Medical Research Archives

European Society of Medicine

OPEN ACCESS

Published: March 31, 2024

Citation: Vomer RP, York ER, et al., 2024. The Rubber Band Man: An Unexpected Reason for Breathing Difficulty, Medical Research Archives, [online] 12(3). https://doi.org/10.18103/mra.v12i3.5082

DOI
https://doi.org/10.18103/mra.v12i3.5082

ISSN: 2375-1924

RESEARCH ARTICLE

The Rubber Band Man: An Unexpected Reason for Breathing Difficulty

1. **Rock Paul Vomer, DO, DPT:** Department of Family Medicine, Avance Care, Wilmington North Carolina, 28411 & Department of Family Medicine, Mayo Clinic, Jacksonville, FL, 32224. ***Corresponding author:** rockduq3409@gmail.com
2. **Emma Richards York, DO:** Department of Family Medicine, Prisma Health University of South Carolina, Columbia SC.
3. **Sara Memon:** Department of Family Medicine, Mayo Clinic, Jacksonville, FL, 32224.
4. **George GA Pujalte, MD, FACSM, FAMSSM:** Department of Family Medicine & Department of Orthopedic Surgery, Mayo Clinic, Jacksonville, FL, 32224.

ABSTRACT

A 73-year-old male presented to his physician with difficulty breathing. The patient described a nagging inability to take a deep breath, and his wife noted what appeared to be a mass on his back, that she believed had gotten bigger. He had a notable family history of soft tissues masses in numerous family members. Physical examination revealed non-painful, soft, and movable masses along the inferior edges of both scapulae, consistent with elastofibroma. Pulmonary function testing and cardiac evaluation revealed normal results. The masses remained stable in size across repeat Magnetic Resonance Imaging and were managed conservatively due to their non-impeding nature and lack of cosmetic concerns. This case highlights the importance of considering elastofibroma in patients with soft tissue masses, the significance of their physical symptomatology and the decision-making process involved in selecting appropriate management strategies based on individual patient characteristics and clinical presentations.

Introduction

Elastofibroma, also known as elastofibroma dorsi (ED), is a slow-growing soft tissue tumor.[1] It typically presents as a soft, poorly defined mass that arises on the thoracic wall and frequently occurs bilaterally.[2] They are most commonly located at the inferior pole of the scapula, beneath the rhomboid major and lattisimus dorsi, with some considering this location to be pathognomonic.[3] Most typically the tumors are bilateral, but when presenting unilaterally, more commonly occurring on the right side of the body.[3] Individuals are typically above the age of fifty-five at the time of diagnosis.[4] Generally, elastofibromas are painless and asymptomatic, and due to such, they are frequently discovered incidentally on computed tomography (CT) or Magnetic Resonance Imaging (MRI). However, when symptoms are present, they can include pain, swelling, limitations in chest wall motion, and scapular snapping.[5] If a patient presents with history and physical exam concerning for elastofibroma, ultrasound evaluation is the first-line recommended imaging of choice. Ultrasound evaluation, which is limited by body habitus, typically reveals a mass without defined borders, a heterogenous fasiculated pattern, with a mix of hypo and hyperechogeneous striae. Doppler will show similar vascularization to that of the surrounding musculature. Xray will demonstrate discrete findings of elastofibroma including a widened scapula thoracic space and possibly elevated appearance of the scapula under which elastofibroma is located. If CT is obtained, ED will appear as a poorly defined soft tissue mass without surrounding bone abnormalities. Biopsy is typically unnecessary for diagnosis if the typical sub-scapula location is present along features are demonstrated on imaging.[1]

ED is classified as fibroblastic/myofibroblastic tumors by the WHO Classification of Tumors of Soft Tissue and Bone.[6] They are benign lesions, with no potential for transformation into malignancy.[1] Histologically the mass is made up of collagenous tissue, fat and abnormal elastic fibers.[7] The pathogenesis of elastofibroma is still unclear, with some evidence suggesting they form as a result of repetitive microtrauma between the thoracic and scapular walls.[8] Other potential etiologies include alterations in fibrillogenesis due to chronic irritation or trauma.[6] Management is either surgical excision or conservative monitoring.

