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Reference:

De Weerdts Glenn, Verhoeven Veronique, Vrints Ina, Thiessen Filip, Tondou Thierry.- Elastofibroma dorsi : a case report of bilateral occurrence and review of literature
Acta chirurgica Belgica - ISSN 0001-5458 - Brussels, Acta medica belgica, 2019, 5 p.
Full text (Publisher's DOI): <https://doi.org/10.1080/00015458.2019.1642595>
To cite this reference: <https://hdl.handle.net/10067/1606980151162165141>

Elastofibroma Dorsi: a Case Report of bilateral Occurrence and Review of Literature

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Elastofibroma Dorsi: a Case Report of bilateral Occurrence and Review of Literature

This report presents the case of a man with bilateral elastofibroma dorsi (ED). He first presented at the age of 49 with a subscapular ED on the right hand side and again at the age of 53 with a subscapular ED on the left hand side. At both times, diagnosis of ED was histopathologically confirmed after surgical resection. And, again at both times, the postoperative course was characterized by development of a seroma. Furthermore this report shows a brief review of literature on ED. It contains a summary of the current data on prevalence, etiology, clinical presentation, diagnosis, histopathological findings, surgical treatment and postoperative management.

Keywords: elastofibroma dorsi, tumor, subscapular, connective tissue, seroma

Background

Elastofibroma dorsi (ED) was first described by Jarvi and Saxen in 1961 [1]. It is a rare, slow growing soft tissue tumor of mesenchymal origin with benign characteristics. ED is most frequently located between the inferior scapular angle and the posterior thoracic wall and has the highest prevalence in middle aged to older women [1-5]. Although clinical reports are scarce, prevalence is estimated to be as high as 2% when looking at retrospective CT studies or even as high as 11-24% in autopsy studies of males and females over 55 years of age [6-8]. In 12-73% of the cases ED presents bilaterally [2,4,5]. The exact aetiology of ED however, is currently unknown. Nevertheless, hypotheses involving friction, trauma, tissue degeneration and genetic predisposition have been reported in current literature [5].

Case presentation

Our patient, a 49 year old man, presented with a prominent subscapular soft tissue tumor on the right hand side which appeared as an ED on magnetic resonance imaging (MRI). Nevertheless, differential diagnosis with other benign tumors (e.g. neurofibroma) and malignant tumors (e.g. fibrous histiocytoma) had to be made.

Therefore, and in order to relieve the patient of his symptoms, excision of the tumor was planned and carried out through a transverse incision over the right latissimus dorsi muscle and by mobilization of the subscapularis muscle and the inferior scapular angle.

The soft tissue tumor, 10x3x9cm in size and 225g in weight, was excised in total macroscopically and was sent for histopathological examination. Microscopic examination showed fibroadipous tissue with large adipocytes and strains of acellular collagen. No atypical cells were found in the specimen, nor were signs of abnormal mitotical activity seen, excluding malignancy. After elastin staining, in order to differentiate with lipoma, the diagnosis of ED was confirmed histopathologically.

Postoperatively the procedure was complicated by development of a seroma in the long term follow up. Drainage via reoperation was mandatory. During this procedure 700cc serosanguinolent fluid was drained and a latissimus dorsi muscle flap, originating from the lateral fibers of the ipsilateral muscle, was created to cover the remaining defect.

Further recovery proceeded without any mentionable complications.

4,5 years later, at the age of 53, the same patient presented again with a similar subscapular tumor on the left hand side. New MRI investigation showed a mass, suggestive for ED, deep to the left anterior serratus muscle. On the right hand side no signs of recurrence were observed on imaging. An excision of the tumor was carried out via a procedure identical to that of the first operation on de right hand side. Since histopathological findings were similar to those of the contralateral mass, the diagnosis of ED, with a weight of 109g and dimensions of 9,5x8x4cm, was confirmed. This time

the surgical procedure was complicated by development of a seroma too. Due to the volume of the fluid accumulation reoperation was necessary for drainage. A similar procedure was carried out. No complications were seen hereafter. The patient recovered well.

Discussion

ED is a benign, connective tissue tumor with a slow growth pattern. In most cases (99%) it is located subscapular, between the inferior scapular angle and the posterior chest wall. More precisely, it's most frequent topology it is near the 6th, 7th and 8th rib deep in the latissimus dorsi, serratus anterior or rhomboids muscles [2,5,8].

Nevertheless, other locations of ED have been described: in the spine (ligamentum flavum, intraspinally), axilla, foot, hand, thigh and buttock (greater trochanter, iliac crest, ischial tuberosity), intra-articular, abdominally (stomach, rectum, omentum), near the olecranon or in the deltoid muscles or even the tricuspid valve and sclera [4,9,10].

ED is said to be a rare entity, but the actual prevalence may be higher than assumed clinically. Brandser et al. [7] estimated that the prevalence of ED, based on CT screenings, is about 2% in a patient population with age older than 60 years. Autopsy studies by Jarvi et al. [6] showed higher prevalences of ED with rates of 11,2% in males and 24,4% in females older than 55 years. Current findings thus result in the general assumption that ED is more likely to occur in middle aged to elderly females over the age of 50 years [1-5]. In 12 to 73% of the cases, subscapular ED presents bilaterally [2,4,5].

Patients with subscapular ED usually present with symptoms of pain and functional limitation of the ipsilateral shoulder. Not infrequently, the condition is associated with snapping of the scapula and impingement-like features. Therefore, adequate history

taking and meticulous physical examination is imperative to prevent misdiagnosing this condition as a tear of the rotator cuff or subacromial bursitis [11-13]. An ED larger than 5cm might be clinically visible as a lump. A deeply located ED however, is not clinically notable unless it reaches a size of 5cm or larger [5].

