Case Report

Imaging of Venolymphatic Malformation in a Child Extending from Inguinoscrotal Region to Thoracic Region: Case Report with Review of Literature

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Abstract

Vascular malformations (VM) are structural malformations of vascular development causing soft-tissue abnormality with functional and esthetic impairment. They are named by their predominant vessel type as arterial, venous, lymphatic, or mixed types. VM extending from the inguinoscrotal to the thoracic region are extremely rare presentation. We present a rare case of veno-lymphatic malformation in the inguinoscrotal region, which is extending superiorly up to the right thorax in a 14-year-old male child who presented with a large swelling in the bilateral inguinoscrotal region and reddish–brown colored skin patches over the right anterior and lateral thoracoabdominal region. The diagnosis was suggested by ultrasonography and confirmed by computed tomography and magnetic resonance imaging.

Keywords: Inguinoscrotal pathology, lymphatic malformations, vascular malformation, veno-lymphatic malformation, venous malformations

Introduction

Veno-lymphatic malformations (VLMs) combine dysplastic lymphatic and venous vascular architecture.[1] These are tumor-like lesions. Based on the primary vessel involved, these lesions can be further divided into arterial, capillary, venous, lymphatic, and combinations of these features.[2] Venous malformations, in particular, are more likely to affect muscle groups in adulthood; however, they can also affect the skin and mucosa and are most commonly located in the head-and-neck region.[3] Clinical manifestations may be an asymptomatic mark at birth to a hemorrhage that poses a serious risk to life. Even the most skilled physician may find it difficult to link any of these incredibly diverse presentations in a patient to a vascular abnormality.[4]

The development of "problematic" vascular abnormalities can cause severe functional and esthetic damage, depending on their size and location.[5]

Veno-lymphatic abnormalities are present at birth, increase in size as the child grows and never involute like other vascular malformations (VM).[6] When more than one vessel type is affected by a malformation, the combined lesions are named after the affected vessel, such as capillary–venous malformation and capillary-VLM.[7]

Since VM are usually detected in children, ultrasound (USG) plays a major role in the diagnosis, classification, and characterization of VM. Furthermore, the primary management of lymphatic malformation is sclerotherapy which can be very well done under USG guidance.[8]

Case Presentation

A 14-year-old male child came to the pediatric outpatient department with complaints of painless swelling in the inguinoscrotal region since birth, gradually increasing in size [Figure 1]. The patient also had reddish–brown colored

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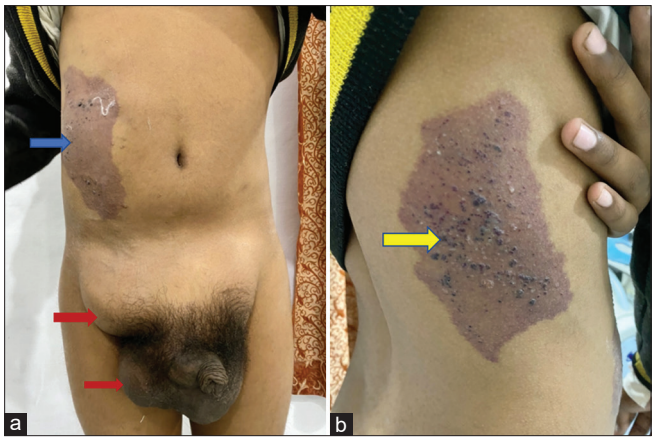

Figure 1: Clinical photograph showing (a) inguinoscrotal swelling (red arrow) with reddish–brown colored patches over the right thoracoabdominal region (blue arrow) and (b) right lateral chest wall (yellow arrow)

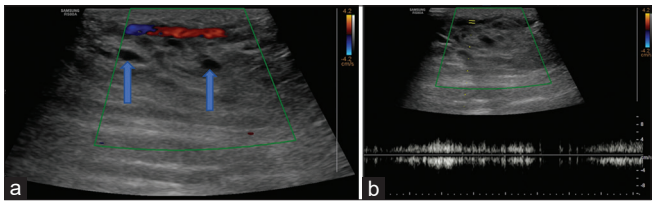

Figure 3: Ultrasound images showing (a) a vessel (red and blue color flow) is seen traversing between the lymphatic channels (blue arrows) in the anterior abdominal wall in the subcutaneous plane, (b) spectral Doppler confirmed it to be a vein, having a monophasic flow

