



ELASTOFIBROMA DORSI: A RARE TUMOR PRESENTING AT AN UNUSUAL AGE WITH EMPHASIS ON IN AND OUT PHASE MR SEQUENCES AS AN ADJUNCT TO CONVENTIONAL MRI.

Radiology

Dr. Saurabh Gupta	(M.D.) Assistant Professor, Department of Radiology, Dayanand Medical College and Hospital, Ludhiana, Punjab, India.
Dr. Jatinder Pal Singh*	(FRCR, DMRD, DNB), Associate Director, Department of Radiology, Medanta- The Medicity, Gurgram, Haryana, India. *Corresponding Author
Dr. Aneet	Assistant Professor, Department of Anaesthesiology, Dayanand Medical College and Hospital, Ludhiana, Punjab, India

ABSTRACT

Elastofibroma dorsi is a rare, slow growing benign proliferation of fibro fatty tissue seen characteristically in subscapular location in elderly patients. Imaging characteristics of this tumor in elderly individuals in different modalities have been described in literature. We present imaging findings of this rare soft tissue lesion in a young male adult of 19 years age, who presented with a symptomatic unilateral painful lump in left infrascapular location, with emphasis on the role of in and out phase MRI imaging in diagnosing such lesion.

KEYWORDS

Elastofibroma Dorsi; in and out phase MRI sequence; young male; rare tumor

INTRODUCTION

Elastofibroma dorsi with its highly characteristic location is a rare, slow growing, benign proliferation of fibro fatty tissue that is most commonly seen in elderly population. It was first diagnosed in 1958 and then reported first by pathologists Jarvi and Saxn in 1961 [1]. The characteristic Ultrasound, Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) findings in this tumor have already been described in elderly patients [2-4]. We present imaging findings of this rare soft tissue lesion in a young male adult of 19 years age, who presented with a symptomatic unilateral painful lump in left infrascapular location, with emphasis on the utility of in and out phase MRI imaging in diagnosing such lesion, never described in literature before.

CASE REPORT

A 19-year-old young male with a painful lump in left infrascapular region (Fig. 1) and clunking of the scapula on shoulder abduction was referred to sarcoma surgeon at our institute for further management of the lesion. He had undergone MRI and Fine Needle Aspiration Cytology (FNAC) outside our institute prior to the referral and was diagnosed as infiltrating intramuscular lipoma. No fever, weight loss or other significant medical history was present. Clinical examination revealed a mass, which was rubbery in consistency, with regular margin, and adherent to the chest wall, but not to the skin. No features of inflammation were noted. The laboratory parameters were unremarkable. The clinical differential diagnosis included lipoma or primary soft tissue sarcoma. Chest radiograph (Fig. 2a) showed soft tissue opacity in left infrascapular region. A non-contrast CT scan (Fig. 2b) of the chest, requested by the referring clinician at our institute and a complimentary ultrasound (Fig. 2c) was performed and an imaging diagnosis of elastofibroma dorsi was made. Considering the young age of the patient, a CT guided biopsy (Fig. 2d) was performed and histopathology revealed collagen fibers with interposed adipose tissue and interspersed elastic fibers in between the collagen bundles. Immunohistochemistry study was also performed on given sample. The histopathology findings (Fig. 3) were compatible with the diagnosis of elastofibroma dorsi. Since the patient was symptomatic with the lesion, therefore surgical excision was planned. However, prior to surgery a contrast enhanced MRI (Fig. 4) was performed in order to better delineate the lesion margins for surgical planning. In addition to routine MR sequences in and out phase MR imaging was also performed (Fig. 5). Subsequently the patient underwent surgical excision of the lesion at our institute and the histopathology of the surgical specimen also confirmed the diagnosis of elastofibroma dorsi.

DISCUSSION

Elastofibroma dorsi is nonencapsulated lesion and essentially always occurs in the subscapular area, anterior to the serratus anterior but rarely is found in other sites such as greater trochanter, ischial tuberosities, olecranon, hands, deltoid muscle, axillae, foot, inguinal region, orbits, stomach, greater omentum and intraspinal spaces [5-8].

