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Venous malformation of External Jugular Vein : A case report

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ABSTRACT

Vascular malformations arising from the wall of the external jugular vein are rare. These distinct clinical entities may be easily and optimally treated with surgical excision without any resultant morbidity. Herein, we report a rare case of venous malformation originating from the external Jugular vein and surgically excised with no recurrence.

Introduction:

Venous malformations may present as isolated neck masses in adults. Cervicofacial venous malformations (VMs) arising from the external jugular vein (EJV) are very rare with less than 10 cases described in the literature.[1] Treatment is sought on account of the swelling for cosmetic reasons. Sclerotherapy is the popular treatment for cervicofacial malformations. The major complications of sclerotherapy include skin necrosis, residual lesions, skin pigmentation, peripheral nerve palsies and hypotensive crisis, as well as fatalities such as cardiac arrest and pulmonary emboli.[1] VMs as distinct clinical entities lies in the fact that they may be easily and optimally treated with surgical excision without any resultant morbidities. Single-stage surgical excision was done in our case with no recurrence.

Case report:

A 45 years old female presented to the Out Patient Department (OPD) of our Institute with complaints of swelling on the right side of neck since 2 years. The swelling progressively increased in size. No fever or other constitutional symptoms were noted. On physical examination, there was a 2-3cm soft, non tender mass in the right posterior triangle of the neck

(Fig.1). It was mobile and not attached to overlying skin or underlying muscle. No pulsation or thrill was demonstrated. There was no other palpable mass or lymph node detected in the rest of the neck. Ultrasonography showed an elliptical, solid, hypoechoic 28mm noncalcified mass inseparable from the left external jugular vein. Computerised Tomography with angiography showed a soft tissue density lesion 28* 20 mm in middle third of right side of the neck (Fig.2). There was no evidence of calcification. Sonography and CT Angiography findings were compatible with the diagnosis of a slow-flow VM intimately connected with the EJV, which demonstrated normal flow. Intraoperatively, VM arising from the EJV was identified. Multiple feeding vessels were identified which were ligated (Fig.3), and lesion completely excised (Fig. 4) with preservation of normal EJV. Macroscopically, the excised lesion was a well-circumscribed and brown in colour. Microscopically, the lesion was composed of ectatic vascular spaces with a thin wall and lined by a layer of endothelium. Smooth muscle was found in the wall of some vessels. Histopathological analysis was compatible with VM.

Discussion:

The nomenclature of cutaneous vascular lesions in the past had always been perplexing, with no consensus until 1982, when Mulliken and Glowacki [2] proposed their revolutionary classification scheme. This scheme, based on endothelial cellular characteristics, correlates well with the biologic behavior of such cutaneous vascular lesions. Vascular lesions can be divided into two major types with distinct clinical characteristics. The first type is designated as hemangiomas. These lesions are present at birth 40% of the time, usually appearing as a small red mark, with a female predilection of 5:1. They typically show rapid neonatal growth characterized by endothelial cellular hyperplasia and proliferation in the proliferative phase, followed by slow regression to a variable extent characterized by diminished cellularity and fibrofatty deposition in the involuting phase.The second type is designated as vascular malformations. These lesions are recognized at birth 90% of the time with no sex predilection. They grow commensurately with the child and do not regress. Histologically they are characterized by “mature” endothelium that are not hypercellular and show a normal endothelial cell cycle. They may have any combination of capillary, venous, arterial, and lymphatic components, with or without fistulas, although most of these lesions are predominantly venous in type. This cell-oriented analysis has since gained wide acceptance because of its diagnostic applicability, which helps in planning therapy. [3,4]

Venous malformations have an incidence of one to two in 10,000 births, prevalence of 1%, are present at birth and grow proportionately with age causing various clinical presentations. Slow-flow VM closely associated with the EJV is rare.[5] They are connected to the EJV by single or multiple veins, mostly of large calibre. Histological abnormalities of the smooth muscle–pericyte component within vascular channel walls of VMs are hypothesised as a potential cause of many VMs. Sclerotherapy has gained popularity over last decade as a treatment in cervicofacial vascular malformations. The most important variables predicting successful sclerotherapy are sclerosant concentration and dwell time, especially when there are multiple wide channels of communication between the VM and its draining vein as in our case.[6] Keeping the risk of skin necrosis, unsuccessful sclerotherapy and proximity of the EJV in mind, surgical excision was planned in our case. Complete surgical excision and subcuticular suturing gave excellent cosmetic results in our patient. 6 months follow up of the patient is uneventful with no recurrence.

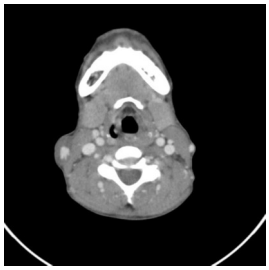

Fig 1 : CT image showing venous malformation

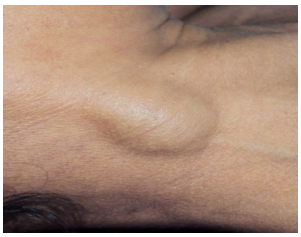

Fig 2: Showing right sided neck mass

Conclusion:

In vascular malformations arising from EJV, the vein is generally normal and can be spared during the surgical excision. Surgical excision of these lesions is simple and straightforward and should be considered as the first line of treatment whenever this clinical entity is recognized. Surgery ensures least down time, morbidity and recurrence rate. Clinicians dealing with vascular problems should be aware of this lesion where early surgical intervention provides optimal results.

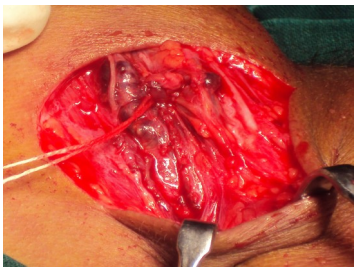

Fig 3 : Showing ligation of vascular malformation

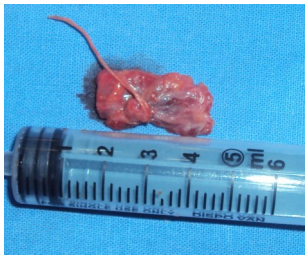

Fig 4 : Excised mass

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