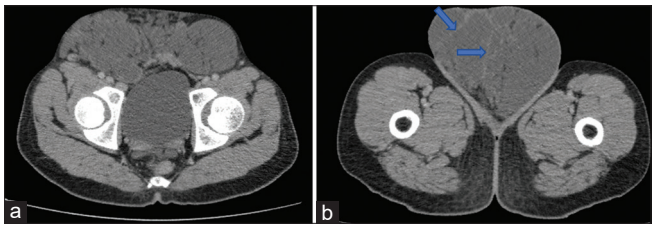

Figure 5: Axial postcontrast computed tomography scan showing dilated lymphatic channels (a) in the bilateral inguinal region and its extension up to scrotum (b) with enhancing septae (blue arrows) within it

patches over the right thoracoabdominal region and right lateral chest wall [Figure 1]. There was no positive family history or medical history. On clinical examination, the swelling was seen extending from the bilateral inguinal region to the scrotum. The swelling was soft, nonpulsatile, nontender, nonfluctuant, and compressible. There was no thrill or bruit over the swelling. There was no associated lymphadenopathy.

On USG by a curvilinear transducer of frequency 1–7A and high-frequency linear transducer of frequency 3–12A, a large well-defined multiseptated cystic lesion was noted in the subcutaneous plane in the pelvic and scrotal region with anechoic channels extending up to the right lumbar and right hypochondriac region, showing no vascularity on color Doppler [Figure 2]. A vein traversed the subcutaneous plane in the right hypochondriac region and extended up to the cystic

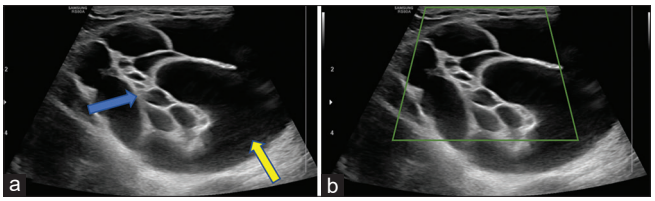

Figure 2: Ultrasound images showing (a) large cystic lesion (yellow arrow) in the inguinoscrotal region with multiple thick and smooth septae within it (blue arrow), (b) on color Doppler, it did not show vascularity suggesting dilated lymphatic channels

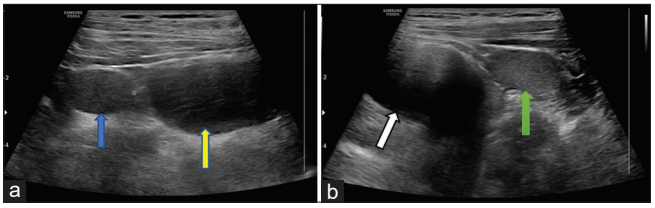

Figure 4: Ultrasound images showing bilateral undescended testes, (a) the right testis (blue arrow) was visualized in the right inguinal region adjacent to the urinary bladder (yellow arrow), (b) the left testis (green arrow) was visualized in the left inguinal region adjacent to the urinary bladder (white arrow)

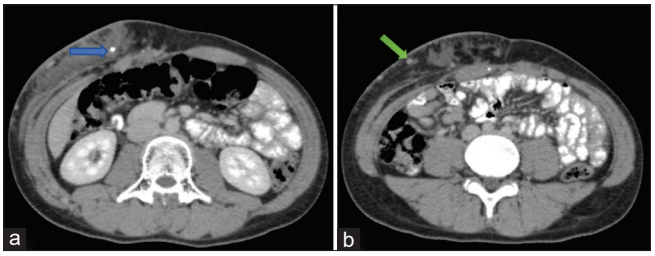

Figure 6: Computed tomography (a) showing calcific focus (blue arrow) suggestive of phlebolith, signifies that it is a venous malformation, and (b) an enhancing vein (green arrow) running through the lymphatic channels in the anterior abdominal wall on the right side

lesion [Figure 3]. The patient also had bilateral undescended testes, which are noted in the bilateral inguinal region just adjacent to the urinary bladder [Figure 4].

USG findings were suggestive of VLM involving the bilateral inguinoscrotal region and extending up to the right thoracic region. The patient was further evaluated by computed tomography (CT) scan [Figures 5 and 6] and magnetic resonance imaging (MRI) [Figure 7] for confirmation of diagnosis with localization and extent of the lesion.

