

CASE SERIES AND REPORTS

# Intra-operative sclerotherapy for treatment of a head and neck venous malformation

## *Trattamento di una malformazione venosa del distretto testa-collo mediante scleroterapia intraoperatoria*

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### SUMMARY

Venous malformations of the head and neck are congenital lesions that grow steadily without spontaneous regression. We describe the management of a 47-year-old woman with an extensive subcutaneous venous malformation of bilateral submandibular regions and the entire tongue, refractory to multiple surgical excisions and percutaneous sclerotherapy sessions. The tumour lacked prominent feeding arteries for embolisation, but maintained high blood outflow via a few substantial venous branches. Sclerotherapy to the lesion was prevented by major communicating branches from the mass to the internal jugular vein bilaterally. Our approach entailed direct surgical access to the malformation, ligation of these communicating veins and intraoperative sclerotherapy with ethanol injection into the vessel stumps.

KEY WORDS: Head and neck • Venous malformation • Recurrent • Sclerotherapy • Intra-operative

### RIASSUNTO

*Le malformazioni venose della testa e del collo sono lesioni congenite che crescono costantemente, senza regressione spontanea. Presentiamo il caso clinico di una donna di 47 anni con una estesa malformazione venosa sottocutanea delle regioni sottomandibolari e della lingua refrattaria a più escissioni chirurgiche ed a sedute di scleroterapia percutanea. Il nostro approccio ha permesso un accesso chirurgico diretto alla malformazione, la legatura delle vene comunicanti, e la scleroterapia intraoperatoria con iniezione di etanolo.*

PAROLE CHIAVE: *Malformazioni venose della testa e del collo • Scleroterapia intraoperatoria*

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## Introduction

Venous malformations are an ill-defined subgroup of congenital vascular anomalies that affect one in 22 children, primarily in the head and neck. In contrast with haemangiomas, which involute soon, vascular malformations persist into adulthood, being the source of distressing functional and aesthetic afflictions<sup>1</sup>.

Treatment options for venous malformations classically include compression garments, laser therapy, percutaneous sclerotherapy and surgery<sup>2</sup>. Whereas superficial lesions can be addressed successfully by the majority of these modalities, deeper subcutaneous masses might be amenable to surgical excision only. However, the latter approach is technically challenging, may compromise other vital structures and carries the risk of significant intraoperative haemorrhage<sup>3</sup>.

In the case of the extensive deep head and neck vascular malformation described here, we needed to utilise the combined approach of intra-operative sclerotherapy.

## Case report

Our patient, a 47-year-old female, has been under our care for the past 7 years. She initially presented with a very long history, beginning in infancy, of a large neck vascular mass, extending to the lower face, which did not involute in childhood. She had undergone multiple surgical procedures, including repeated bilateral neck dissections, left parotidectomy and significant tongue resections. In spite of that, she was never disease-free, and presented to our Department with the complaint of progressing swelling and tenderness over the left parotid and upper neck area, as well as the tongue, odynophagia and severe episodic tongue bleeding.

On the initial physical exam, the oral cavity demonstrated notable hypertrophy of the anterior tongue, which was also tethered to the floor of mouth, due to post-operative scarring. Palpation of the neck, apart from extensive scarring, did not reveal any distinct masses, but rather diffuse soft enlargement of the left parotid and upper cervical re-

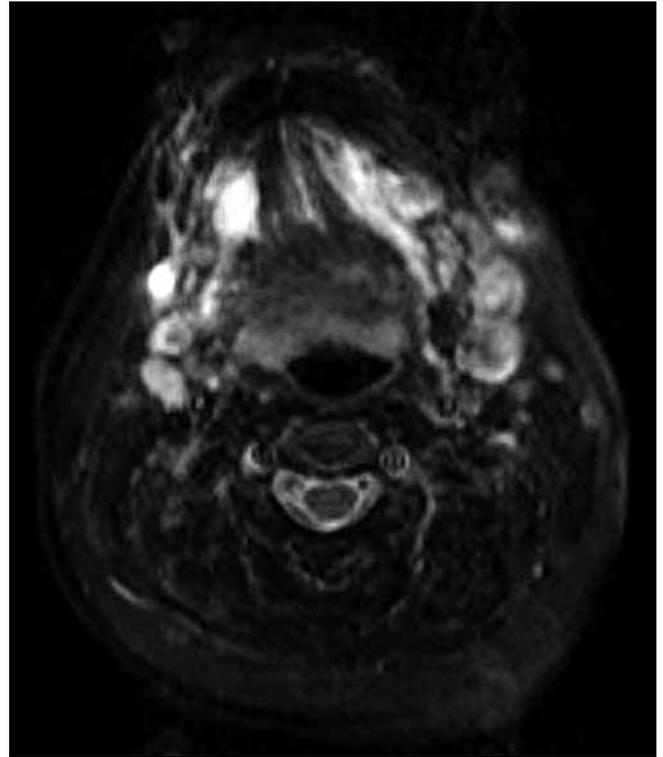
gion. Yet, the affected areas did not show any cutaneous lesions or pigmentation changes.

