Recurrent Cervical Venous Malfomation: An Endless Dilemma

Tekrarlayan Servikal Venöz Malformasyon: Sonsuz Bir İkilem

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ABSTRACT

Venous malformation is considered to be the most common type of congenital vascular malformation with a prevalence of 1%. Diagnosing and managing venous malformation remains a conundrum to attending physician till date. Hereby, we present our experience in dealing with a case of recurrent venous malformation of the neck in a 37-year-old woman, initially treated successfully with sclerotherapy. Patient noticed recurrence 2 years later. Albeit a challenge, recurrent venous malformations in the neck can be treated with multimodal treatment.

Key Words: Venous malformation; recurrence; sclerotherapy; head neck

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ÖZET

Venöz malformasyon% 1 prevalansla en sık görülen konjenital vasküler malformasyon türü olarak kabul edilir. Venöz malformasyonun teşhisi ve yönetimi, bugüne kadar hekime başvurmak için bir muamma olmaya devam ediyor. Bu vesileyle, 37 yaşında bir kadında, başlangıçta skleroterapi ile başarılı bir şekilde tedavi edilen, boyunda tekrarlayan venöz malformasyon olgusuyla ilgili deneyimimizi sunuyoruz. Hasta 2 yıl sonra nüks fark etti. Zor olsa da, boyunda tekrarlayan venöz malformasyonlar multimodal tedavi ile tedavi edilebilir.

Anahtar Sözcükler: Venöz malformasyon; tekrarlama; skleroterapi; baş boyun

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INTRODUCTION

Venous malformation (VM) comprises about 40% of incidence in the head and neck region. It is a benign lesion, with a low flow rate and mostly occurs sporadically. High morbidity and recurrence rate are reported in the literature as it can involve various anatomical spaces and encase important neurovascular structures. Transcutaneous sclerotherapy is the gold standard treatment modality especially the usage of sclerosants like ethanol and sotradecol. Unfortunately, recurrence is quite frequent following sclerotherapy. Hereby, we present our experience in dealing with a case of recurrent venous malformation in the neck.

CASE REPORT

A 37-year-old woman with no comorbid presented to us with 1-year history of painless left neck swelling. According to the patient, the swelling was initially size of a pea however, it progressively increased over the past 1 year. There was no associated obstructive symptoms or any dysphagia or odynophagia. In addition to that, patient claims there were no fever or constitutional symptoms.

Upon examination, patient was sitting comfortable under room air. A non-pulastile left neck swelling was seen occupying level II of neck. It was soft, non-tender, not fixed to underlying structures, measuring 2 x 2 centimetre. No other neck swelling were palpable. Intraoral and otoscopic examinations were unremarkable. Flexible nasopharyngolaryngoscopy performed was normal.

A neck ultrasound was done which was suggestive of a cystic lesion, likely a branchial cyst. Simultaneously, fine needle aspiration cytology performed was unsatisfactory. Subsequently, MRI neck was carried out which showed subcutaneous vascular lesion at the left lateral neck likely a venous aneurysm, originating from the retromandibular vein and draining into the external jugular vein. (Figure 1) Patient was then referred to a vascular center and sclerotherapy was done under general anesthesia for a type I venous malformation. The sclerosing agents used were Thrombovar and Lipiodol. During the procedure, a hypoechoic lesion was seen, which represented multiple venous channels within the subcutaneous tissue at the left lateral neck region. Post-procedure, the swelling reduced progressively in size for the past 2 years.

However, patient claims she started noticing similar swelling over the same site which started to progressively increase in size. (Figure 2& 3) As before, she was asymptomatic. A repeat MRI neck was done which demonstrated a vascular lesion measuring about 2.1x 1.2cm with communication between the superficial part of the lesion and external jugular vein with no filling defect to suggest thrombosis. (Figure 4) Patient was then referred back to the vascular center for intervention. However, she refused to repeat sclerotherapy in view of possible recurrence and is currently under expectant management.

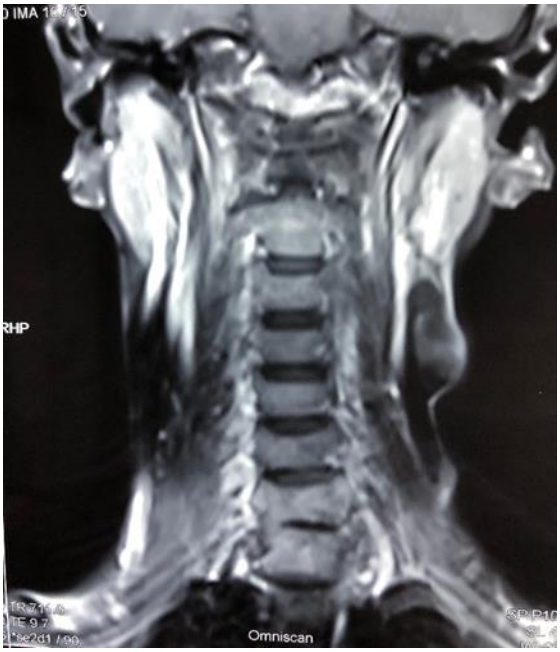

Figure 1 Coronal view of MRI neck prior to first sclerotherapy, showing left neck venous aneurysm

