Bilateral elastofibroma dorsi: A case report and mini-review of the literature

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Abstract. Elastofibroma dorsi (EFD) is a rare benign lesion characterized by elastic fiber proliferation in a collagenous stroma with adipose tissue. It is typically asymptomatic and found in the infrascapular region. The present study describes the case of a patient with bilateral symptomatic EFD, which caused pain and restricted the daily activity of the patient. The patient was a 51-year-old female who presented with a 4-month history of bilateral masses in the scapular region. She complained that the lesion was limiting her daily activities and was painful. She underwent surgery for the excision of both masses. This type of lesion is usually found incidentally since it is asymptomatic. However, symptomatic cases do exist, with pain being the most common complaint. As the clinical presentation of this type of tumor is not always clear, imaging is required for the diagnosis. The main modality for managing symptomatic EFD is the surgical excision of the mass. On the whole, the present study demonstrates that EFD is a rare benign tumor, that can be symptomatic.

Introduction

Elastofibroma dorsi (EFD) is a rare benign, non-encapsulated lesion characterized by the proliferation of elastic fibers in a collagenous stroma with adipose tissue (1). The lesion mostly occurs in the infrascapular region (2). As a result of the unspecific clinical presentation of the mass, it is difficult to make an accurate diagnosis based on clinical signs and symptoms alone (3). EFD is asymptomatic in the majority of cases and is mostly diagnosed incidentally on cross-sectional imaging studies of other indications (4). Pain appears to be the most frequent and main complaint in symptomatic cases, ranging from mild pain to severe disabling pain. The condition was initially described by Jarvi in 1961 as a rare lesion, and is currently classified as a benign fibroblastic/myofibroblastic tumor according to the 2020 World Health Organization (WHO) classification (5,6). The present study describes the case of a female patient with bilateral symptomatic EFD in the scapular region that affected her daily activity. In addition, a literature review of similar cases is presented. All the reviewed and cited studies have been confirmed to be credible (7).

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Case report

Patient information. The patient was a 51-year-old female who presented to the Smart Health Tower (Sulaimani, Iraq) with a 4-month history of bilateral masses on the scapular region. She was complaining of restricted daily activities and pain due to the lesions.

Clinical findings. Upon a physical examination, two palpable masses were noted in the suprascapular region that appeared to be causing conflicting pain, and limiting daily activities and motion. The patient had a history of hypertension and diabetes.

Diagnostic assessment. The patient underwent a thorough diagnostic evaluation, which revealed intermediate vitamin D3 deficiency (20.28 ng/ml; normal range, 30-50 ng/ml) and a normal complete blood count, apart from an elevated lymphocyte count ($3.8x10^9$; normal range, $1\text{-}3.5x10^9$). Her biochemistry evaluation indicated elevated levels of C-reactive protein (0.58 mg/dl; normal range, <0.5 mg/dl) and serum uric acid (6.2 mg/dl; normal range, 2.4-5.7 mg/dl) with normal bilirubin, aspartate aminotransferase, alkaline phosphatase, serum creatinine, urea, blood urea nitrogen, calcium and lactate dehydrogenase levels. An echocardiography revealed no significant lesion and a good ejection fraction of 68%. An ultrasonography revealed evidence of a solid mass of 10x8x3 cm in size at the posterior-lateral aspect of the lower right chest wall. The lesion appeared to be deeply seated on the muscle. A computed tomography (CT) scan demonstrated bilateral suprascapular masses of soft tissue that were poorly

defined, with fat density interspersed between the inferior angle of the scapula/latissimus dorsi muscle and the chest wall. Upon imaging, the mass on the right side measured 6.5x8x2.8 cm, while the one on the left measured 6x7x2.5 cm. The authors were not able to retrieve the CT scan and ultrasonography images of the patient.

