Elastofibroma dorsi: What's new?

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Abstract

Elastofibroma dorsi is a rare slow-growing soft tissues tumor. The lesion usually grows near the shoulder but could also involve other location. Pathogenesis of elastofibroma dorsi is still unknown and in the literature, there are mostly described case report or case series. The aim of our study is to summarize the recent innovation in the histology and immunoistochemical finding about elastofibroma and update the radiological algorithm of diagnosis.

Introduction

Elastofibroma dorsi (ED) was first described by Jarvi and Saxten,[1] it is a rare, slow-growing soft tissues tumor of mesenchimal origins with benign characteristics.

The lesion usually grows beneath the rhomboid major and latissimus dorsi muscles adjacent to the inferior angle of the scapula.[2] The location is so distinctive that some authors consider it almost patognomonic.[3] Others rare location sites of elastofibroma are ischial tuberosity, olecranon area, deltoid muscle, axilla, intraspinal space, greater omentum.[4] It usually is a unilateral lesion, but it can be bilateral in 25% of the case.[5]

Generally elastofibromas are asymptomatic tumors.[6] The clinical symptoms of elastofibroma are palpable mass with swelling, discomfort, functional restriction, occasional pain,[7] clunking of the scapula on moving the shoulder.[8]

Pathogenesis of ED is still unknown and matter of debate.[9,10] Findikcioglu et al believe that changes in the subscapular region and repeated microtrauma predispose to elastofibroma formation.[11] In fact ED is common among physical laborers and this could explain also the right sided dominance,[12] even if this might be related to the patient's dominant handedness.[13] Thus this correlation is still unclear.[14] Nakamura et al proposed that ED arises because of a disturbed fibirillogenesis, due to chronical irritation or trauma.[15] To support this, they demonstrated a minimal change in the aminoacidic composition of the abnormal elastin of ED.

Due to its symptomless being, ED is often incidentally discovered by computer tomography or MRI scan performed for other reasons.

The diagnosis of ED can be made by clinical and radiological findings. Usually needle biopsy or excision is not performed for this kind of lesion, though surgical excision in the option in case of symptomatic masses with a standard antibiotic prophylaxis for oncological patients.[16] Some authors also suggest to evaluate MR enhancement to decide whether proceed to biopsy and consequent tumour excision or not.[17] The aim of this study is to clarify the new findings in histological, immunohistochemical and radiological findings for ED and better understand its complicated pathogenesis.

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Histology

Histological section of elastofibroma shows a mixture of collagen, elastic fibers and adipose tissue together with some blood vessels, rich amorphous extracellular matrix and mature fat cells.[12,18]

Macroscopically, ED present a non-capsulated whitish mass with yellowish foci and elastic consistence.[4,19]

Microscopically it is composed by dense collagen bundels and abundant abnormal elastic fibers.[15,18]

Immunoistochemical findings

Immunohistochemical studies showed positive Cd34 fibroblasts, both in the cytoplasm and in the space between the cells and collagen fibers.[9,19] These cells may produce both abnormal elastic fibers and collagen fibers as they normally have a central role is the regeneration of tendon and ligaments.[19-20] Kakudo and Hemmi found positivity also for vimentin.[9,21] Factor XIIIa has been identified among stromal cells markers;[22] it is a necessary factor in the coagulation pathway that stabilizes clot formation by cross linking fibronectin to collagen.[18]

Kuroda et al also identified TGF-B in the fibroblastic cell cytoplasm, suggesting that this could be the reason why fibroblast may produce collagen and abnormal elastic fibers.[19]

Harigopal et al and Yamazaki et al found the presence of lysozyme.[12,18] This enzyme has been shown to prevent elastic fibers degeneration and is a marker of elastic fiber damage.[23]

Some authors describe the presence of myofibroblasts while other authors deny their presence. [19, 24]

Di Vito et al found periostin and tenascin-c positive stromal cells.[25] Most of them are concentrated more intensively around peripheral vessels. These two proteins seem to be involved in fibrosis pathogenesis, also in other body districts. Di Vito et al also identified mast cells tryptase-positive abundant throughout the lesion.[25] These cells play a role in inflammatory and auto-immune diseases and seem to have a regulatory role on cell differentiation.[26]

Ultrastructural findings

Most of authors agree that ED is mostly made of abnormal globular elastic fibers, and that most of them are distributed randomly.[12] The electron microscopy shows how ED cells are rich in endoplasmatic reticulum and in vimentin stromal production.[27]

Radiological findings

Nowadays the most used imaging technique are: ultrasound (US), computed tomography (CT), magnetic resonance (MR).