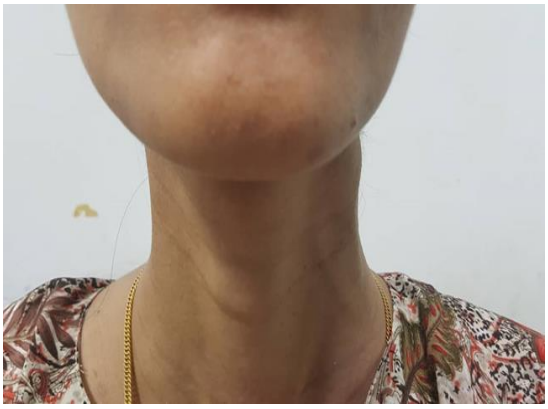

Figure 2 Recurrent, soft,non-pulsatile swelling at left level II of neck (anterior view)

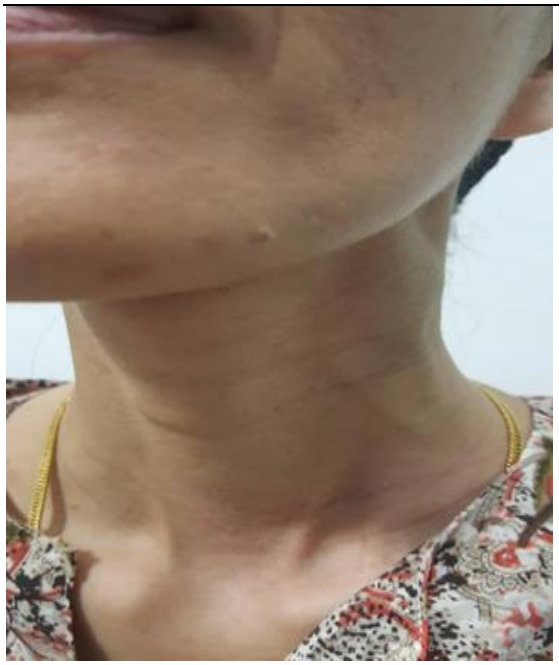

Figure 3 Recurrent, soft,non-pulsatile swelling at left level II of neck (left lateral view)

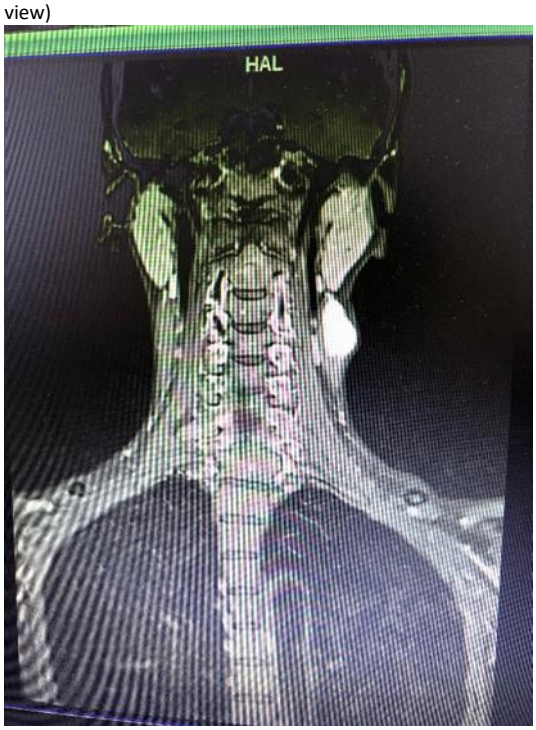

Figure 4 Coronal view of MRI neck showing a recurrent left neck venous aneurysm

DISCUSSION

Vascular malformations can generally be distinguished into arterial, venous, lymphatic, arteriovenous, and also combined according to the Hamburg classification. It can also be subdivided into truncular and extratruncular, circumscribed, and infiltrating malformations (1). However, in 1996, International Society for the Study of Vascular Anomalies (ISSVA) has categorized vascular anomalies into 2 main types; vascular tumours or hemangiomas and vascular malformations. Vascular malformations is then further divided into slow-flow and fast-flow types (2). Generally, 14-65% of cases of vascular malformations develops in the head and neck region.

