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SHORT COMMUNICATION

Elastofibroma dorsi

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Summary

We present our experience on 6 patients (4 females, 2 males) with elastofibroma dorsi. The diagnosis was based on imaging studies along with clinical examination. Surgery was decided due to the symptomatic nature of the tumors along with the consent and willingness of the patients. All patients had an uncomplicated course and long term follow up did not show any disease recurrence.

Key words: *benign, elastofibroma dorsi, surgery, symptoms, tumor*

Introduction

Elastofibroma dorsi is a benign tumor of mesenchymal origin localized mainly in the periscapular region. Histologically, an interference of eosinophilic fibers along with elastic fibers is found mixed with gathering of mature adipose cells and fibroblasts. The typical location is the top of the scapula between the thoracic wall and the muscles of the area (serratus anterior, lattisimus dorsi muscle) to which is gripped firmly. It is also found in women above 50 years as a unilateral mass of the back and rarely bilateral which is concrete, hard, movable, painless on palpation creating symptoms of local oedema, stiffness and dysfunction of the involved shoulder joint and rarely pain due to pressure.

Methods

During the last 8 years (2004-2012) 6 patients (4 females – 2 males) with elastofibroma dorsi of the back were admitted to our service. The mean patient age was 55 years (range 49-60). In 4 of them, the mass was unilateral, but in 1 (one female) it was bilateral. Location was right to left 5:2. On admission the mass was visible only on prone position with the upper limbs on extension. On standing position, there was only an asymmetry on the back view of the thorax. The appearance of the mass was suggestive of a chronic condition. None of the patients reported recent or past injury of the involved area neither had intense physical exercise. All patients had a chest X-ray (f/p) and a computed tomography of the thorax. In some patients an MRI was ordered as well. Imaging reports were suggestive of elastofibroma as a well-defined, of uneven density mass of different dimensions located under the subscapular region, between thoracic cage and serratus anterior muscle.

Results

All patiens were surgically managed including complete removal of the mass with its pseudoblaze and the surrounding muscles to normal tissues. The dimensions of the masses are shown in Table 1. A closed loop diversion and pressure dressing was applied in all patients for a median of 10 days (range 5-12).

The postoperative course was uncomplicated and the patients were discharged after a median of 4 days. The operated shoulders were immobilized for a short period (1-2 weeks) followed by rehabilitation and physical therapy. Monitoring included clinical examination every 6 months for the first year and yearly for the next years.

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Table 1. Patient data

Patients	*Age, years*	*Gender*	*Location*	*Mass size (cm)*
1	50	M	R	13x 9 x 2.5
2	59	F	R	6 x 5 x 4
3	60	F	L	10 x 6.5 x 3
4	56	F	R	8 x 7 x 2
5	49	F	R	10 x 6 x 2.5
6	66	F	R-L	8.5 x 5 x 2.5

R: right, L: left, M: male, F: female

Imaging examination was ordered if symptoms were developed associated with the previous lesion. None of the patients relapsed during the study period. All of them recovered to full physical activity within 6 months without reporting limitations or dysfunction of the related upper limb/shoulder. The characteristics of the patients and the tumors are shown in Table 1. Figures 1 and 2 show the lesion on CT and MRI in one of our patients.

Histology revealed hard-fibrous, elastic and myxoid structure with adipose differentiation on places (Figure 3). The presence of degenerated hub-structured with cogged edges elastic fibers was confirmed with the use of Van-Gieson, Weigert and hematoxylin-eosin staining. All surgical margins were free of infiltration.

Discussion

Elastofibroma dorsi was first described by Jarvi and Saxen [1] in a series of 4 patients and is historically considered by Brandser et al.[2] as a rare and benign soft tissue pseudotumor located in the scapular region and found predominantly in the elderly. This rarity was contradicted by the Nagamine et al. [3] with a large series of cases and shortly after again by Jarvi and Lansimies [4] who found an incidence up to 24% in women and 11% in men aged over 55 years . Giebel et al. [5] found an incidence of elastofibroma around 13 % with 81% of the patients having pre-elastofibroma lesions.

Regarding the localization of elastofibroma, the vast majority of them (93%) [6] are located in the lower angle of the scapula between the thoracic wall, serratus anterior, lattisimus dorsi muscles. Other locations are also described, including ischial tuberosities, sciatic, upper limbs, lower limbs, intraspinal spaces, stomach and the peritoneum/omentum [3,7,8]. It is bilateral in 10% of the cases.

World Health Organization (WHO) of soft tissue tumors taxonomy in 2002 characterises

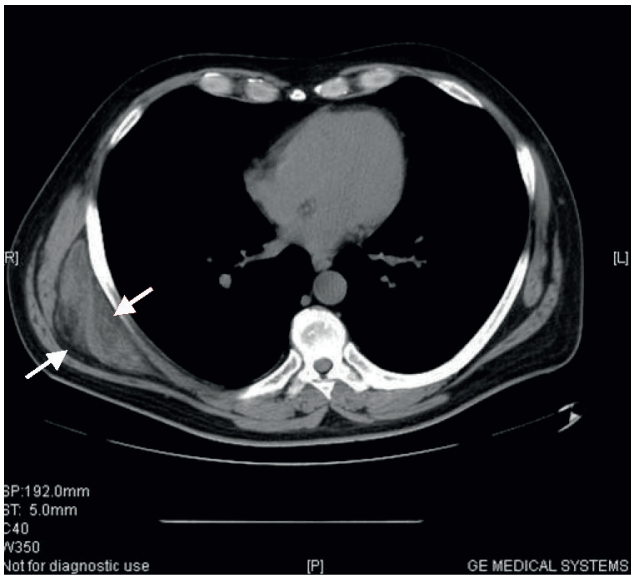

Figure 1. CT image of elastofibroma dorsi (arrows).