While originally thought to be rare, estimations of prevalence vary tremendously. In one study examining a series of 265 autopsies of patients over fifty-five, prevalence of tissue changes similar to elastofibroma were found in 24.4% of females and 11.2% in males.[9] Another study which sought to examine prevalence via CT scan found prevalence to be around 2%, which, while much lower than that found by autopsy is still higher than expected given the common belief that elastofibromas represent "rare" tumors.[1]

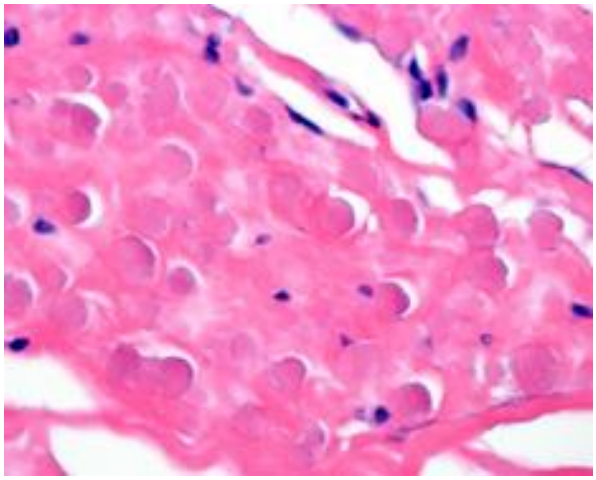

Figure 1. Micrographic image of an Elastofibroma

Case presentation

HISTORY

A 73-year-old male presented for mild difficulty breathing, and presence of an enlarging mass on his back. He described a sensation of being unable to inhale deeply. The patient's wife also reported that she subjectively felt that the masses had been enlarging. The patient was able to continue his current activities of daily living without limitations and exercise daily with 40-minute walks without exacerbating his breathing difficulties. The patient also noted a family history of soft tissue masses in his mother, sister and daughter, some of which required surgical removal. Prior Magnetic resonance imaging (MRI) demonstrated anterior subscapular masses approximately 7.5 x 3.5 x 12.0 cm on the right and 7.0 x 3.0 x10.5 cm on the left.

PHYSICAL EXAM

Cervical active/passive range of motion did not elicit any pain. The shoulder demonstrated full active and passive range motion for flexion, abduction, internal and external rotation. The patient demonstrates 4/5 gross strength for the shoulder. Prominent soft masses, approximately 7-8cm in length, were observed along the lower borders of both scapulae. These masses were soft, not painful, mobile, and did not show any changes in the skin. Examination of the lungs revealed clear lung fields, normal breathing, and no tenderness in the chest wall. The pulmonary examination indicated a steady heart rate and rhythm, with no presence of murmurs, rubs, or gallops.

DIFFERENTIAL DIAGNOSIS

During initial patient encounter, a range of potential differential diagnoses were considered for the observed physical exam findings. These included, but were not limited to, lipoma, fibroma, sarcoma of unspecified type and elastofibroma dorsi.

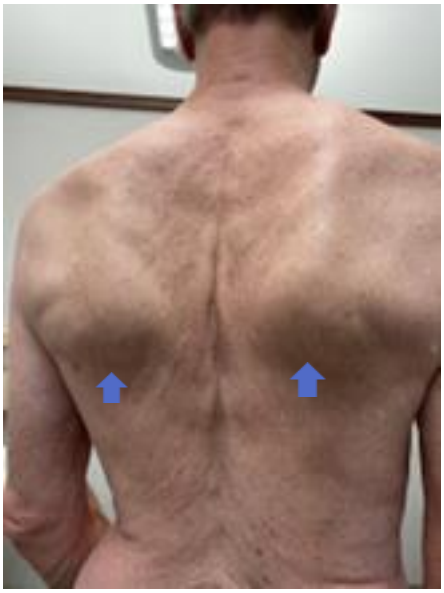

Figure 2. Photograph from initial physical exam. Arrows demarcate 7-8cm soft tissue masses appreciated on palpation.

