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Original Research Paper

Surgery

A RARE CASE OF :BILATERAL ELASTOFIBROMA -BENIGN SOFT TISSUE TUMOR

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ABSTRACT Elastofibroma is a rare benign, soft-tissue slow-growing tumor seen predominantly in elderly females. The most common location is the infrascapular region. These benign tumors require resection only in symptomatic cases. We present a case of elastofibroma in a 56-year-old female. She presented with gradually increasing soft-tissue swelling of 7 cm × 5 cm in the right inferior subscapular region and 6 cm × 5 cm in the left inferior subscapular region for the last 10 years. She underwent tru-cut biopsy and the histopathology was reported as elastofibroma. Microscopically, lesions are composed of paucicellular collagenous tissue and large numbers of elastic fibers. The elastic fibers are large, coarse, deeply eosinophilic and fragmented into small, linearly arranged globules or serrated discs with 'beads on string' appearance. Entrapped lobules of mature fat cells are seen. A definitive diagnosis helps to avoid unnecessary wide and radical resection.

KEYWORDS : Elastic fibers, elastofibroma, soft-tissue, Verhoeff's Stain,'beads on string'

INTRODUCTION

Elastofibroma is a rare benign soft-tissue tumor. It is a slow-growing tumor, predominantly seen in elderly females. It often has a bilateral location in the thoracic wall. The most common site is in the infrascapular region beneath the muscular tissue. In magnetic resonance imaging (MRI), a preferable noninvasive investigation, elastofibroma typically appears as a solitary, poorly circumscribed, heterogeneous, soft-tissue mass. In the absence of a reliable preoperative diagnostic investigation, histopathological examination of biopsy is necessary to confirm the diagnosis in cases of unilateral elastofibroma.

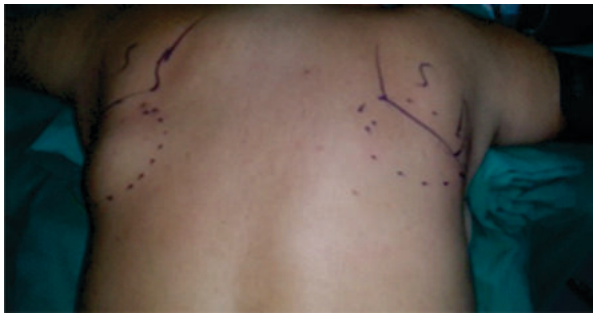


Image 1- Bilateral Elastofibroma With Marking

Case Report

A 56-year-old female presented with the complaint of diffuse soft-tissue swelling in the back. The swelling had been present since the last 10 years and was gradually increasing in size. Patient also complained of pain Which was spontaneous in origin ,dull aching in nature, intermittent (on and off) for the past 3 years,radiating to the right lateral chest ,aggravated by exercise and relieved on rest and medication. In recent months the lesion had increased in size and caused discomfort and even pain in response to exercise. Physical examination revealed bilateral masses that became manifest on abducting and flexing arms .Both lesions were grossly rounded, of rubber-like consistency, not adhering to the skin but adhesive to deep structures, measured 7 cm × 5 cm on right and 6cm × 5 cm on the left. The surface of the swelling was smooth. The routine hematological and biochemical examinations were within normal limits. Ultrasound reported , the lesions show inhomogeneous lesions in intramuscular planes of the medial aspect of the scapula on both sides.

The MRI reported ill-defined irregular marginated heterogeneous lesions in the posterior chest wall(deep to the serratus serratus anterior muscle.) in the infrascapular region on both sides.

The patient underwent a tru-cut biopsy which reported elastofibroma. No signs of malignancy were observed, particularly absence of bone invasion. The MRI data were compatible with elastofibroma dorsi.due to restricted and painful shoulder movements.

Surgical excision was performed under general anesthesia on a prone position and with abducted arms. An incision was made over the palpable masses and the ill-defined tumors were removed and sent for histo pathological examination. Histo pathology reported Macroscopically, the lesions were non encapsulated, with whitish fibrous tracts alternating with yellowish tissue of fatty consistency. The histological study on both sides revealed a lesion composed of paucicellular collagenous tissue and large numbers of elastic fibers. The elastic fibers are large, coarse, deeply eosinophilic and fragmented into small, linearly arranged globules or serrated discs with 'beads on string' appearance. Entrapped lobules of mature fat cells are seen. Necrosis, nuclear pleomorphism and mitosis are not seen. The postoperative course was without complications, and the patient was discharged 3 days after surgery.

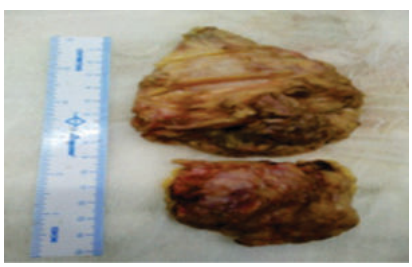


Image 2 The two excised masses

DISCUSSION

The term elastofibroma was first described by Jarvi and Saxon in 1961. It is a rare, benign slow growing soft-tissue tumor. But there is geographic variation and elastofibroma is common in the Japanese population.In Indian medical literature, the data regarding elastofibroma is scant. Though it has a marked predilection for elderly females, it is often reported in the age group ranging from 35-94 years. The typical location is the infrascapular region, occurring predominantly on the right side, but is bilateral in 30% of the cases. Majority of the patients are asymptomatic.Rarely, when symptoms are present, they are pain, scapular snapping, limitation of motion, clunking sensation in the shoulder adduction-abduction movement.Majority of the patients are asymptomatic.