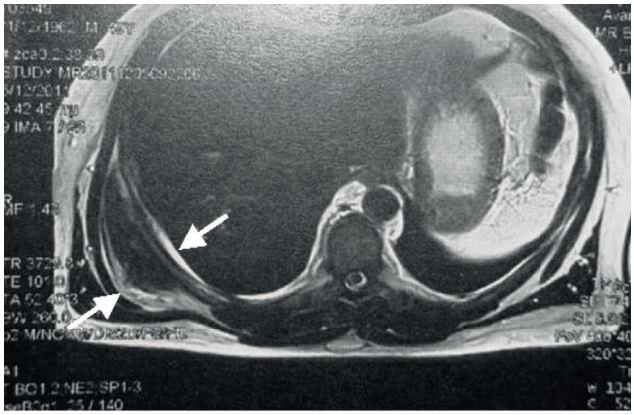

Figure 2. MRI image of elastofibroma dorsi (arrows).

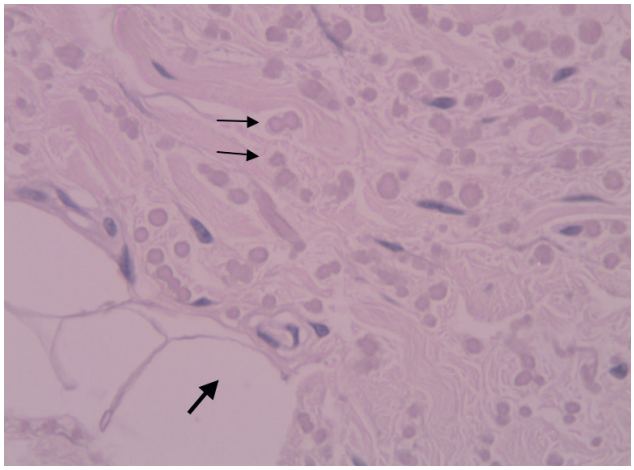

Figure 3. Altered elastic fibers (thin arrows) in a collagenous matrix with a minor mature fat component (thick arrow). Hematoxylin- eosin x400.

elastofibroma as a benign fibroblastic/muscle-fibroblastic tumor. Despite this, elastofibroma is frequently referred to hyperplasia or metaplasia of fibroblasts due to chronic inflammation which is the result of friction or injury of the elastic tissue [8]. The pathogenesis remains unsolved but there are three main dominant theories regarding its etiology. The first suggests that chronic friction due to repeated mechanical stress results to injury that leads to hyperproduction of elastic tissue from the stimulated fibroblasts. The description of hard-working men (repetitive manual labor) was compatible with this theory [1]. Thereafter, however, the emerging diagnosis of the disease in females led to the second theory of reactive fibroblastic hyperplasia and secondary degeneration of elastic fibers due to hypoperfusion [10]. A third theory suggests an underlying enzymatic disorder or deficiency [11] as the cause of development of these tumors, based on observations with familial distribution and also in a patient with double location including the stomach and the scapula [12,13].

Of interest was the case of a male patient, 50-year-old office employee, with mild physical activity and a history of familial multiple symmetrical adipose syndrome. This patient had a known inherited syndrome with multiple subcutaneous lesions in several parts of the body but also had a relatively rapid growing tumor in his back that had different features from the known ones. Histology revealed subcutaneous lesions as being of lipomatous origin and the scapular mass as elastofibroma. However, there are reasonable questions whether both lesions were related and in which way.

The clinical examination of elastofibroma starts with macroscopic characteristics during inspection and palpation and is completed with imaging studies (ultrasound, CT scanning and MRI) of the regions in question. These imaging studies are the gold standard examinations for diagnosis, whereas MRI remains the examination of choice for the thorax because of its specific imaging features allowing the differential diagnosis between sarcoma, liposarcoma, hemangioma, hematomas, lipomas and several other lesions including fluid collections. MRI will reveal a well defined lesion of uneven density without oedema, the muscles' marks and the mass in which longitudinal adipose fascicles are identified.

It was accepted that following the diagnostic and therapeutic algorithm suggested by Muratori [14] , the diagnosis is definite, but the most important point was to exclude the presence of other diseases such as sarcoma or lipomas which require other therapeutic approach and have worse prognosis. Characteristically, an open biopsy is suggested instead of a fine needle aspiration (FNA) because the latter is not diagnostic due to the absence of cells regarding elastofibromas.

The method of choice regarding surgical therapy is resection on normal tissue boundaries -R0 (negative macroscopic and microscopic margins) - including the pseudoblaze without large resection of the surrounding healthy tissues without infiltration [15]. The prognosis is excellent with extremely low relapses [16]. The patients are considered free of disease but a sparsely clinical follow up seems to be reasonable. It must be noted that surgical intervention should be considered as a therapy of choice in patients with symptomatic disease or in patients with diagnostic doubts. In this case an open excisional biopsy is mandatory. In asymptomatic patients a clinical follow up in due course is the option of choice.

Conclusions

Elastofibroma is a rare, benign soft tissue tumor, most commonly found in the subscapular area. Its growth is slow and has specific characteristics on imaging studies. Our experience with 6 cases that were surgically managed is in accordance with the literature which suggests that the therapeutic option of choice in symptomatic patients is surgical - R0 excision - of the tumor. Due to the very good prognosis of this disease, the selection of appropriate therapy (conservative vs surgical) is a matter of discussion between the attending physician and the patient.

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