

# A Rapidly Enlarging Soft Tissue Lesion of the Back

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**A** 17-year-old male, with no comorbidities, presented with a mass from the left lateral chest extending to the back. At age seven, he had a small mass in the left lateral chest wall at the mid-axillary line which was excised in a periphery hospital (no official medical report). Six years after the surgery, another mass appeared just above the scar of the previously excised mass. The size of the mass increased dramatically over four years; therefore, the patient came to our center. On examination, the swelling was soft, lobulated, cystic, not attached to the overlying skin, extending

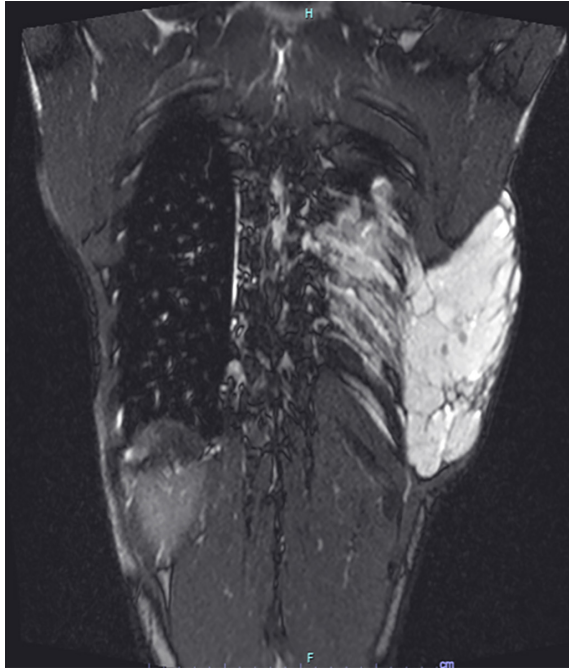
from the left lateral chest to the back, measuring  $13 \times 7$  cm anteriorly,  $7 \times 4$  cm on the lateral chest, and  $28 \times 23$  cm posteriorly [Figure 1]. Magnetic resonance imaging (MRI) revealed a large heterogenous mass measuring  $17 \times 6 \times 20$  cm, extending from the 4<sup>th</sup> to the 10<sup>th</sup> rib involving the left paravertebral muscles [Figure 2].

## Questions

1. What is the diagnosis?
2. What are the MRI findings of this lesion?
3. How will you treat this patient?



**Figure 1:** Showing a lobulated mass extending from lateral chest to the back measuring  $13 \times 7$  cm anteriorly,  $7 \times 4$  cm laterally, and  $28 \times 23$  cm posteriorly.



**Figure 2:** MRI showing a huge heterogenous mass (measuring 17 × 6 × 20 cm) extending from the 4<sup>th</sup> to 10<sup>th</sup> rib and involving the left paravertebral muscles.

**Answers**

1. Lymphovascular malformation.
2. Fat-saturated T2-weighted coronal MRI of the

chest wall showed a large, relatively well-defined solid soft tissue lesion in the left posterolateral chest wall extending to several intercostal and extrapleural spaces. The lesion showed marked T2 hyperintensity with few thin hypointense septa. Few rounded T2 hypointense foci were seen, which corresponds to calcification. No cystic spaces or fluid level and no fat within the lesion were observed. Post-contrast dynamic images reveal delayed heterogeneous enhancement of the lesion but no arterial enhancement, enlarged arterial feeders, or early draining veins. The presence of multiple calcific foci with central lucency within the mass is suggestive of phleboliths. Overall MRI features were 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 (CVMs) and vascular tumors. This classification has been revised by the International

**Table 1:** International Society for the Study of Vascular Anomalies (ISSVA) classification for vascular anomalies approved at the 20<sup>th</sup> ISSVA Workshop, Melbourne, April 2014, last revision May 2018.

Vascular tumors	Vascular anomalies			
	Simple	Combined <sup>o</sup>	Major named vessels	Associated with other anomalies
Benign	Capillary malformations (CMs)	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 syndrome 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; CVM: capillary-venous malformation; CLM: capillary-lymphatic malformation; LVM: lymphatic-venous malformation; CLMV: capillary lymphatic venous malformation; CAVM: capillary arteriovenous malformation; CLAVM: capillary-lymphatic-arteriovenous malformation.*



**Figure 3:** Lymphovascular malformation after the third session of sclerotherapy.

Society for the Study of Vascular Anomalies [Table 1].<sup>1,2</sup> Whereas vascular tumors may regress with age, CVM never regresses on its own. They are subdivided into venous, lymphatic, arteriovenous, and combined malformations.<sup>3</sup> Lymphovascular malformations (LVMs), also known as hemangioma-lymphangiomas, are low-flow vascular malformations that consist of malformed lymphatic channels and account for about 12% of vascular anomalies.<sup>4</sup> They can be classified into macrocystic (> 1 cm), 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 levels.<sup>3</sup> Our patient had a macrocystic type of LVM. Diagnostic modalities for LVM include ultrasound, MRI, and angiography. MRI is considered the best modality because of its soft tissue resolution. LVMs are usually well defined, lobulated, hyperintense on T2-weighted MRI, and hypointense to isointense on T1-weighted images.<sup>2,4</sup> Macrocystic lesions are effectively treated by sclerotherapy, as in our case. Commonly used sclerosing agents are sodium tetradecyl sulfate, 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 each session. The size of the lesion decreased significantly after the third session; 8 × 8 cm anteriorly, 7 × 4 cm on the lateral chest, and 22 × 22 cm posteriorly [Figure 3]. Surgical intervention is reserved for patients that fail to respond to sclerotherapy. In contrast, microcystic LVMs 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% with complete resection.<sup>2,5</sup> Despite the advancements in managing vascular anomalies, CVMs remain a diagnostic and therapeutic challenge to many physicians requiring a multidisciplinary approach.

#### Disclosure

The authors declared no conflicts of interest. Verbal consent was obtained from the patient and his father to publish the images and the clinical findings.

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