffuse increase of collagenous tissue.

Several studies have suggested other factors leading to the formation of elastofibroma, including degenerative changes in collagen (8), an enzymatic defect (9), and disturbance of elastic fibrillogenesis by periosteal-derived cells (10) and vascular insufficiency (11). In addition, a recently reported case showed four members of a family with bilateral elastofibroma of the scapula suggesting a genetic component (12). Our patient had no family history of elastofibroma.

On the other hand, the observation of chromosome instability and clonal abnormalities in elastofibroma suggests that this lesion may represent a neoplastic rather than a reactive process (13-15). Moreover, a systemic involvement is suggested by the finding of elastofibroma like changes in the wall of a gastric ulcer with bilateral subscapular elastofibromas (16). Recently, Hisaoka suggested that elastofibroma may represent clonal fibrous proliferation (17). Additional studies are needed to define the potential biological consequences of these abnormalities in elastofibroma.

Elastofibroma has ill-defined margins and is often confused with soft tissue tumors. Differential diagnosis includes lipoma, fibrolipoma, liposarcoma, malignant fibrous histiocytoma and extraabdominal fibromatosis (1,6,8,18). Although the clinical behavior and histologic appearance definitely distinguish the elastofibroma, all cases should undergo biopsy to rule out malignancy. Sarcomas are generally more cellular and contain more mitotic figures. Fibromatosis is frequently more cellular and usually recurs if not completely excised. And elastofibromas do not exhibit a proliferation of small vascular channels lined by prominent endothelial cells that was seen in reactive fibrous tissue. Elastic tissue stain (Verhoeff) will be helpful, because the most important clue to the correct diagnosis may be the appearance of thick irregular wavy fibers and globules scattered in the collagenous stroma. Also the radiological appearances on CT or MRI may strongly support the diagnosis (6).

The pathogenesis of these lesions remains controversial. Accurate diagnosis should be made preferably by biopsy and histopathologic evaluation before additional treatment is administered. Marginal resection is curative in patients with symptoms. Despite its low incidence, this pseudotumoral lesion should be known generally to differentiate it from malignant tumors and to avoid unnecessary wide surgery.

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