Therapeutic intervention. A pre-operative biopsy was performed using an image-guided core biopsy, which revealed tissue cores composed of spindle cells with entrapped fat, elastic fibers and a rope-like beaded structure in a light pink collagenized background. Under general anesthesia, the surgical excision of the masses was conducted, with a section of each sent for histopathological evaluation. A post-operative histopathological examination was performed. This was performed on 5-μm-thick paraffin-embedded sections. The sections were fixed with 10% neutral-buffered formalin at room temperature for 24 h and then stained with hematoxylin and eosin (Bio Optica Co.) for 1-2 min at room temperature. The sections were observed under a light microscope (Leica Microsystems GmbH). The results of the histopathological examination revealed ill-defined proliferation composed of collagen bundles with large, thick elastic fibers in a loose stroma that were infiltrating into the surrounding fibrofatty tissue, confirming the diagnosis of bilateral EFD (Fig. 1).

Follow-up and outcomes. The post-operative outcome was satisfactory, and the patient is in a good condition.

Discussion

The present study performed a mini-review of the literature in order to identify relevant studies on this type of tumor. The literature review involved a search on Google Scholar, employing key words, including 'Elastofibroma dorsi' and 'bilateral elastofibroma dorsi' to identify relevant studies. The literature review was also assessed based on the CASP checklist. A total of 15 studies on EFD were identified, including a total of 38 cases (Table I). The youngest case documented in the literature was that of a 39-year-old male who was 44 years younger than the oldest documented case, who was an 83-year-old female (4,8). As was expected, a predominance of the female sex was observed for the lesion, with a 2:1 female-to-male ratio. Although the majority of the identified studies stated that most of the cases were asymptomatic, only 4 cases out of the 38 (10.5%) cases exhibited no symptoms (2,9,10). Pain was the chief complaint in symptomatic individuals with other symptoms, such as discomfort and fatigue. Imaging analyses, such as ultrasound, CT scan and magnetic resonance imaging (MRI) were frequently used for diagnosis (11-17). Some studies utilized more than one imaging technique to further analyze the lesion and made a comparison between them (3,9-12,16). As regards the location of the lesion, infrascapular EFD was the most common in the identified literature, as also indicated by many other studies (7,11,12). Other sites included the suprascapular, subscapular and other regions of the back (Table I). As regards the laterality of the lesion, bilateral EFD was the most frequent. Surgery was the management of choice in the majority of the cases (89.5%).

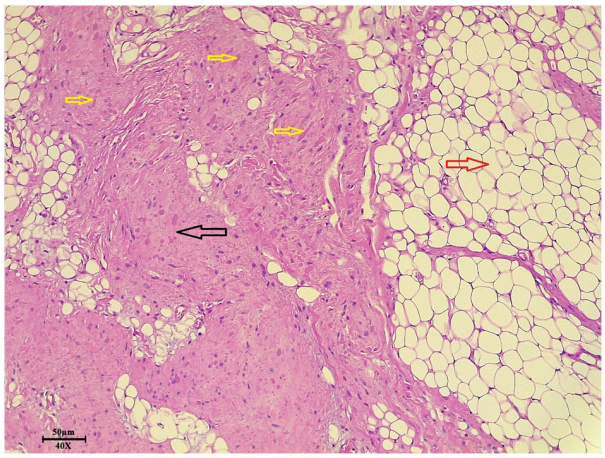

Figure 1. The image of the section illustrates thick, densely eosinophilic elastin bands (yellow arrows) mixed with collagen (black arrow) and adipose tissue in the background (orange arrow) (magnification, x40).