The initial magnetic resonance imaging (MRI) scan displayed multiple contrast-enhancing lesions, diffusely expanding into the tongue mass, as well as the submandibular and deep parotid spaces, mainly on the left side (Fig. 1). The aberrant vascular network seemed to be adherent to the internal jugular vein (IJV). A computed tomography angiography (CTA) showed a normal arterial branching pattern, but prominent venous drainage during the late phase. Next, we performed a direct venogram, which displayed a large varicose branch, merging into the IJV bilaterally (Fig. 2). Based on the above findings, interventional radiology proceeded with percutaneous sclerotherapy, as no obvious feeding arteries could be identified for embolisation. The sclerosing agent was injected into the submandibular and parotid regions. There was symptomatic improvement for two years, but eventually the swelling and bleeding recurred, and a second sclerotherapy session followed, with new temporary regression of the malformation. Unfortunately, both attempts were limited by the immediate drainage pattern from the lesion to the IJV, via bilateral venous branches. During the past year, the symptoms had been progressing slowly again.

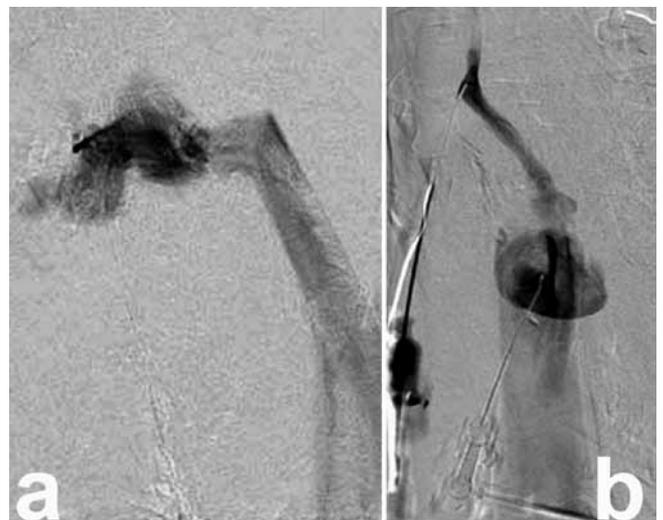
Keeping in mind the multiple attempts to control the disease, both by either surgery or sclerotherapy alone, the diffuse infiltrating spread pattern along the deep and subcutaneous tissues and the large venous trunk draining into the IJV, we chose to perform direct intra-operative sclerotherapy through the main lesion vein, after division of the venous trunk, to avoid infusion of the sclerosing agent systemically. In detail, surgical exploration of the neck revealed the inferior aspect of the vascular mass and its large draining vessel, which was dissected down to its termination into the IJV (Fig. 3a). Three smaller venous branches were traced from the venous malformation to more superior points of entry into the IJV. The four branches were ligated. Then, interventional radiology performed sclerotherapy via the open neck wound. Specifically, into the proximal ligated end of the principal lesion vein, 4 mL of alcohol were injected (Fig. 3b). The entire procedure was also performed on the right side, where three considerably smaller veins than those on the left, were dissected between the vascular anomaly and IJV, and suture-ligated. Subsequently, 3 mL of alcohol were injected into the largest branch. Several minutes later, sclerosis of the vascular spaces was palpable. The patient's post-operative course was fortunately uneventful. Six months later, depletion of the malformation volume could be noted both clinically and radiographically (Fig. 4).