TESTING/ RESULTS

MRI of the chest, both with and without contrast, revealed presence of bilateral vague fatty and fibrous masses anterior to the scapula, consistent with elastofibroma (Figure 3). There was no significant change in size compared to the previous MRI examination. Pulmonary function tests revealed normal results.

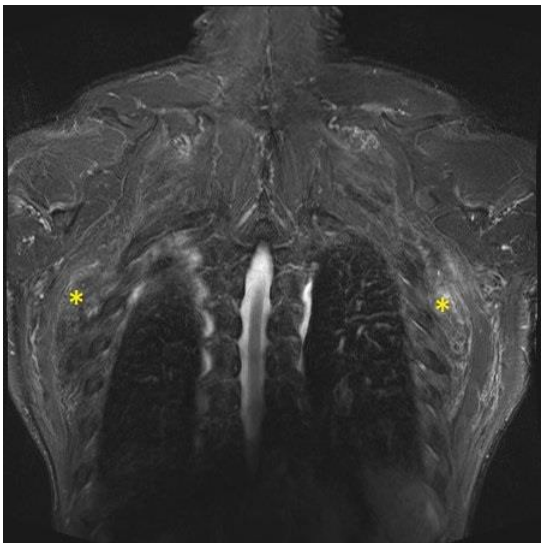

Figure 3. Frontal and Coronal T1 weighted MRI. Star demarks the elastofibroma which is located between the posterior rib cage serratus anterior.

FINAL DIAGNOSIS, TREATMENT AND OUTCOMES

Final diagnosis was stable, bilateral elastofibroma. The patient was referred to cardiology for a cardiac evaluation which demonstrated no evidence of arrhythmia. A surgical consultation was also arranged to assess the potential necessity of a surgical procedure. Given the stable size of his elastofibromas demonstrated on repeat MRI, lack of interference with rib movement, and its minimal cosmetic impact on the patient, the patient opted for a conservative, non-operative approach.

Discussion

Elastofibroma dorsi (ED) is a soft tissue mass characterized by slow growth, composed mainly of collagenous tissue, fat, and abnormal elastic fibers.[6] In this case, the patient presented with mild difficulty breathing and subjectively enlarging bilateral masses on his back, which prompted a thorough evaluation of his symptoms, medical history and physical exam. The clinical presentation of the patient, along with the palpation findings, raised suspicions of an underlying soft tissue mass. The masses were soft, non-painful, and poorly defined, consistent with the typical characteristics of elastofibroma.[1] In addition, the location of the tumors at the base of the scapula also raised suspicion for elastofibroma, as this represents their most common location. The family history of soft tissue masses in the patient's relatives, some of which required surgical removal, could indicate a potential genetic predisposition for this patient. ED is most commonly diagnosed with the help of imaging, such as in this patient. The MRI obtained in this case revealed bilateral subscapular fibrous and fatty masses, consistent with ED.[3] These findings were in line with the previously reported dimensions of the masses, indicating stability over time.

The patient's symptoms of mild difficulty breathing, particularly the sensation of being unable to inhale deeply, could be attributed to the proximity of the masses to the ribcage. However, it's noteworthy that the patient was able to engage in daily exercises involving 40-minute walks without exacerbating his breathing difficulties. This suggests that the masses were not significantly impairing respiratory function. Pulmonary function testing, which yielded normal results, further supported the absence of substantial respiratory compromise due to the masses. Due to the stable nature of the elastofibromas, the absence of impingement on rib motion, and the lack of significant cosmetic concerns, the patient opted for conservative management without surgical resection. Regular follow-up visits, coupled with periodic imaging studies will be performed if the patient's symptoms change to track

any changes in the size or characteristics of the masses.

