

Young patient with swelling of mandible

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PART A

A 10-year-old boy presented at the Oral and Maxillofacial Surgery Outpatient Clinic with painless swelling of the left mandible. His medical history was unremarkable and clinical examination revealed a soft and compressible lesion with unclear boundaries in the left mandible with slight pink to red skin discolouration.

The lesion enlarged slowly and commensurately with the patient. Due to its increasing size and the correlated cosmetic problems, the patient required medical treatment. Face photo, ultrasound (US), magnetic resonance (MR) imaging and digital subtraction angiography (DSA) images are shown.



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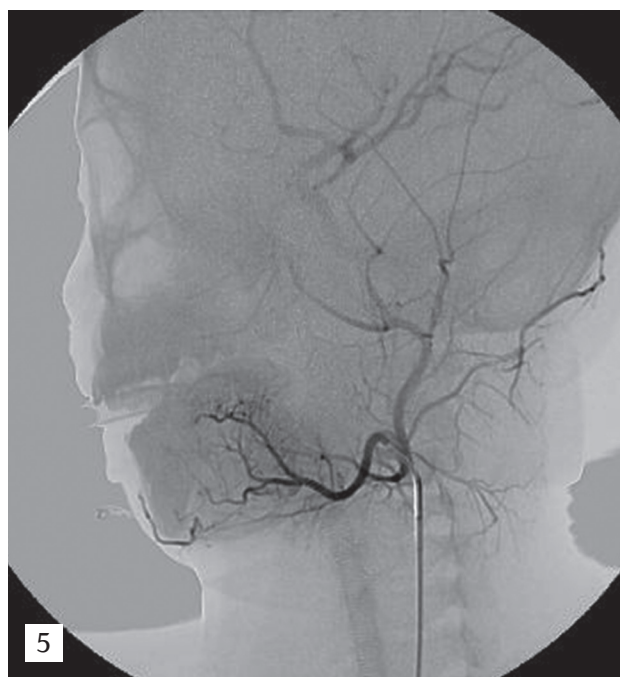
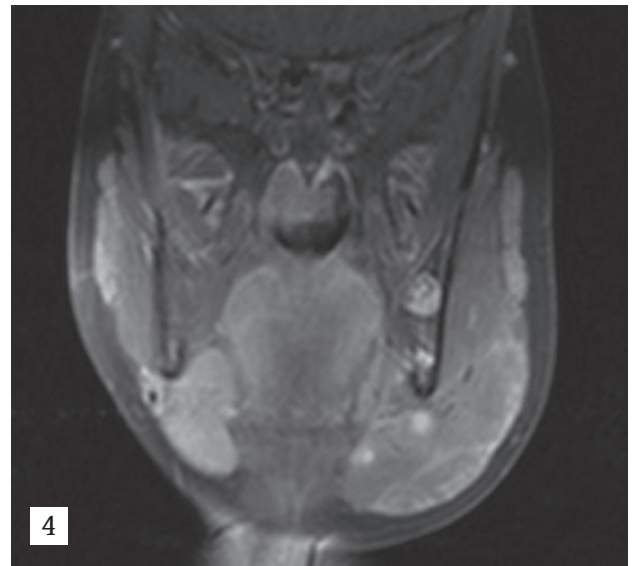
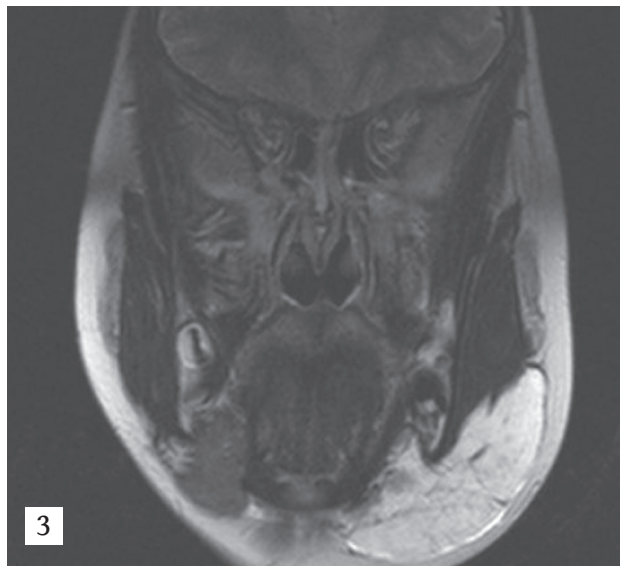
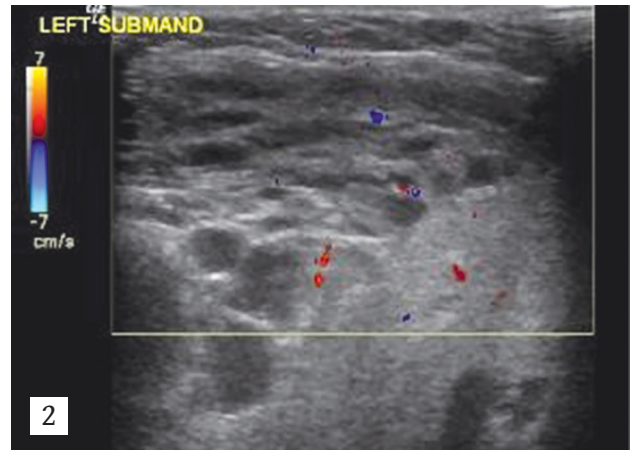


