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Elastofibroma Dorsi: An Uncommon Soft Tissue Tumor: Case Report

Elastofibroma Dorsi: Nadir Bir Yumuşak Doku Tümörü

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ABSTRACT Elastofibroma Dorsi (ED) is a benign, slow-growing lesion seen predominantly in elderly females. The tumor is non-encapsulated, hypo cellular in nature, composed of elastic fibers, fat and collagen. ED is asymptomatic in over 50 % of the cases and is incidentally discovered by imaging modalities. Differential diagnosis includes lipomas, fibro lipomas, hemangiomas and malignant tumors. ED is often overlooked or may be misinterpreted as a neoplastic process. The typical location of the tumor, long-term symptoms, and typical radiological findings, particularly MRI, are generally sufficient for ED diagnosis. We present a case of elastofibroma in a 66-year-old female. Elastofibroma dorsi can also show 18F-fluorodeoxyglucose (18F-FDG) uptake on positron emission tomography combined with computed tomography (PET/CT) imaging.

Keywords: Female; positron-emission tomography/computed tomography; soft tissue neoplasms

ÖZET Elastofibroma Dorsi (ED) genellikle yaşlı kadınlarda görülen, benign, yavaş büyüyen bir lezyondur. Tümör elastik lif, yağ ve kollajen dokudan zengin, kapsüllü olmayan, hücresel açıdan fakir bir yapıdadır. ED yaklaşık %50 olguda asemptomatiktir, görüntüleme tetkiklerinde rastlantısal olarak saptanır. Lipom, fibrolipom, hemangiom, malign tümörler ile ayırıcı tanıya girer. ED genellikle malign bir oluşum sanılabilir. Tipik tümör lokalizasyonu, uzun süren semptomlar, tipik radyolojik bulgular, özellikle MR görüntüleme ED tanısı için yeterlidir. Biz elastofibroma dorsi tanısı konulan 66 yaşında kadın hastayı sunuyoruz. Elastofibroma dorsi bilgisayarlı tomografi ile kombine pozitron emisyon tomografi (PET/BT) tetkikinde 18F-florodeoksiglukoz (18F-FDG) tutulumu gösterebilmektedir.

Anahtar Kelimeler: Kadın; pozitron-emisyon tomografi/bilgisayarlı tomografi; yumuşak doku neoplazileri

The term elastofibroma was first described by Jarvi and Saxon in 1961.[1] It is a benign, slow-growing lesion classically presenting as an ill-defined mass at the inferior pole of the scapula and the posterior chest wall, hence the term elastofibroma dorsi (ED). ED is frequently observed in middle-aged and older females. It is often bilateral. The etiology remains unclear and is a source of ongoing debate. Typically, symptoms may include the presence of a mass, pain, stiffness, scapular snapping and impingement like features. The tumor is non-encapsulated, hypo cellular in nature, composed of elastic fibers, fat and collagen.[2]

Differential diagnosis includes lipomas, fibro lipomas, hemangiomas and malignant tumors. Its radiological aspect has been documented with

ultrasonography (USG), computerized tomography (CT) and magnetic resonance imaging (MRI). ED is often overlooked or may be misinterpreted as a neoplastic process.

CASE REPORT

A 66-year-old female presented with back pain and swelling under the left scapula. These symptoms were presented since the last 4 years and stated progressive enlargement during the last 6 months. On examination, the mass was present in the left inferior subscapular region. It measured 2 cm×4 cm in size. The surface of the swelling was smooth. The routine hematological and biochemical examinations were within normal limits. The thoracic MRI was reported 25x56 mm mass in left anterior scapular muscles plans, significant enhancement and malignant potential.