Frequently, ED is found incidentally when chest imaging is obtained and is thought to be present in up to 2% of asymptomatic adults who are over the age of 60.[9] Most commonly they are a slow growing mass that can be seen unilaterally or bilaterally, but more frequently are bilaterally.[9] Typically, patients will complain of a mass that has appeared on their back, or for those patients who do report symptoms, mid back pain or a clunking as the arm is abducted or adducted is the most common.[10]

In patients presenting with findings concerning for ED, a careful family history should be obtained as prior studies have noted up to one third of patients had a family member with a similar soft-tissue mass.[8] In cases of familial inheritance, it is thought to be due to instability of the 1st chromosome.[11] While genetic inheritance has not been proven, other causes for ED are still unclear as well. Some thought the tumor may be due to repetitive microtrauma, however, this not been found to be consistent in literature.[12,13]

After a thorough history and physical is obtained, Imaging is typically the next step to further clarify the diagnosis, if it has not already been obtained. While xray is a reasonable first step, due to the soft tissue make up of these masses, they are not well-seen on xray other than indirect signs such as an enlarged scapula thoracic space or a slightly raised scapula on the side where the mass is located.[10] Therefore, advanced imaging with MRI or CT is the recommended next step. While MRI is the preferred imaging type, due to having a higher diagnostic confidence, ED can also be found incidentally on CT chest as well.[10] Lesions that are found in a similar location include hemangioma, malignant histiocytoma and lipoma, thus MRI with contrast is considered the preferred method of imaging to further differentiate between benign and malignant lesions. As our patient had previously obtained an MRI Magnetic Resonance Imaging (MRI), another MRI was obtained to be able to best compare the lesions on the two images. Advanced imaging of ED is able to characterize the size, location and the density of the tumor. As the subscapular location at the inferior pole of the scapula is thought to be pathognomonic and our patient had similar appearing masses bilaterally of similar appearing size to the prior imaging obtained, the diagnosis of ED was made without obtaining a biopsy. Frequently, a diagnosis of ED is able to be made by history, exam and imaging alone. If there is concern about malignant etiology, a PET-CT can be obtained or proceed to biopsy of the lesion which can confirm both the diagnosis as well as the benign nature of the lesion.

Biopsy can be performed under local anesthesia with image guidance with ultrasound. Histological evaluation will reveal a mix fibroblastic stromal cells and collagenous stroma as well as a mix of blood vessels, fat cells and amorphous extracellular matrix.[10,14] If a lesion is removed, macroscopically, it will appear as a white colored mass with yellow foci.

Depending on patient symptoms and preference, a conservative or an operative approach can be undertaken. A conservative approach is appropriate for those who are asymptomatic or minimally symptomatic or those who don't want to undergo surgical removal, such as our patient. If surgical removal is pursued, common complications include seroma formation, which occurs in 36-88% of patients due to the size of the mass that is removed and creation of dead space upon its removal.[9] Inadequate margins during surgical excision, can lead to recurrence of ED.[15]

While ED is often considered to be a rare tumor, with increased understanding of their presentation, appearance and imaging findings, we can decrease the occurrence of unnecessary workup including biopsy and surgical removal of a benign growth in largely asymptomatic patients. It is important to include ED in the differential in patients presenting with soft tissue masses, particularly in the subscapular region, as they are considered benign tumors that can be diagnosed clinically with the aid of imaging such as MRI.

Conclusion

This case highlights the clinical presentation, diagnostic evaluation, and management considerations for elastofibroma. While surgical excision remains a viable option for symptomatic cases, a conservative approach can be appropriate for stable elastofibromas that do not significantly impact the patient's daily life. Ongoing monitoring is essential to ensure that any potential changes are promptly addressed.