## Discussion

“Vascular anomaly” is a generic, non-descriptive term, embracing a group of diverse lesions, which indicates dis-

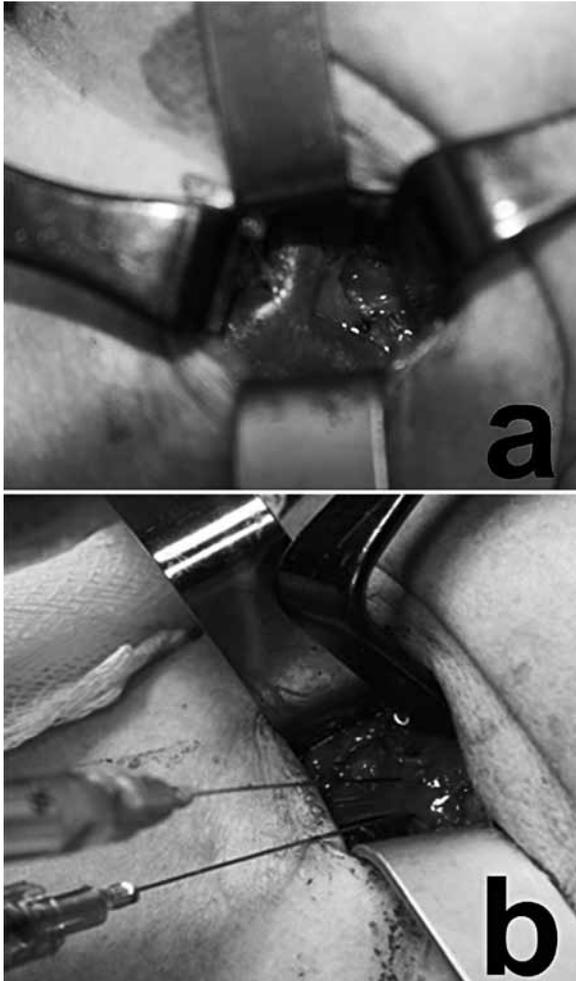


**Fig. 1.** This axial T2 fat-suppressed MRI scan depicts high signal intensity lesions surrounding the ramus and body of the left mandible. The lesion extends into the left lateral tongue. A smaller lesion involves the corresponding structures on the right. This “bunch of grapes” arrangement with septations is pathognomonic of a venous malformation.

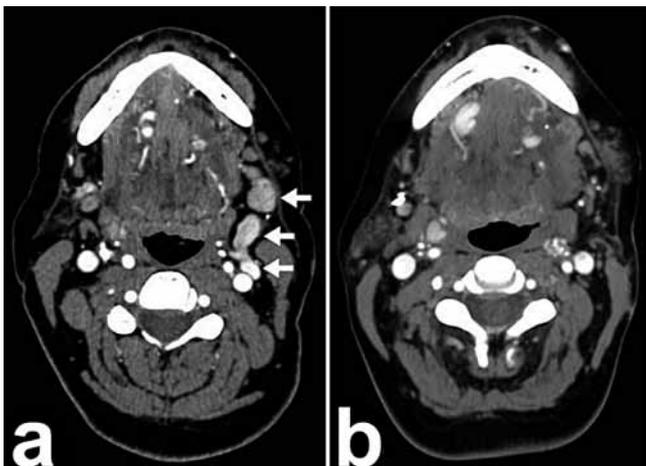


**Fig. 2.** After the percutaneous injection of contrast into the vascular mass, digital venograms show a large varicose venous trunk on each side, draining briskly into the ipsilateral IJVs. The left malformation (a) is larger than the right (b).

tinct approaches. Therefore, a clear-cut classification precludes their misdiagnosis and mismanagement. According to the criteria established by Mulliken and Glowacki, vascular malformations are non-neoplastic, ectatic vessels, of venous, arterial or mixed origin<sup>1</sup>. Their architecture can



**Fig. 3.** Intraoperative snapshots from our approach: (a) The left IJV receives the venous outflow from the malformation, via a large-calibre vessel, which is about to be ligated. (b) After the ligation, ethanol is injected directly into the stump, and disseminates in the circulation of the lesion, to exert its toxic effect on the vascular endothelium.



**Fig. 4.** (a) Axial CTA section before the intra-operative sclerotherapy: large enhancing vascular spaces (arrows) of the left submandibular triangle, which communicate readily with the IJV. (b) Post-treatment CT-image: the terminal venous branch of the malformation has been ligated and the direct injection of the sclerosing agent has led to substantial contraction of the abnormal vasculature.

be either well-defined, producing a sharply demarcated lesion, or as in the case of our patient, might demonstrate infiltrating margins into the deeper structures of the head and neck<sup>4</sup>. Perioral localisation can hamper food intake and permanently deform the lips.