On ^{18}F-FDG PET/CT examination this pathological mass SUVmax was 5.5 (Figure 1). Following this, she underwent surgical excision. Microscopically, the lesion was poorly circumscribed and showed an admixture of collagenous tissue with mature adipocytes. The elastic fibers were large, coarse, and deeply eosinophilic in nature. There was no area of necrosis or increased mitotic activity (Figure 2). The Van Giesen's elastic stain confirmed the elastic nature of the fibers (Figure 3). Based on the characteristic histopathological features and histochemical staining, the lesion was diagnosed as a case of elastofibroma. No further surgical complications occurred in postoperative period and no recurrence was observed during one- year follow up.

DISCUSSION

Elastofibroma dorsi is a rare clinical entity. Elastofibroma dorsi typically arises from the connective tissue between the chest wall and the lower portion of the scapula beneath the serratus anterior and the latissimus dorsi muscles in 99% of the cases but can also occur in extrascapular locations.[3] Other rare locations are, forefoot, hand, breast, stomach and mediastinum.

The literature about ED usually depends on case reports and small series. Brandser et al. found its incidence in normal population to be 2% in the CT screenings.[4] The rates of elastofibroma seen in the autopsy series were 11.2% in males and 24.4% in females older than 55 years.[5] The majority of the patients were housewives in the present series and 37.2% of the cases showed bilateral ED but only 19.6% of the cases underwent resection. The bilaterality rates was between 12- 73% in the clinical series of the literature.[6]

There are several scenarios to explain the pathogenesis of ED. Minor micro traumas due to friction between the inferior border of scapula and chest wall may cause reactive fibromatosis. Hypoperfusion by the enlargement of the lesion, enzyme deficiency, elastotic degeneration have also been discussed for pathogenesis.[7] 30% of the patients had ED history in their families suggest the genetic causes of the disease. Hernandez et al. found DNA sequence losses in 1p, 13p, 19p, and 22q chromosomes.[8] Imanishi et al. stated that transforming growth factor b and basic fibroblast growth factor may be effective in the

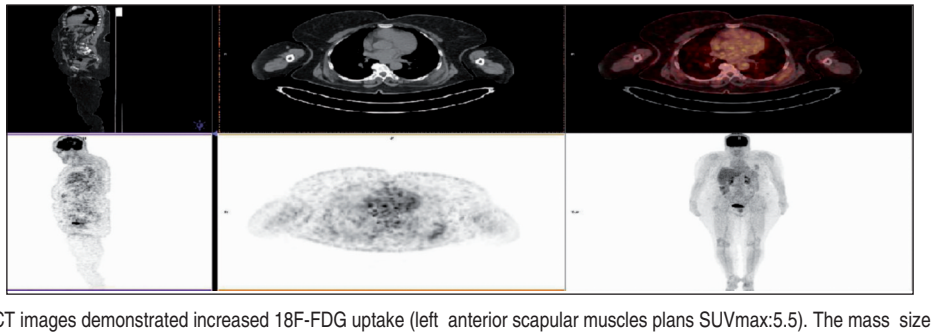

FIGURE 1: PET/CT images demonstrated increased 18F-FDG uptake (left anterior scapular muscles plans SUVmax:5.5). The mass size was 25x56 mm.

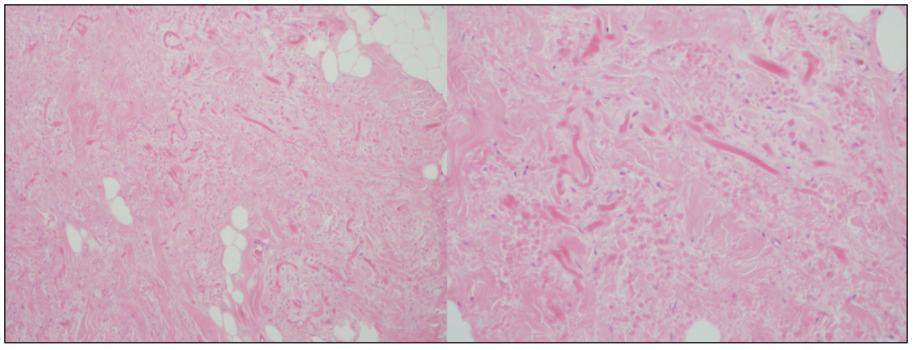

FIGURE 2: Lesion with admixture of collagen fibers and adipose tissue with characteristic elastic fibers (Hematoxylin and eosin 100× and 200x).