Elastofibroma dorsi is usually a disease of elderly patients. The

average age range has been reported to be between 49 and 71 years. It was originally described at the 12th Congress of Scandinavian Pathologists by Jarvi and Saxn in elderly patients in 1959, and subsequently reported in 1961 and 1969 [1, 9]. Nagamine et al in 1982 reported a study of 170 cases in Okinawa and found the average age at the time of diagnosis was 70 years and of 170 only 3 patient were less than 25 years of age, however no cross sectional imaging features have discussed in this age group [5]. Go et al reviewed the literature from 1980 to 2009 and found 330 cases of elastofibroma with mean age at presentation being 62 years [10].

It is thought to result from mechanical friction between the scapula and chest wall, hence is considered reactive rather than neoplastic [4]. The WHO defines elastofibroma as a benign, ill-defined proliferation of elastofibrous tissue characterized by an excessive number of abnormal elastic fibers.

Elastofibroma dorsi histologically is made up of fibrous tissue with internal linear fatty streaks [11, 12]. The imaging characteristics correlate with the tissue composition of the tumor. At ultrasonography, they demonstrate multilayered linear alternating hypoechoic fat tissue and echogenic fibroelastic tissue, which has been described in pathologically proven elastofibromas [2]. On CT, they are seen as poorly defined soft-tissue mass with attenuation similar to that of the adjacent skeletal muscle and internal striations of fat attenuation. However, smaller elastofibromas may also have homogeneous attenuation at CT without visible internal foci of decreased attenuation [4]. MR imaging features also correspond to the tissue characteristics with the majority of the mass being isointense to the skeletal muscle, as the fibrous component is isointense to muscle, with interspersed linear or curvilinear increased signal intensity due to fat component on T1 weighted images. On post contrast images these show heterogeneous enhancement with enhancing and non-enhancing areas [4].

The differential diagnosis of such periscapular lesions, with imaging characteristics similar to that of skeletal muscle on CT and MR imaging, includes lesions with decreased cellularity and abundant collagen such as extraabdominal desmoid, neurofibroma, cicatricial fibroma and malignant fibrous histiocytoma. The treatment of elastofibroma dorsi depends on the patient symptoms. Surgical excision is sufficient to cure symptomatic individuals. In asymptomatic individuals the lesions can be left as such after malignancy is ruled out on histopathology.

Our case showed many of the classic findings of elastofibromas- sub or infrascapular location, composed of soft tissue with attenuation and signal intensity on CT and MR imaging resembling those of skeletal muscle interwoven with strands of fat with heterogeneous enhancement following gadolinium administration. Lack of intermuscular fatty tissue made the identification of lesion difficult and effacement of fat planes suggested presence of an underlying lesion. In and out phase sequence used to study the characteristics of this fibro fatty tumor showed India ink artifact near the interface on out phase

imaging, confirming the fatty content of the lesion. This could be of value in diagnosis of smaller lesions that do not show characteristic imaging characters on conventional MR sequences and are difficult to identify due to less intrinsic and extrinsic fatty tissue in younger age group patients.

To conclude, elastofibroma dorsi should be considered as clinical diagnosis for patients presenting with infrascapular lesion in all age groups. In and out phase MR imaging can be used to complement the usual conventional MR sequences in smaller lesions with less fatty content.

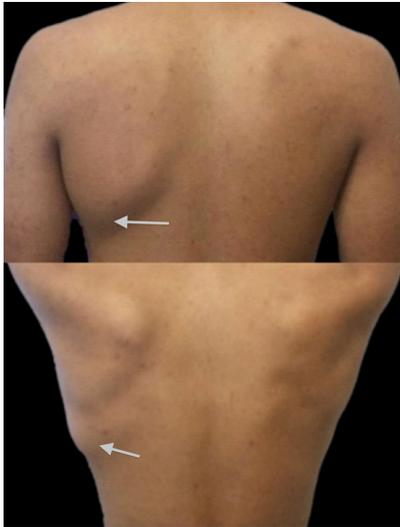


Fig. 1: Pictures of a 19-year-old young male reveals an infrascapular mass lesion (Grey arrow), which becomes more prominent on shoulder abduction.

