



Subcutaneous Venolymphatic Malformation in the Neck: A Unique Presentation

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Abstract

Venous malformation is one of the most common type of congenital vascular malformation which normally involves the lymphatic system. It is a common type of congenital vascular lesion with recorded incidence of 0.01%, from this 0.01%, 40% is from the neck region, 40% occurs in the trunk, and another 20% in the extremities. The pathogenesis remains unclear. This article elaborates on the type of venous malformation, imaging modality assisting to come to a definitive diagnosis. Surgical excision is the best compared to non-operative method in managing superficial well encapsulated localized venolymphatic malformation.

Keywords: Venolymphatic Malformation; Congenital; Imaging; Surgery

Introduction

Venolymphatic malformation is a congenital lesion presents since birth due to error in endothelial development [1]. The pathogenesis is unclear. Initially classified by Virchow subsequently by Mulliken and Glowacki in 1982 [4], a clinically practical classification system thence supported by biochemical, clinical and radiological studies. This classification has been modified and adapted by The International Society for the Study of Vascular Anomalies (ISSVA) classification system. The venous malformation can be categorized into superficially, subcutaneously as in this case and deep intramuscular lesions. The management of vascular malformation still challenging to highly experienced practitioners. Imaging tools like ultrasounds and magnetic resonance imaging will benefit in determining definitive diagnosis of vascular malformation [1,2]. There are various treatment approach towards vascular malformation

which will be discussed in this article. We would like to present a 14 years old girl with left subcutaneous venolymphatic malformation in the neck and our ENT team approach towards managing this patient.

Case Report

A 14 year old girl, presented with a two years history of left neck swelling measuring 4 cm x 3 cm which was soft ovoid in shape with healed scar over the swelling. Swelling was not fluctuant nor pulsatile. There was no pain or tenderness on palpation. She denied any history of trauma or tuberculosis contact. The swelling gradually increasing in size. There was no underlying skin color changes. Patient claims had similar swelling over the same side since birth where an excision was done at the age of 4 by the surgical team. However unsure of the findings. Patient denied any family history

of malignancy. The unique presentation of this swelling is, it will increase when the patient lies flat and back to normal size when patient stands as shown in figure 1a and 1b.



Figure 1a: Neck mass which not obviously seen while in sitting position.



Figure 1b: Neck mass seen when patient lying flat.

There was no other palpable mass and oropharynx examination was normal.

Ultrasound doppler of the neck revealed low flow vascular malformation with suspicious infiltration of the underlying sternocleidomastoid muscle.

Further assessment with Magnetic Resonance Imaging (MRI) was conducted and suggestive of a left neck subcutaneous venolymphatic malformation measuring 4 x 3 cm with no extension of the lesion to the deep neck spaces (Figure 2a-2c).

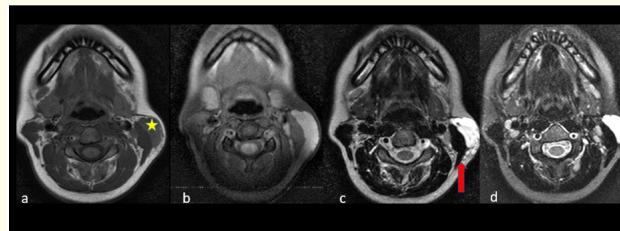


Figure 2a: Images from MRI neck at level of oropharynx; a: T1W, b: T1W with fat saturation, c: T2W, d: T2W with fat saturation. A multilobulated lesion within subcutaneous plane of left neck region adjacent to left sternocleidomastoid muscle. Overlying skin is intact.



Figure 2b: IMRI neck in T2W images; a: In coronal plane, b: in axial plane (red arrow)- showing the hyperintense signal of left subcutaneous neck lesion adjacent to left sternocleidomastoid muscle. No abnormal signal noted within the muscle to suggest involvement.

Figure 2c: MRI neck post contrast with fat saturation; a: In axial plane, b: in coronal plane (red arrow)- showing the hyperintense signal of left subcutaneous neck lesion adjacent to left sternocleidomastoid muscle. No abnormal signal noted within the muscle to suggest involvement.