Discussion

VLMs are slow-flow lesions made up of both lymphatic and venous components. Previously, they went by the names lymphangiohemangioma or hemangiolymphangiomas.[1] A histological classification scheme distinguished vascular neoplasm from tumor-like vascular dysplasias and labeled the latter as VM.[2] There are also combined variants, such as arterial–venous–lymphatic and veno-lymphatic abnormalities.[3]

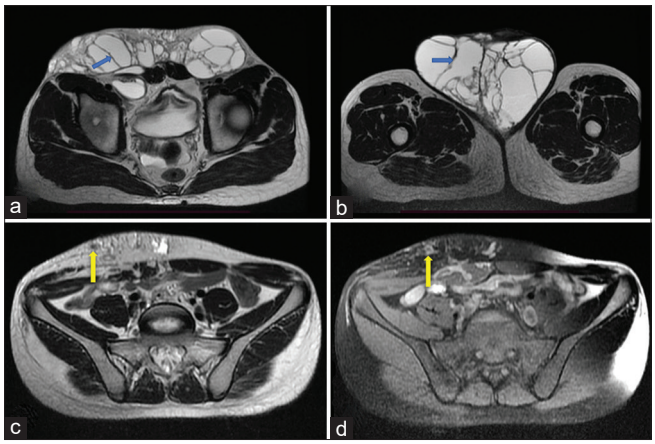

Figure 7: Magnetic Resonance Imaging (MRI) (a) T2 weighted image showing dilated lymphatic channels in the bilateral inguinal region, and (b) extending up to the scrotum with septae (blue arrows) within it. (c) T2 weighted and (d) T1 weighted fat sat images showing flow voids (yellow arrows) in the subcutaneous region surrounding the lymphatic channels suggestive of traversing vessels

Arteriovenous malformations, congenital arteriovenous fistula, and posttraumatic and acquired arteriovenous fistula are some examples of high-flow malformations. Venous malformations, lymphatic malformations, and mixed lesions are examples of slow-flow lesions.[4] Congenital groups of ectatic lymph vessels that create endothelial-lined cystic areas are known as lymphatic malformations.[5] Rare lymphatic–venous anomalies have been observed in the mediastinum, lower limbs, large and small intestine, and craniofacial area.[6] These VM are clinically characterized as soft and movable masses that expand slowly and exhibit few if any, symptoms.[3] When symptoms do occur, they are usually due to the persistent nature of the tumor as well as the size that leads to compression of adjacent structures.

The initial diagnosis has been supported by USG and color Doppler imaging, which demonstrates a slow-flowing lesion, the presence of phleboliths, and a vein that passes through the lesion. MRI has proven to be the gold standard in the diagnosis and to accurately outline the complete extent of the lesion, which is needed to plan the treatment, even though a CT scan with contrast is an option.[1] In addition to analyzing osseous involvement and phleboliths, contrast-enhanced CT and CT angiography also provide information on enhancement, thrombosis, calcification, vascular architecture, and involvement of nearby tissues. US and MRI are the main noninvasive imaging modalities utilized in the assessment of vascular abnormalities to avoid ionizing radiations.[8]

Conclusion

VLMs of the inguinoscrotal region extending up to the thoracic region are very rare lesions and are present since birth with gradually increasing size as age advances. Usually, these lesions are asymptomatic; however, symptoms may occur due to compressive pathologies of such a large-sized lesion. CT and MRI are essential to confirm the diagnosis and the extent of the lesion and invasion to plan the management accordingly.

Declaration of patient consent

The authors certify that they have obtained appropriate patient's guardian consent form. In the form, the guardian has given the consent for the child's images and other clinical information to be reported in the journal. The guardian understands that the child's name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Khatib Y, Dande M, Patel RD, Gite V. Venolymphatic vascular malformation of the parotid gland extending into the parapharyngeal space: A rare presentation. J Oral Maxillofac Pathol 2016;20:308-11.
2. Kadota Y, Utsumi T, Kawamura T, Inoue M, Sawabata N, Minami M, *et al.* Lymphatic and venous malformation or "lymphangiohemangioma" of the anterior mediastinum: Case report and literature review. Gen Thorac Cardiovasc Surg 2011;59:575-8.
3. Fakhry N, Varoquaux A, Michel J, Giovanni A. Venolymphatic vascular malformation in the parapharyngeal space. Otolaryngol Head Neck Surg 2012;147:796-8.
4. Yakes WF. Diagnosis and treatment of low-flow veno-lymphatic vascular malformations. Ces Radiol 2008;62:131-45.
5. Buckmiller LM, Richter GT, Suen JY. Diagnosis and management of hemangiomas and vascular malformations of the head and neck. Oral Dis 2010;16:405-18.
6. Ewing MJ, Zreik RT, Donner LR, Zehr KJ. Large lymphaticovenous malformation resection. Interact Cardiovasc Thorac Surg 2013;17:205-6.
7. Dompmartin A, Vikkula M, Boon LM. Venous malformation: Update on aetiopathogenesis, diagnosis and management. Phlebology 2010;25:224-35.
8. Nosher JL, Murillo PG, Liszewski M, Gendel V, Gribbin CE. Vascular anomalies: A pictorial review of nomenclature, diagnosis and treatment. World J Radiol 2014;6:677-92.