EFD is a rare benign tumor of mesenchymal origin typically occurring in middle-aged women and elderly individuals (3). It is characterized by elastic fiber proliferation inside a stroma of collagenous adipose tissue (1). In a previous retrospective study on 4,435 chest CT scans, EFD was found incidentally in eight of them, indicating a prevalence of 0.8% (18). The lesion exhibited a predominance for the female sex, with a 2:1 female-to-male ratio (according to the authors' review) and a mean age of 76.6 years (18). The specific rationale behind the higher incidence of EFD in females remains unclear. However, Metin *et al* (19) observed that among the 72 females in their case series, the majority (66.6%) were housewives. This observation may indicate a potential association with the substantial time spent by housewives engaging in household activities that require the repetitive use of the upper extremities, such as cleaning and cooking (19). The same study revealed that the lesion was located on the right side in 49 of the total number of patients (n=84; 58.3%), on the left side in 16 (19%) of the patients, and was bilateral in the remaining 19 (22.6%). There appeared to be a statistically significant association between the dominant hand and the mass forming on the same side (19). A theory suggests that chronic and repetitive mechanical stress results in microtrauma, leading to the overproduction of elastic tissue from stimulated fibroblasts. This could explain the predilection for the dominant hand, given its more extensive use in daily activities (16). A previous study demonstrated there was no statistically significant association between either the sex or age of the patient and the size of the mass (18). The pathophysiology of the lesion remains unclear and is a matter of debate (3). In another study on 258 elderly individuals who had a chest CT scan, the prevalence of EFD was found to be 2% (20). On the other hand, another study reported the mass to be the most common, and it was found in 66% of the studied patients (5). As the lesion tends to be generally asymptomatic and has a slow grow rate, it may remain undiagnosed (2). However, the mass can still be identified in patients who experience symptoms, with pain being the most common complaint (3). In the case described herein, apart from pain, the mass also affected the daily activities of the patient, and she was not able to function normally. A number

Table I. Summary of the data from the studies identified in the literature search.

Author, year of publication	Age/ mean age, years	Sex of included patient(s)	No. of cases	Presenting complaint			Imaging used for diagnosis			Location of lesion				Side of lesion(s)		Management	(Refs.)
				Pain	Asymptomatic	Other symptoms	CT	MRI	US	Infrascapular	Suprascapular	Subscapular	Others	Left	Right		
Kourda *et al*, 2009	66	Female	1	Yes	No	No	No	Yes	No	No	No	Yes	No	Yes	Yes	Surgery	(1)
Karti *et al*, 2022	55	2 Females 1 Male	3	1	1	1	Yes	No	No	No	1	1	1	Yes	Yes	Surgery	(2)
Ngoy *et al*, 2023	50	Female	1	Yes	No	Yes	Yes	Yes	No	No	Yes	No	No	Yes	Yes	Surgery	(3)
Almutlaq *et al*, 2022	39	Male	1	No	No	Yes	Yes	No	No	No	Yes	No	No	Yes	Yes	Surgery	(4)
Fabien *et al*, 2022	56	Male	1	Yes	No	No	Yes	No	No	No	No	No	Yes	Yes	Yes	Surgery	(5)
Abdullah *et al*, 2022	83	Female	1	Yes	No	Yes	Yes	No	No	Yes	No	No	No	Yes	Yes	Surgery	(8)
Yoshida *et al*, 2022	74	Male	1	No	Yes	No	Yes	Yes	No	No	No	No	Yes	Yes	No	Unknown	(9)
Muratori *et al*, 2008	61	7 Females 1 Male	8	No	2	6	Yes	Yes	Yes	No	No	No	Yes	Yes	Yes	Six cases: Surgery Two cases: Conservative	(10)
Sarici *et al*, 2014	62	Female	1	Yes	No	Yes	Yes	Yes	No	No	Yes	No	No	Yes	Yes	Surgery	(11)
Parratt *et al*, 2010	60.9	8 Females 7 Males	15	Yes	No	Yes	Yes	Yes	No	Yes	No	No	No	Yes	Yes	Surgery	(12)
Nadeem *et al*, 2021	54	Female	1	Yes	No	Yes	No	Yes	No	Yes	No	No	No	Yes	No	Conservative	(13)
Limaeim *et al*, 2022	63	Female	1	Yes	No	Yes	No	Yes	No	No	No	Yes	No	No	Yes	Surgery	(14)
Falidas *et al*, 2013	65	Female	1	Yes	No	No	No	Yes	No	Yes	No	No	No	No	Yes	Surgery	(15)
Karrakchou *et al*, 2017	53	Female	1	Yes	No	Yes	No	Yes	Yes	No	No	No	Yes	Yes	Yes	Surgery	(16)
Neagoe *et al*, 2021	65	Female	1	Yes	No	No	Yes	No	Yes	No	No	Yes	No	No	Yes	Surgery	(17)