References

1. Nishio J, Nakayama S, Nabeshima T, et al. Current update on the diagnosis, management and pathogenesis of Elastofibroma Dorsi. *Anticancer Research*. 2021;41(5):2211-2215. doi:10.21873/anticanres.14997
2. Jena A, Patnayak R, Settipalli S, et al. Elastofibroma: An uncommon tumor revisited. *Journal of Cutaneous and Aesthetic Surgery*. 2016;9(1):34. doi:10.4103/0974-2077.178543
3. Hayes AJ, Alexander N, Clark MA, et al. Elastofibroma: A rare soft tissue tumor with a pathognomonic anatomical location and clinical symptom. *European Journal of Surgical Oncology (EJSO)*. 2004;30(4):450-453. doi:10.1016/j.ejso.2004.01.006
4. Brandser EA, Goree JC, El-Khoury GY. Elastofibroma Dorsi: Prevalence in an elderly patient population as revealed by CT.. *American Journal of Roentgenology*. 1998;171(4):977-980. doi:10.2214/ajr.171.4.9762978
5. Maj J, Gracia I, Doncel A, et al. Elastofibroma Dorsi as a cause of shoulder pain or snapping scapula. *Clinical Orthopaedics and Related Research*. 2001;388:200-204. doi:10.1097/00003086-200107000-00027
6. Sbaraglia M, Bellan E, Dei Tos AP. The 2020 who classification of soft tissue tumors: News and perspectives. *Pathologica*. 2020;113(2):70-84. doi:10.32074/1591-951x-213
7. Kakudo N, Morimoto N, Ogawa T, et al. Elastofibroma Dorsi: A case report with an immunohistochemical and ultrastructural studies. *Medical Molecular Morphology*. 2015;49(1):42-47. doi:10.1007/s00795-015-0112-5.
8. Findikcioglu A, Kilic D, Karadayi Ş, et al. A thoracic surgeon's perspective on the Elastofibroma Dorsi: A benign tumor of the deep infrascapular region. *Thoracic Cancer*. 2013;4(1):35-40. doi:10.1111/j.1759-7714.2012.00139.x
9. Neagoe O, Faur Cl, Ionică M, et al. Elastofibroma Dorsi, a rare condition, with challenging diagnosis. Case report and literature review. *Medicina*. 2021;57(4):370. doi:10.3390/medicina57040370.
10. Oliva MS, Smimmo A, Vitiello R, et al. Elastofibroma Dorsi: What's new? *Orthopedic Reviews*. Published online July 1, 2020. doi:10.4081/or.2020.8708.
11. Järvi OH, Länsimies PH. Subclinical elastofibromas in the scapular region in an autopsy series. *Acta Pathologica Microbiologica Scandinavica Section A Pathology*. 1975;83A(1):87-108. doi:10.1111/j.1699-0463.1975.tb01361.x
12. Hoffman JK, Klein NA, Mclnerney VK. Bilateral elastofibroma. *Clinical Orthopaedics and Related Research*. 1996;325:245-250. doi:10.1097/00003086-199604000-00030
13. Daigeler A, Vogt PM, Busch K, et al. Elastofibroma Dorsi – differential diagnosis in chest wall tumors. *World Journal of Surgical Oncology*. 2007;5(1). doi:10.1186/1477-7819-5-15
14. Yamazaki K. An ultrastructural and immunohistochemical study of elastofibroma: CD 34, MEF-2, prominin 2 (CD133), and factor xiiia-positive proliferating fibroblastic stromal cells connected by CX43-type gap junctions. *Ultrastructural Pathology*. 2007;31(3):209-219. doi:10.1080/01913120701350365
15. Briccoli A, Casadei R, Di Renzo M, et al. Elastofibroma Dorsi. *Surgery Today*. 2000;30(2):147-152. doi:10.1007/pl0001006