US investigation is widely accepted as first line imaging modality. Elastofibroma appears as deep-seated pseudonodular area with poorly defined borders, fasciculated structures with hypo and hyperechogeneous striae of variable thickness, within few millimetres, parallel to chest wall similar the ones of muscular tissue but coarser and slightly more disorganized in comparison are the main characteristic of the lesion. The Color and power Doppler show vascularization patterns similar to surrounding muscle, intravenous contrast agents, when used, do not shows sign of abnormal vascularization.[28]

The limitation of US is represented by patient body build and the possible underestimation of its real size.

Some authors also use elastosonography to identify the malignancy pattern of soft-tissue tumors, but this kind of technique has not been applied to ED yet.[29]

X ray shows only indirect signs of ED presence like raising of scapula of affected side on chest x-ray and enlarged scapula thoracic space.[10]

On CT scan ED appears as poorly defined, heterogeneous, lentiform shaped, soft tissue mass, with attenuation similar to that of skeletal muscle and linear streaks of decreased attenuation suggesting accumulation of adipose tissue are constantly present within the lesion.[10] No bone abnormalities are detected.

MR has a higher diagnostic confidence compared to US and CT, on MR imaging ED appears as a semilunar, soft tissue mass abutting the rib cage posteriorly with well-defined margins. The mass shows low signal intensities which reflect fibrous tissue and is similar to normal muscle on both T1 weighted and T2 weighted sequences. In T2FS or STIR the lesions show partial or regional high signal intensity due to a varying degree of layered fat streaks.[7] Underlying bone are intact and there is no evidence of chest wall infiltration.[5] Usually no peritumoral edema is present.

Diffusion Weighted magnetic resonance imaging DWI represents a promising new diagnostic tool for ED providing functional information to define the diagnosis as benign.[10]

Contrast administration usually is not necessary to characterize the lesion when typical findings are present at MR imaging, as contrast enhancement is fairly variable, ranging from a mild to avoid enhancement.[2]

Fludeoxyglucose (FDG-PET) is not actually used to diagnose ED but to evaluate painful subscapular mass when suspect of metastasis in oncologic patients,[11] more often ED is incidentally detected during cancer staging, mimicking malignancy it could interfere with the correct diagnosis and patient management.[30] ED in the frontal MIP projection is represented by an oval image with moderate or low intensity uptake of FDG at the unilateral or bilateral pectoral level.

18F-FDG uptake patterns remains stable after chemoradiotherapy and a stable, slow growing aspect which may help to discriminate malignant lesions.[8]

The mechanism of uptake is uncertain but may reflect a combination of high vascularity and increased metabolic avidity within the mass. Although is not necessary to perform PET-CT imaging to obtain diagnosis of ED, it remains important to detect ED to avoid misdiagnosis especially in lung breast and chest wall malignant tumors.

ED should be distinguished from other soft tissue masses as sarcomas, lipomas, hemangiomas, myositis ossificans, giant cell tumors or peripheral nerve sheath tumors.[31,32] Imaging studies combined with its clinical futures such as typical location of the tumor and its growth pattern are sufficient to allow the definitive diagnosis and differentiate ED from other lesions.

Conclusions

ED is a tumor of still uncertain pathogenesis. Histology and immunoistochemical findings are deepening our knowledges about this tumour; the combined processes of local microtrauma and elastin degeneration could have a central role in the pathogenesis of this tumour. Modern nuclear medicine techniques, such as PET-TC, give us further information but the diagnosis is still based on clinical symptoms and MRI findings.

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