CT, computed tomography; MRI, magnetic resonance imaging; US, ulstrasound.

of risk factors have been shown to be associated with EFD; however, as aforementioned, the exact pathogenesis is not yet completely understood (2,3,16). The risk factors associated with the lesion can be both environmental and genetic (4). The friction of the lower scapula against the thoracic wall as a result of either manual labor or repetitive minor trauma is considered to promote the development of the lesion (3). This can be supported by the higher prevalence of the disease among manual workers and elderly patients (4).

Both clinical evaluation and imaging can aid in the diagnosis (2). Given that the majority of individuals do not exhibit symptoms, this lesion is typically discovered serendipitously during MRI and CT scans or incidentally during surgical procedures performed for other purposes (4). However, this may not involve bilateral EFD, as the authors' review of the literature revealed that most of the bilateral EFD cases had symptoms. An ultrasound is generally the first line of examination, as it poses no radiation damage to the patient, followed by CT and MRI scans. MRI, although not routinely required, can elevate diagnostic confidence as a result of its good soft tissue contrast (1,13). As per the standard diagnostic approach, an ultrasound was initially performed in the patient described in the present study, which revealed a solid mass with a size of 10x8x3 cm at the posterior-lateral aspect of the lower right chest wall. The mass was heterogeneously echogenic with no features suggesting any underlying rib destruction. The diagnosis was further aided by a CT scan, which revealed not one, but two bilateral suprascapular soft tissue masses that were both poorly defined and exhibited no infiltration of the adjacent structures nor any destruction of adjacent ribs. The definitive method for confirming the diagnosis is a histopathological examination. This examination typically reveals a mesenchymal tumor characterized by dense collagen bands interspersed with numerous irregularly arranged elastic fibers, separated by mature adipose islets (1). The results of the histopathological analysis of both masses in the patient described herein revealed proliferations composed of collagen fibers and large thick elastic fibers within a loose stroma infiltrating into the surrounding fibrofatty tissue. No cellular atypia or mitotic activities were found in the specimen.

As regards treatment options, these vary based on the presence or absence of symptoms, since simple observation is sufficient in asymptomatic cases (1). Surgical resection is reserved for cases where the tumor is sufficiently large in size and symptomatic (4). The case in the present study was managed surgically by removing the masses, as they affected the quality of life of the patient. The recurrence rate following surgical resection is between 0.06 and 4.5%, and hematoma and seroma formation are among the most common complications (19). While wide or radical excision is not deemed necessary, curative marginal resection is recommended, as incomplete excision has been reported to be associated with local tumor recurrence (1). Nevertheless, to the best of our knowledge, no instances of malignant transformation were identified in the existing literature (1,15). In the present study, the surgical outcome of the patient was good, with no recurrence and complications. The present study however, was limited by the absence of an MRI and immunohistochemical examinations, as well as the non-retrieval of CT scan images.

In conclusion, EFD is a rare benign tumor that can be symptomatic. Imaging is required for its diagnosis, and it typically forms within the upper back region. While the occurrence of this mass is infrequent, the prognosis is generally favorable. Post-operative recovery is typically uncomplicated, and the recurrence rates are low.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' contributions

SKA and MS were major contributors to the conception of the study. FHK designed the study. ASA and HOA participated in preparing and drafting the manuscript and were involved in the conception of the study. FHF, JIH were involved in the study design, literature review and processing of the figures. AMA, BAA, and HMH critically revised the manuscript and were involved in the study design. All authors contributed equally to the manuscript, and have read and approved the final version of the manuscript. FHK and SKA confirm the authenticity of all the raw data.

Ethics approval and consent to participate

Written informed consent was obtained from the patient for participation in the present study.

Patient consent for publication

Written informed consent was obtained from the patient for the publication of the present case report and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

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