The challenge in the management of such lesions is determining how far we can go with interventions, and which treatment modality is appropriate. Whereas the consensus regarding the treatment of these benign anomalies is to resect the malformation aggressively, for fear of recurrence, the magnitude of some cervicofacial lesions determines a debulking, palliative approach<sup>5</sup>.

Laser photothermolysis is the treatment of choice for mucosal or skin malformations. Subcutaneous masses may respond to interstitial Nd:YAG laser, but vessels deeper than 7-8 mm will not be coagulated sufficiently due to the limited penetration of the light beam. Moreover, when targeting beyond the superficial layers, the risk of injury to the facial and hypoglossal nerves should be considered<sup>6</sup>. High-flow arteriovenous malformations communicate profusely with the arterial tree and are amenable to embolisation either as a monotherapy, or an adjunct to surgery<sup>7</sup>. However, in our patient, no prominent arterial branch was identified angiographically, to be targeted for occlusion.

Percutaneous intra-lesional sclerotherapy is a useful adjunct to surgical resection. The sclerosing agent produces a successful fibrous change in the consistency of the venous anomaly, but only after prolonged contact with the endothelial lining of the vascular spaces<sup>8</sup>. By using novel sclerosing substances, such as sodium tetradecyl sulphate foam, transoral or percutaneous injection, significant regression of the malformation can be achieved in up to 83% of lesions. The advantage of a sclerosant with a foamy texture is its slow-paced clearance from the deposition site<sup>9</sup>. In the case presented here, several wide-calibre vessels, as outlined in the venogram, maintained a brisk out-flow of blood from the malformation to the IJV, thus rapidly clearing the sinusoidal compartment of the sclerosing substance. This also explains the temporary effect of the two sclerotherapy sessions performed in our institution.

Surgical intervention is the definitive treatment for small to moderate venous malformations<sup>10</sup>. In the case presented here, the operation would be a major undertaking with significant morbidity. Taking into account the multi-planar, infiltrating extent of the tortuous lesion, radical resection would require near-total glossectomy and extended submandibular resection. Consequently, the functional restoration of deglutition and articulation, as well as the aesthetic reconstruction would, in all probability, be sub-optimal, even with use of free tissue transfer.

Nevertheless, a minimal surgical intervention provides valuable direct access to the vein trunks of the mass. A sclerosing agent is then injected under direct vision

into the ligated vessels and saturates protractedly the dysplastic venous channels. The concept of depriving a vascular lesion of its main drainage pathways by surgical ligation, while addressing the numerous minute vascular spaces by long-standing sclerotherapy, has been efficiently applied in urology to treat varicoceles<sup>11</sup>. In the head and neck however, to our knowledge, intra-operative sclerotherapy of venous malformations is first reported here. It has successfully treated an aneurysmal cyst of the sphenoid bone, however<sup>12</sup>. Of course, a less complicated alternative to ligating the draining veins prior to the injection of ethanol would be simply to hold pressure transiently on the IJV, allowing the sclerosis to take place. Yet in our opinion, the combination of direct sclerotherapy with the elimination of main flow connections, provides a long-term effect on vascular space obliteration as the dilated malformed veins may not degenerate while still communicating widely with the circulation. The capacity of the sinusoidal vessels<sup>2</sup> and the blood reflux from the IJV<sup>13</sup> would synergistically perpetuate the anomaly.

Naturally, direct injection of ethanol into the bloodstream carries a risk of substantial morbidity, which is already documented in percutaneous sclerotherapy. Local adverse effects include deep venous thrombosis, ulceration and necrosis, while systemic toxicity may escalate to pulmonary embolism and ventricular fibrillation<sup>14 15</sup>. In the case described here, the major outflow pathways were surgically eliminated prior to ethanol administration to avoid rapid washout of the sclerosing substance. We carefully monitored our patient in the ICU for 24 hours, and fortunately her post-operative course was uneventful. Other sclerosants, such as sodium tetracycl sulphate, have demonstrated a safer adverse effect profile<sup>9</sup>.

The case we present demonstrates a multidisciplinary approach to a complex problem. Extended venous malformations which are not amenable to radical excision might be appropriate candidates for intra-operative sclerotherapy. Clearly, the modality described here is not recommended as a first-choice treatment, but rather as a backup technique only in selected cases that are refractory to transcatheter sclerotherapy. It is evident that in every aspect of this approach, close collaboration between the surgeons and interventional radiologists is cardinal.

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