Ultrasound of the left neck region showed a lobulated soft tissue lesion within the subcutaneous plane. Poor plane seen between the lesion and underlying sternocleidomastoid muscle raising the suspicion of possible involvement. In Doppler mode, no arterial flow noted within the lesion.

MRI neck performed 5 months later showed similar size of the subcutaneous lobulated left neck lesion, measuring approximately

1.3 x 3.6 x 4.7 cm. Lesion demonstrated homogenous T2W hyperintense signal with no significant area of fat suppression. Post contrast administration, heterogenous enhancement of the lesion suggestive of venous component. The non-enhancing component represents lymphatic component. Lesion was seen indenting left sternocleidomastoid muscle with no abnormal signal within the muscle to suggest infiltration/involvement. Overlying skin appeared intact. Adjacent left parotid gland was not infiltrated and no extension of the lesion into deep neck spaces. No abnormal dilated vessels or arterial supply to the lesion appreciated. Based on the imaging features, a slow flow venous malformation with venolymphatic component was suggested.

Excision of the left neck venous lymphatic malformation performed. Intraoperatively noted a subcutaneous mass over the left level II and III, just superficial and not involving the sternocleidomastoid muscle. Specimens as showed in figure 3a and 3b. The length of the lesion was approximately 6 cm in length.



Figure 3a: Left neck venous lymphatic mass excised 6cm in length with Great auricular nerve preserved.

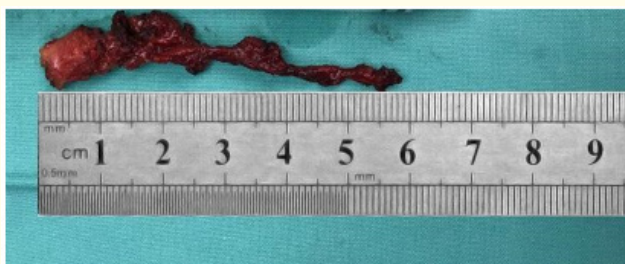


Figure 3b: The lymphatic mass.

Histopathological findings shows a hemorrhagic vascular channel with organized intravascular thrombus and proliferation of fibroblasts. Hemosiderin interposition withing the vessel wall, and the left neck mass in keeping with venolymphatic malformation.

Patient is currently well with no signs of recurrence after 6 months post excision.

Discussion

Venous malformation is a uncommon type of congenital vascular lesion with recorded incidence of 0.01%, from this 0.01%, 40% is from the neck region, 40% occurs in the trunk, and another 20% in the extremities [1]. It can cause serious morbidity and intolerable pain if left untreated [5]. Clinically, vascular malformation appears bluish to purplish as superficial lesions but in our patient there is no skin discoloration. Patient was having intolerable pain on palpation. This patient passes the criteria ISSVA classification system where the swelling was well define, compressible, pain and this swelling was categorized as macrocytic lesion which located subcutaneously [3]. According to studies conducted by Vikula., *et al.* 94% of venous malformation are sporadic type, 5% are non-inherited glomuvenous malformation and 1% dominantly inherited cutaneomucosal venous malformation [3].

Out of these, MRI provides maximum information specially in deep lesions while USG-doppler is the initial investigation [4]. Ultrasound can differentiate vascular tumors from vascular malformations. On Doppler ultrasound, monophasic low flow rate clinches the origin of vascular malformations. MRI helps us to further characterize the size and depth of the lesion which determines the ideal treatment modalities of medical or surgical intervention.

First line treatment for venous malformations are sclerotherapy. Main objective of sclerotherapy is to destroy the vascular channel which affects the endothelium which lead to inflammation and fibrosis. In this case, we proceeded with surgical excision rather than sclerotherapy [3]. The venous malformation in our patient located superficially which have a higher risk leading to skin necrosis and damage to surrounding tissues with worst case scenario higher chances of causing systemic problems which can lead to pulmonary collapse and cardiovascular arrest due to excessive shunting [5].

From the studies conducted, 75% cases with well-defined and localized venous malformation which excised surgically shows

good results, despite chances of significant bleeding from feeding veins or arteries. In our patient there was minimal blood loss [4,5].

Conclusion

Vascular anomalies remain challenging with various therapeutic options. Although the first line treatment for venous malformation is non-operative, those patient with well-defined and localized superficial lesion can be considered for excision for favorable clinical outcomes.

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