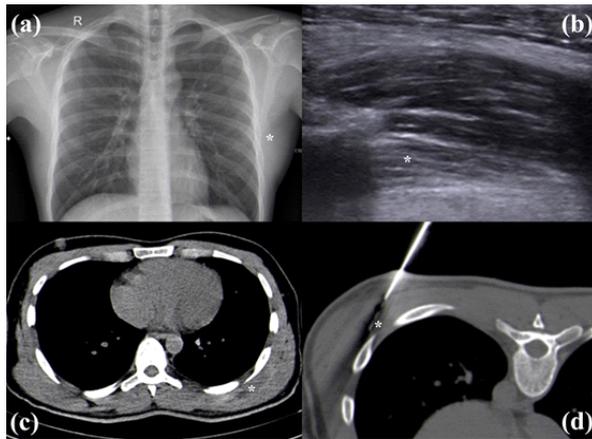


Fig. 2: Elastofibroma Dorsi (asterix) of left infrascapular region seen on different imaging modalities: (a) Chest radiograph of the patient shows soft tissue prominence. (b) High resolution ultrasound shows alternating hypoechoic fat tissue and echogenic fibroelastic tissue. (c) Non contrast CT scan of the patient reveals a sheet of soft tissue thickening deep to the serratus anterior and latissimus dorsi muscles, along with thickening of the fascia overlying the serratus anterior muscle fibers. (d) CT guided biopsy of the lesion.

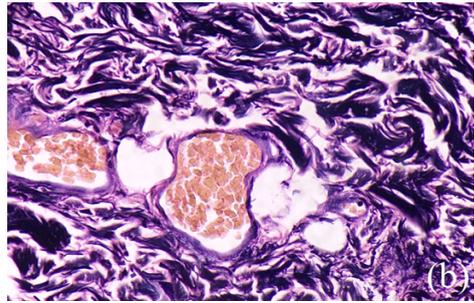
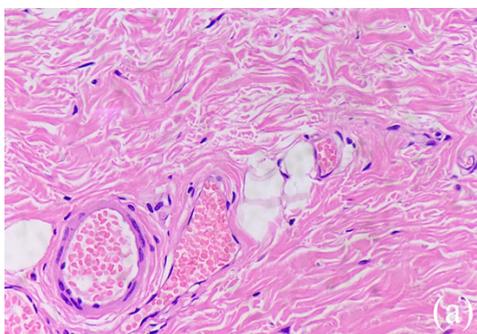


Fig. 3: Elastofibroma Dorsi microphotographs: Hematoxylin and Eosin 400X (a) and Verhoeff-Van Gieson (b) stains show loose collagenous tissue admixed with fragmented elastic fibers and adipose tissue.

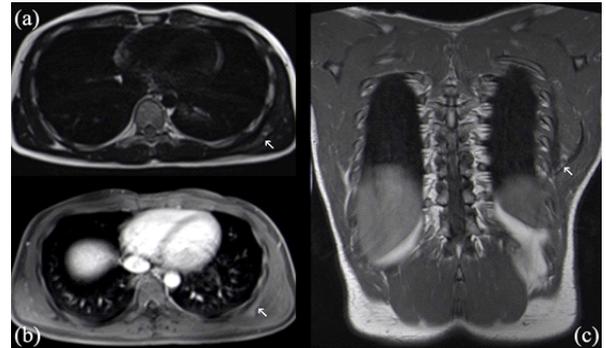


Fig. 4: Elastofibroma Dorsi (arrow) of left infrascapular region on MRI: T1 weighted pre contrast axial (a) & coronal (c) sections and post contrast fat suppressed axial (b) sections show effacement of the fat plane between the serratus anterior and latissimus dorsi muscle with elliptical soft tissue thickening.

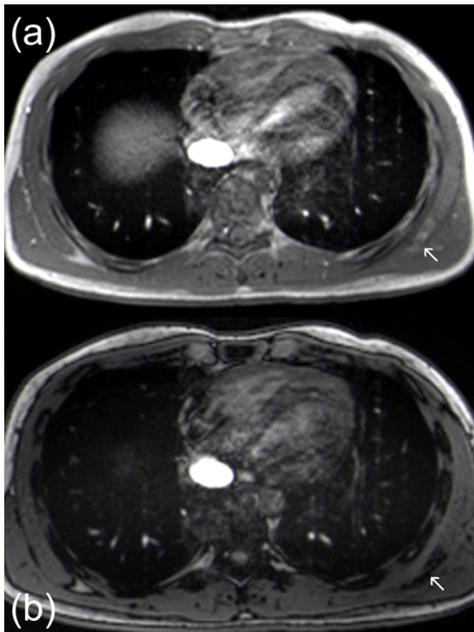


Fig. 5: Elastofibroma Dorsi (arrow) of left infrascapular region on MRI- in and out phase sequence. In (a) and out (b) phase sequence showing India ink artifact on out phase, consistent with the fatty content of the lesion.

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