A Rapidly Enlarging Soft Tissue Lesion of the Back

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Received: 14 July 2021
Accepted: 12 October 2021

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DOI 10.5001/omj.2023.12

Introduction

Seventeen-year-old boy, with no comorbidities, presented with a large mass in the back extending to the chest. At the age of seven he had a small mass in the left lateral chest wall at the mid-axillary line which was excised in one of the periphery hospitals (no official medical report). Six years after the surgery, another mass appeared just above the scar of the previously excised mass, it increased in size dramatically over a period of four years and therefore he came to our center. On examination, The swelling was soft, lobulated, cystic, not attached to the overlying skin, extending from left lateral chest to the back and measuring 13x7 cm anteriorly, 7x4 cm on the lateral chest and 28x23 cm posteriorly, figure 1 (A, B). MRI revealed a large heterogenous mass measuring 17X6X20 cm, extending from the 4th to the 10th rib involving the left paravertebral muscles, figure 2.

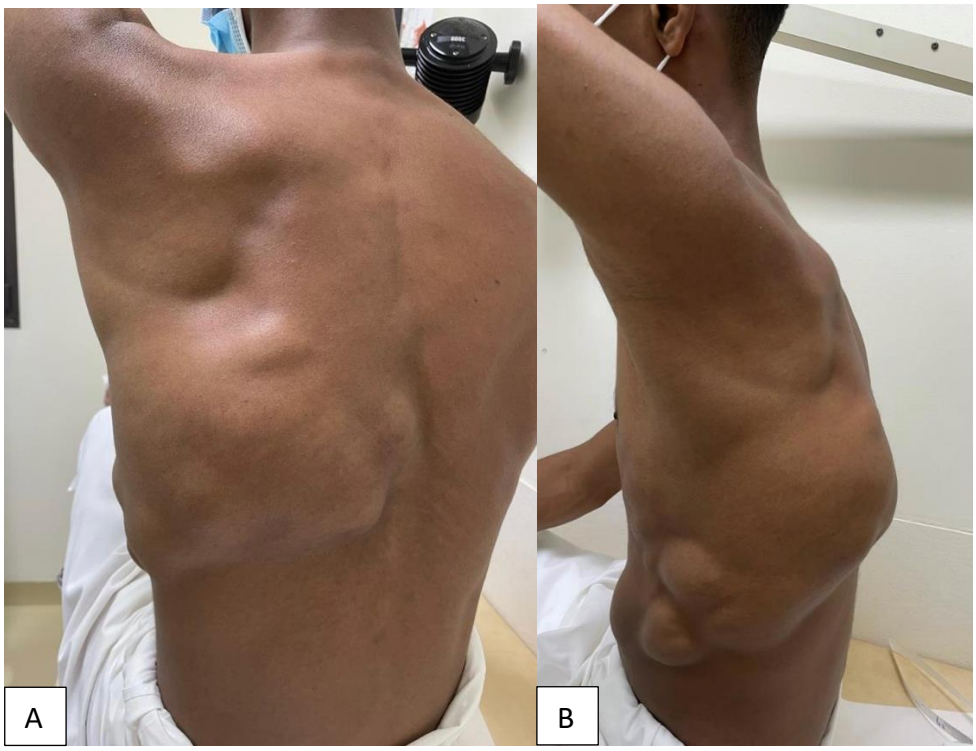

Figure 1

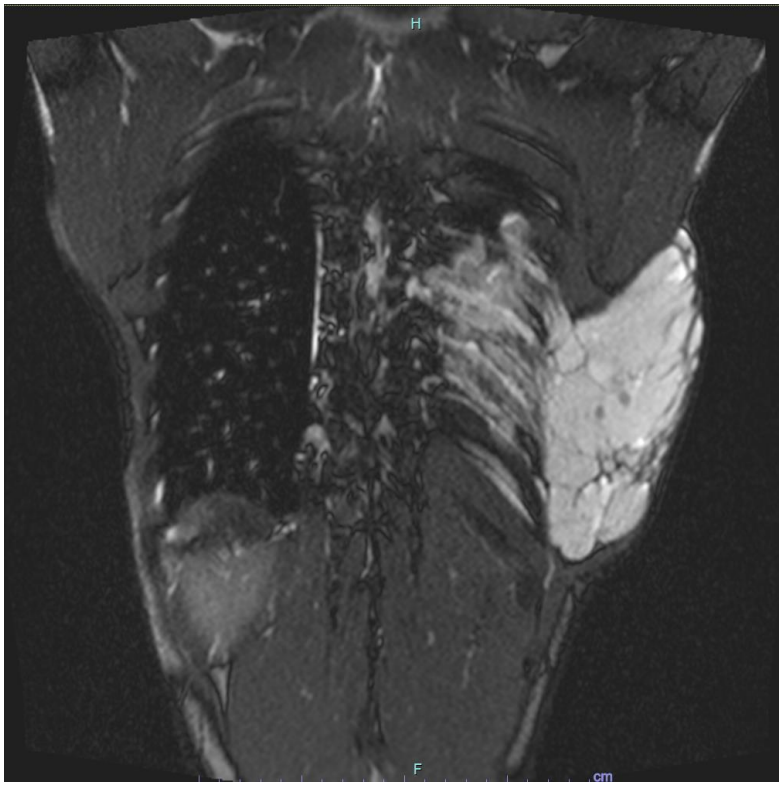

Figure 2

Questions

1- What is the diagnosis?

2- What are the MRI findings of this lesion?

3- How will you treat this patient?

Answers

1- Lymphovascular malformation

2- FAT SAT T2W coronal MRI of the chest wall shows a large, relatively well-defined solid soft tissue lesion in the left posterolateral chest wall- extending to several intercostal and extrapleural spaces. The lesion shows marked T2 hyperintensity with few thin hypointense septa. Few rounded T2 hypointense foci are seen, which corresponds to calcification. No cystic spaces or fluid level. No fat within the lesion. Post-contrast dynamic images reveal delayed heterogeneous enhancement of the lesion. No arterial enhancement. No enlarged arterial feeders or early draining veins. Presence of multiple calcific foci with central lucency within the mass suggestive of phleboliths. Overall MRI features are of slow-flow vascular malformation with venous and lymphatic components.

3-Sclerotherapy with bleomycin injections.

Discussion

Vascular anomalies affect up to 10% of newborns; they were classified in 1982 into congenital vascular malformations (CVM) and vascular tumors .This classification has been revised by the International Society for the Study of Vascular Anomalies (ISSVA) (figure 3) (1,2) .

Whereas vascular tumors may regress with age; CVM never regress on their own. They are subdivided into venous, lymphatic, arterio-venous and combined malformations (3). Lympho-vascular malformations (LVM), also known as haemangio-lymphangiomas, are low flow vascular malformations that consist of malformed lymphatic channels and they account for about 12% of vascular anomalies (4). They can be classified into macrocystic (>1cm), microcystic (<1 cm) or mixed cystic according to their cystic appearance. Moreover, they can be generalized or localized at the cutaneous, subcutaneous, fatty or intramuscular level (3). Our patient had a macrocystic type of LVM . Diagnostic modalities for LVM include ultrasonography, MRI and angiography. MRI is considered the best modality because of its soft tissue resolution. LVM are usually well defined, lobulated and hyperintense on T2 weighted MRI, and hypointense to isointense on T1 weighted images (4,5). Macrocystic lesions are effectively treated by sclerotherapy as in our case, commonly used sclerosing agents are sodium tetradecyl sulfate (STS), bleomycin (>50% reduction in lesion size) and doxycycline, there is no consensus regarding the best sclerosing agent in treating macrocystic LVM. Our patient received three sessions of sclerotherapy; there was a gap of one month between one session and another. The size of the lesion has decreased significantly after the 3d session ; 8x8 cm anteriorly, 7x4 cm on the lateral chest and 22x22 cm posteriorly (figure 4) . Surgical intervention is reserved for patients who fail to respond to sclerotherapy. By contrast, microcystic LVM are difficult to treat and are typically managed conservatively. However, symptomatic lesions are treated surgically. Surgery is considered the first line of treatment as sclerotherapy is ineffective in treating microcystic lesions; although recurrence happens in 40% of patients with incomplete excision and 17% of patients with complete resection (5,6). Despite the advancements in the management of vascular anomalies, CVMs remain a diagnostic and therapeutic challenge to many physicians therefore requiring a multidisciplinary approach.

Figure 3: ISSVA Classification for Vascular Anomalies Approved at the 20th ISSVA Workshop, Melbourne, April 2014, last revision May 2018.

Vascular Anomalies				
Vascular Tumours	**Vascular Malformations**			
	Simple	**Combined°**	**Of Major Named Vessels**	**Associated with Other Anomalies**
Benign	Capillary malformations	CVM, CLM	Affecting arteries, veins, and lymphatics of large diameter.	Klippel-Trenaunay Syndrome Parkes Weber syndrome Servelle-Martorell syndrome Sturge-Weber syndrome Limb CM + congenital non-progressive limb overgrowth Maffucci syndrome Macrocephaly – CM Microcephaly – CM CLOVES syndrome Proteus syndrome Bannayan-Riley-Ruvalcaba sd CLAPO syndrome
Locally aggressive or border line	Lymphatic malformations	LVM, CLVM		
Malignant	Venous malformations	CAVM*		
	Arteriovenous malformations	CLAVM*		
	Arteriovenous fistula	Others		Others

° Defined as ≥ 2 vascular malformations found in one lesion

***** High flow lesions

Abbreviations: Capillay-Venous Malformation (CVM); Capillary-Lymphatic Malformation (CLM); Lymphatic-Venous Malformation (LVM); Capillary-Lymphatic-Venous Malformation (CLVM); Capillary-Arteriovenous Malformation (CAVM); Capillary-Lymphatic-Arteriovenous Malformation (CLAVM).