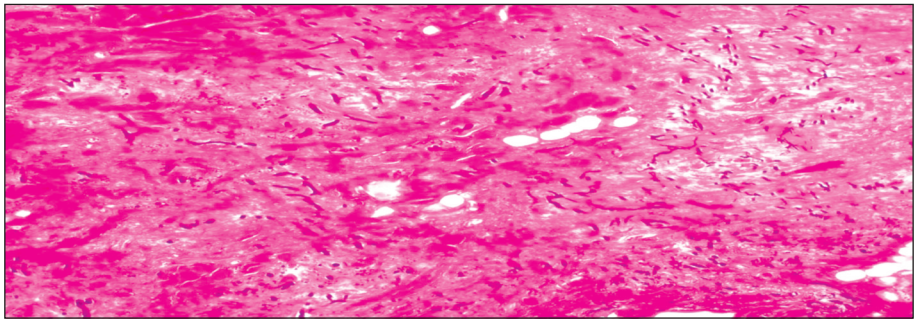

FIGURE 3: Van Giesen's elastic stain showing elastic fibers (Van Giesen's elastic stain 100x)

development of ED.[9] Taşlı et al. considered that the lesion originates from primitive dermal mesenchymal cells due to Factor XIIIa and CD34 positivity.[10]

Elastofibroma dorsi is asymptomatic in over 50% of the cases and is incidentally discovered by imaging modalities.[3]When symptoms are present, they are usually mild, consisting of a swelling, clunking of the scapula on moving the shoulder or moderate pain. Hammoumi et al. reported 93% pain, 51.3% functional limitation and 31% click or lump in a series of 76 cases.[11]

Although most cases can be easily diagnosed on the basis of USG, CT and MRI imaging characteristics, ED (especially in lung, breast and chest wall malignant tumors) can be misinterpreted as a neoplastic process, especially for lesion over 5 cm in size. The size of the lesion varies from a few centimeters to 15-20 cm. Malghem et al. described a nonencapsulated lesion at the typical location, between the fibrous tissue and the fat tissue that showed an alternating pattern in MRI.[12] In ultrasound a well-demarcated multilayer eco-signal is diagnostic. The studies conducted using PET/CT found that ED had a diffuse but slight ^{18}F-FDG involvement. The studies found the average SUV to be 2.29.[13]

The typical location of the tumor, long-term symptoms, and typical radiological findings, particularly MRI, are generally sufficient for ED diagnosis. Some authors recommend fine needle aspiration biopsy. There is no evidence about the malignant transformation of ED.

Generally accepted indications for resection are symptomatic cases with lesions bigger than 5 cm. Some authors suggested resection of all symptomatic and asymptomatic cases in order to confirm the diagnosis pathologically.[14] After resection of ED Nagamine et al. reported 0.06% recurrence in his series of 170 cases, and Lococo reported 4.5% recurrence in his series of 71 cases.[3,15]

Our patient was 66-years-old, had a unilateral mass on her left infrascapular border and it was

painful with shoulder movements. Our patient's lesion stated rapid growth during the last 6 months and had no family history. MRI and PET-CT images were present malignant potential. We preferred for surgical excision is curative in both diagnostic.

In conclusion, ED is a rare, slowly growing soft tissue tumor typically located in the scapulothoracic region. Surgical treatment is sufficient in symptomatic cases. Mild or moderate diffused uptake of ^{18}F-FDG is often observed in ED.

We present rare case of elastofibroma with its histopathological and PET-CT findings.

Conflict of Interest

Authors declared no conflict of interest or financial support.

Authorship Contributions

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