VM within the head and neck region comprises about 40%. It's a slow-flow type of benign lesion and mostly occurs sporadically (3). The exact pathogenesis of VM however, remains unclear. It is postulated that some developmental defects in the venous system may have caused these malformations. Apart from that, a defect in the angiopoietin receptor namely (TIE) Tyrosine kinase with immunoglobulin-like and EGF-like domains 1 receptor mutation has been proven to cause a few types of venous malformation syndromes. Commonest locations for development of VM includes eyelids, lips, cheek, mandible, tongue, soft palate, neck, floor of mouth and parapharyngeal space. VM can either occur at a single site or multiple sites (3). When multiple site involvement are seen, one must remember to obtain a proper family history to rule out multiple glomangiomas, an inherited autosomal dominant pattern which is also a subtype of VM (4).

VM usually presents as a soft, compressible, non-pulsatile bluish mass. One can observe an expansion of the mass on compression of the jugular vein or upon doing Valsalva's maneuver. Pain is one of the main symptoms of VM of the head and neck region which occurs as a result of spontaneous thrombosis and phlebitic syndrome caused by the venous stasis in the malformed vessels. It is quite easy to clinically diagnose a venous malformation. But, in the head and neck region, imaging is almost always required. B-scan ultrasonography, computed tomography and magnetic resonance imaging are the essential scans. Some of the invasive diagnostic studies includes selective and super-selective angiography and percutaneous phlebography. However, most of the invasive procedures are usually reserved for treatment planning (1).

Management of VM ought to be tailored according to its size, flow-rate, location and the facilities available. Single treatment modality, especially in larger lesions, perhaps will not be able to produce a favorable outcome. In addition to that, as VM of the head and neck region has been reported to have high recurrence rate, a multimodal approach must be undertaken.

VM can either be treated surgically or non-surgically. Conservative measures which can be under taken for smaller, asymptomatic lesions includes elevation of head of bed to reduce long-standing hydrostatic pressure and also warm compression therapy. Transcutaneous sclerotherapy is considered gold standard treatment for VM. Sclerosants like ethanol and Sotradecol was previously used. Recently, bleomycin and OK-432 has been used with successful outcomes. Besides that, laser therapy, cryotherapy, embolization, and surgical excision can be done based on the size of lesion and surgeon's experience(5).

Laser therapy is indicated for the superficial type of VM. Sclerotherapy, on the other hand, is best done for localized, slow-flow VM or as an adjunct pre or post-operatively in large lesions. Besides that repeated sclerotherapy can be done in lesions which are large or polycystic, which might consume a long treatment time. Embolization is suggested prior to sclerotherapy or surgery for those deep and fast-flow lesions. Surgical excision is preferred in lesions which are moderate in size and well-circumscribed. According to a retrospective study done in Japan, 5 out of 67 patients with venous and capillary malformations had recurrent or residual disease (2).

Sclerotherapy being the mainstream treatment modality for VM. Mechanism of action of sclerotherapy is by producing long term fibrosis within the lesion thus preventing any re-expansion of the malformation. Apart from that, sclerotherapy works by damaging the endothelial cells of the blood vessels, accelerating adhesion of platelet cells causing thrombosis and vascular occlusion to develop, and also promotes coagulation of protein in the blood. However, recurrence is quite frequent after sclerotherapy. This could be attibuted by inadequate dosing and lumen recanalization after the formed thrombus gets absorbed(3). This might be the case in our patient. Essential factor to be considered during the injection is that the sclerosing agent remains inside the lesion and does not get washed away by the blood flow. This is possible with the usage of firm agents like sotradecol foam or a type of glue called Ethibloc. Besides that, the injection can also be mixed with fibrin glue, and ethylcellulose (5). The sclerosing agent with the least rate of recurrence is absolute ethanol, but, it comes with significant toxic effects both locally and systemically. Its high toxicity rate is likely due to the high dose of ethanol given for procedures and its fast rate of diffuseness between the tissues (6). Complications which may occur following sclerotherapy includes skin changes in the form of blister, edema, pain and paresthesia over the injection site. Besides that, alarming complications which may occur in 34% of cases includes hemoglobinuria and paradoxical gas embolism which may lead to stroke(7).

On the other hand, bleomycin, which is often used in lymphatic malformations, can also be included in the treatment of venous malformations. The benefit of bleomycin is its low rate of swelling, thus, making it more useful in lesions which are anatomically challenging and delicate.

Laser ablation therapy has shown some promising outcomes in both VM and infantile hemangiomas. Major advantage of this therapy is that it has no effect on the systemic circulation. The mechanism of action of a laser is by forming direct thermal damage to the endothelium, thus, causing obliteration of the vessels by fibrosis. The frequently used laser types are CO2, Nd:YAG and diode(7).

The end result of treatment and its prognosis dependents mainly on the site and size of the malformation. The morbidity and rate of recurrence rises when the lesion is more diffuse and if any pivotal neurovascular or anatomical structures are within close vicinity to the malformation.

CONCLUSION

Recurrent cervical VM can be quite challenging to treat and requires multimodal level of therapy. However, recent advancement in technology namely in the field of imaging and angiogenesis has witnessed more effective treatment with betterment in the outcome of patients.

Conflict of interest

No conflict of interest was declared by the authors.

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