Apart from clinical investigation, imaging studies play an important role in diagnosing ED. On MRI ED appears as a poorly circumscribed tissue mass which produces a signal similar to that of skeletal muscle and with a rather specific heterogeneous pattern. On T1- and T2-weighted sequences, the fibrous strands in ED produce a low-intensity signal comparable to that of skeletal muscle. The fatty tissue on the other hand, produces a high signal on T1-weighted images and an intermediate signal on T2-weighted sequences. Thus, resulting in a streaky pattern that is rather typical for ED [10]. This gives MRI a positive predictive value of 93,3% and a sensitivity of 100% [14]. Other useful modalities are computed tomography (CT) and ultrasonography (US). CT, like MRI, shows a heterogeneous mass with indistinct margins and an attenuation similar to that of muscle. But CT fails to visualize the typical streaks of fatty tissue. With US alternations of hypo- and hyperechoic zones might be visualized [10]. Nevertheless, US's diagnostic reliability depends on the radiographer's experience and is understood to be variable.

Nowadays it is presumed that imaging studies, in combination with the location of the tumor and its growth pattern, are sufficient for diagnosing ED. This makes invasive diagnostic procedures rather redundant [5,15-17].

At present, the etiology of ED remains unclear. However, it is thought to be caused by repetitive mechanical microtrauma and irritation [4,8,10]. This explain the higher prevalence of ED in manual laborers [2,6,12] and the right sided predominance, as this might be related to the patient's dominant handedness [9]. Also, hypertrophy, secondary

degeneration and elastofibromatous changes were detected in histopathological studies on lesions of human cadavers. These microscopic alterations are all supportive for the hypothesis of mechanical stress as etiological factor [6,18]. Other factors that might contribute to the development of ED are of a genetic kind. Nagamine et al. [2] reported a positive family history for ED in 32% of their investigated cases. However, these findings are not in keeping with later findings of Parratt et al. [12]. Nishio et al. [19] detected increased DNA copy numbers on the chromosomal regions Xq12-q22 and 19 which are potentially associated with development of ED, and DNA-sequences modifications were found by Hernandez et al. [20] at the chromosomal locations 1p, 13q, 19p, and 22q. These findings support the hypothesis of ED being a true neoplasm. On the other hand, their often bilateral occurrence pleads against a malignant origin [9,21]. This is indorsed by the absence of cellular atypia of microscopic examination. However, a true consensus on the etiology of ED has not been reached yet.

When ED is investigated anatomopathologically, a firm ill-defined mass with a gray-white glistening cut surface is perceived macroscopically [9]. On microscopic histopathological investigation hypocellular lesions with spindle cells resembling fibroblasts, and eosinophilic collagen and elastic fibers can be seen. The interspersed mesenchymal adipocytes should not show any signs of cellular atypia. When Elastica-van-Gieson histochemical dye is used, a rich amount of elastine fibers and globules can be visualized [4,5,21,22].

In order to obtain tissue samples for histopathological examination open biopsy or at least core needle biopsy is recommended to obtain sufficient amounts of tissue. Due to the hypocellular characteristics of ED fine needle aspiration is not recommended [21]. Complete surgical excision can be considered when treating subscapular ED as it is effective in eliminating pain symptoms and resetting functionality of the effected

shoulder girdle. Parratt et al. [12] showed that surgical treatment can reduce the visual analogue scale (VAS) for pain from 4,6 preoperatively to 2,4 postoperatively, and that it can enhance the mean forward flexion of the shoulder from 135° to 166° with a mean postoperative Stanmore score of 78,1%.

Surgery, however, is not considered mandatory, especially not in small and asymptomatic tumors (< 5cm) [21,23]. If surgical excision is carried out, the ED is best to be resected completely since incomplete resection may result in recurrence [2,12].

Other complications that might occur after excision of subscapular ED are seroma and hematoma (38,9-87,5% [12,23-25]) and infection [3,12]. The incidence of postoperative seroma and hematoma is associated with the size of the resected ED [23]. This is easily understood when taking under consideration that larger EDs result in larger wound beds and larger dead spaces after they're resected. That is why techniques used by plastic surgeons in latissimus dorsi flap reconstructions, such as quilting sutures and fibrin sealants which reduce the incidence of postoperative seroma at the donor site, might be used in ED surgery as well [23,26,27]. Development of seroma and hematoma after ED resection might also be prevented postoperatively by sufficiently long wound drainage, immobilization of the affected shoulder and by application of a compression bandage [21]. Nevertheless, a protocol or clear consensus about postoperative management hasn't been published in current literature yet [5].

If ED remains unexcised, as mentioned earlier, it most frequently shows a slow growth pattern. Nevertheless, few reports of rapid growing EDs and one report of spontaneous total regression are found in current literature [28]. However, there exist no reports of malignant transformation.

Conclusion

ED is a benign connective tissue tumor that most frequently appears subscapularly as shown in this case. It is not uncommon for it to present bilaterally, though it does not necessarily affect both sides simultaneously. Therefore it is not injudicious to carefully examine the contralateral side. In order to diagnose ED MRI is ought to be sufficient, making tissue biopsy obsolete. Nevertheless, in order to treat this condition and resolve the corresponding symptoms, complete surgical excision should be carried out. If taken under consideration such a surgical procedure, one has to be aware of the high risk of seroma formation, as occurred in this case on both sides.

Disclosure of interest

The authors report no conflicts of interest.

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