The present case is a 56-year-old female who presented with a slow growing tender swelling in the both subscapular region. She complained of pain on movement for 3 years

Elastofibroma dorsi (ED) is used as a synonym and it is the name given to those lesions in the periscapular region. Histogenesis of ED is debatable. Some authors consider it to be a true neoplasm, whereas others presume it a result of reactive hyperproliferation of connective tissue exposed to repetitive minor trauma or friction between the lower scapula and the underlying chest wall. According to Nagamine et al., who reported a large series from Japan, genetic factors are implicated in the pathogenesis.However, our patient did not give any family history of similar infrascapular swelling.

The nature of elastic fibers is disputed. Theories suggest that they may be caused by abnormal elastogenesis or by degeneration as a secondary process, or may be by a combination of both processes.Plain radiographs do not show specific changes apart from a possible soft-tissue signal intensity or elevated scapula. In ultrasonography, the lesions show inhomogeneous lesions in intramuscular planes of the medial aspect of the scapula on both sides. MRI is the investigation of choice and shows ill-defined irregular marginated heterogeneous lesions in the posterior chest wall(deep to the serratus serratus anterior muscle.) in the infrascapular region on both sides. Computed tomography (CT) is less sensitive for visualizing the strands of fatty tissue. On MSCT scan of thorax the possibility of soft tissue mass-possibility of elastofibroma likely has been described. Due to its muscle-like appearance in nearly all the imaging procedures, diagnosis of elastofibroma remains challenging. In the present case, the lesion was located on both sides which is quite rare.

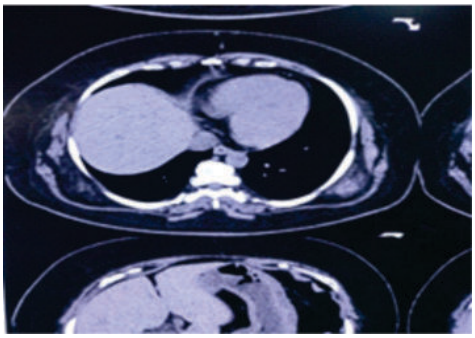


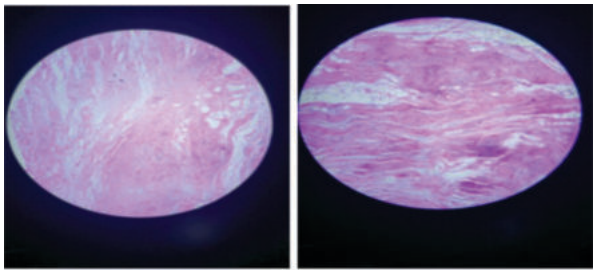
Image 2- MSCT image of bilateral elastofibroma (transversal view)

It is difficult to distinguish elastofibroma from other benign and malignant soft-tissue tumors of thoracic wall due its rarity. Preoperative procedures such as FNAC and frozen sections are not helpful. FNAC is not recommended as these lesions are hypocellular in nature and in frozen sections, the elastic fibers are difficult to detect.

In the absence of a definitive preoperative diagnosis, there is a chance of inappropriate treatment in the form of radical surgery. The clues for the presumptive diagnosis of elastofibroma are advanced age of the patients, the typical localization, female gender, or unilateral/bilateral manifestation.For a definitive diagnosis of elastofibroma, the role of biopsy is indisputable. A diagnostic biopsy helps to avoid unnecessary radical treatment. The differential diagnoses include various benign and malignant soft-tissue tumors such as nuchal fibroma, fibrolipoma, fibroma, spindle cell lipoma, and pleomorphic lipoma, aggressive fibromatosis, sarcoma, and subcutaneous metastasis.

However, the characteristic findings of elastofibroma such as the presence of elastic fibers in a collagenized fibrous tissue with entrapped adipose tissue helps in arriving at the correct diagnosis.

Complete surgical excision in symptomatic patients remains the treatment of choice. These tumors rarely recur and malignant transformation has not been described so far therefore, in asymptomatic lesions there is no need for excision. The role of diagnostic biopsy is to exclude malignancy and to reassure the asymptomatic patient that no radical surgical treatment is necessary. In this case the patient was symptomatic and hence,surgical excision has been done.



Left Side **Right Side**

CONCLUSION

Elastofibroma is an infrequent benign soft tissue tumor which frequently occurs in the subscapular region of elderly women. Although it is frequently asymptomatic, it can be responsible for discomfort and pain in arm mobilization. MRI is the gold standard examination for diagnosis. Biopsy is performed only in untypical elastofibroma dorsi to rule out a malignant tumor diagnosis. Surgical excision is the therapeutic option for symptomatic patients only. Pathological study confirms the diagnosis after surgery. Finally, our experience with this case report is in accordance with the literature.

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