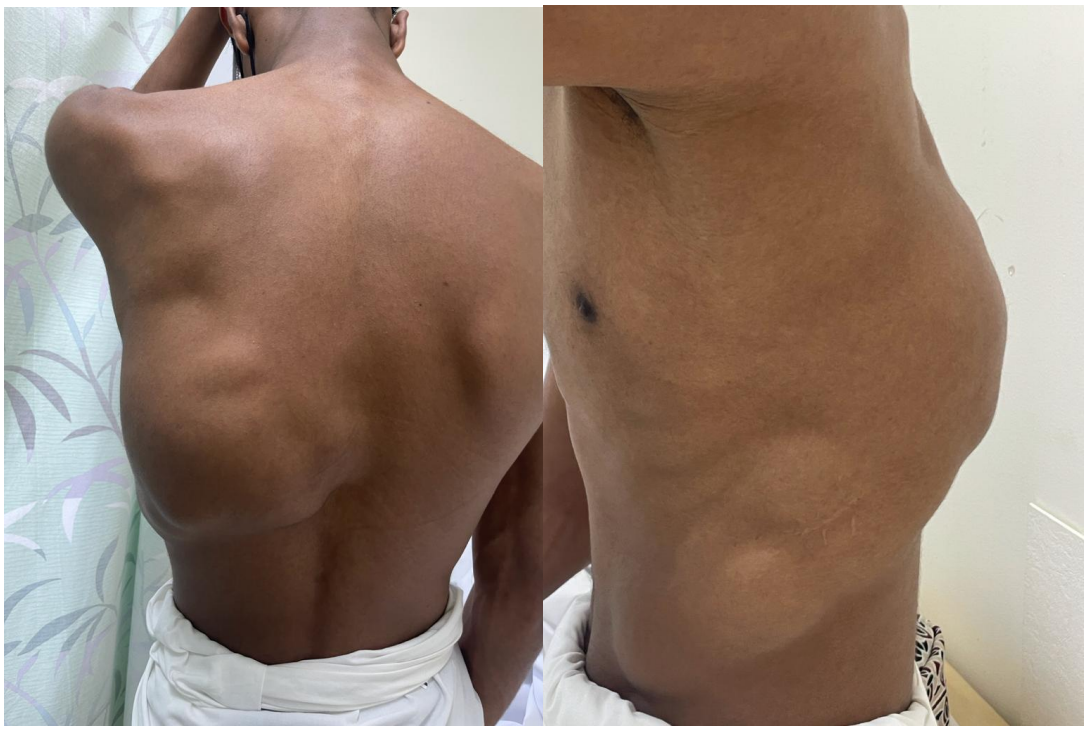

Figure 4: Lymphovascular malformation after the 3de session of sclerotherapy.

Consent statement

Verbal consent was obtained from the patient and his father to publish the images and the clinical findings.

References

1. Mahruqi G, Stephen E, Mawaali H, Al-Musalhi B, al balushi Z, Sukeiti R, et al. Congenital vascular malformations -a quick recap. OMJ . 2020 Nov 19; 10.5001/omj.2021.46.

2. Mulligan PR, Prajapati HJS, Martin LG, Patel TH. Vascular anomalies: classification, imaging characteristics and implications for interventional radiology treatment approaches. BJR. 2014 Mar;87(1035):20130392.

3. Sadick M, Müller-Wille R, Wildgruber M, Wohlgemuth WA. Vascular Anomalies (Part I): Classification and Diagnostics of Vascular Anomalies. Fortschr Röntgenstr. 2018 Sep;190(9):825–35.

4. Sharma M, Mallya V, Khurana N, Kumar P, Duggal R. Lymphovascular Malformation – A Report of Two Cases. J Clin Diagn Res. 2017 May;11(5):ED03–4.

5. Mulligan PR, Prajapati HJS, Martin LG, Patel TH. Vascular anomalies: classification, imaging characteristics and implications for interventional radiology treatment approaches. BJR. 2014 Mar;87(1035):20130392.

6. Sanlialp I, Karnak I, Tanyel FC, Senocak ME, Büyükpamukçu N. Sclerotherapy for lymphangioma in children. Int J Pediatr Otorhinolaryngol. 2003 Jul;67(7):795–800.