Fig. 1. Face photo

Fig. 2. Colour Doppler US image of the left mandible

Fig. 3. Coronal T2-weighted MR image

Fig. 4. Coronal, delayed gadolinium-enhanced fat-suppressed T1-weighted MR image

Fig. 5. Selective external carotid artery DSA

PART B

Diagnosis: Lymphatic-venous malformation treated with US-guided percutaneous sclerotherapy

Vascular Malformations (VMs) include congenital vascular anomalies of veins, arteries and lymph vessels, which can involve any part of the body. These lesions represent the most common cause of paediatric soft-tissue masses [1]. They are present at birth, but are not always evident. They grow commensurately with the child and often enlarge during puberty. VMs are often bluish, soft, compressible lesions and expand when the affected area is dependent or after a Valsalva maneuver. Enlargement can occur with changes in pressure and flow, collateral formation, shunting, hormonal modulation and infection [2]. The first step in the care of a patient with a VM is to determine whether it is a low- or high-flow lesion, according to its flow dynamics. VMs classified in simple malformations (capillary malformations, venous malformations, lymphatic malformations, arteriovenous malformations, capillary-venous, arteriovenous fistula) and combined malformations such as lymphatic-venous, capillary-venous, capillary-lymphatic-venous, capillary-venous-arteriovenous, capillary-lymphatic-venous-arteriovenous malformations. There are also VMs associated with other anomalies (Proteus syndrome, Klippel-Trenaunay syndrome, Parkes Weber syndrome, Sturge-Weber syndrome, CLOVES syndrome) [3].

Diagnosis and classification of a VM is achieved through clinical history, physical examination and imaging studies such as colour Doppler US (CDUS), MRI, computed tomography (CT) and DSA.

CDUS provides a rapid, cheap and noninvasive initial assessment of the lesion morphology and vascular components. It can also be used to detect arterial flow and distinguish haemangioma from VMs which is important for the subsequent management. Haemangiomas are typically seen as well defined soft-tissue masses with both arterial and venous spectra. Their density is also higher in comparison to low-flow VMs. The distinction between haemangioma and high-flow arteriovenous malformation is critical. The presence of solid tissue in combination with lower mean venous peak velocity are characteristics of haemangiomas which aid the differential diagnosis ver-

sus arteriovenous VMs. On the other hand, low-flow VMs appear as well-defined sponge-like collections of vessels (venous VMs), well-defined collection of cysts and stroma (lymphatic malformation) or isoechoic thickening of skin and subcutaneous tissues (capillary malformation) [4].

MR imaging is the most valuable modality for classification of vascular anomalies. It is also helpful to describe the extension of the lesion and the anatomic relationship to adjacent structures [5]. The protocol should include spin-echo (SE) T1-weighted imaging for basic anatomic evaluation and fast SE T2-weighted or short τ inversion-recovery (STIR) imaging for characterisation of the lesion. Contrast-enhanced MR angiography (using 3D T1-weighted fast gradient echo (GRE) sequence) after intravenous administration of gadolinium is important for the evaluation of the lesion's perfusion. Venous malformations are usually seen as lobulated lesions without mass effect with low signal intensity (SI) on T1WI, high SI on T2WI, no flow voids on SE images, no arterial or early venous enhancement and diffuse enhancement on delayed images. Occasionally, haemorrhage or high protein content may cause internal fluid-fluid levels. Typically, lymphatic malformations are lobulated masses with similar features with venous malformations (low SI on T1WI, high SI on T2WI, no flow voids on SE images). Lymphatic malformations are usually infiltrative and involve multiple tissues. There is rim and septal enhancement in macrocystic lymphatic malformation but no significant enhancement on microcystic ones. As far as capillary malformations are concerned, skin thickening and subcutaneous thickness are the only MR findings [6, 7].

These imaging studies can define the extent and depth of the VM and give us information about the vascularity of the lesion. Nevertheless, the complete characterisation of the angioarchitecture of the VM can only be made with selective or superselective DSA, aiming to exclude any arterial component of the lesion [8].

Low-flow VMs can be treated by percutaneous injection of sclerosing drugs such as ethanol, polidocanol, sodium tetradecyl sulfate (STS), morrhuate sodium and ethanolamine with good to excellent results in the majority of patients. It's a safe, effective treatment that may be repeated if necessary. As for the sclerosing drugs, ethanol is a cheap, easy to obtain and

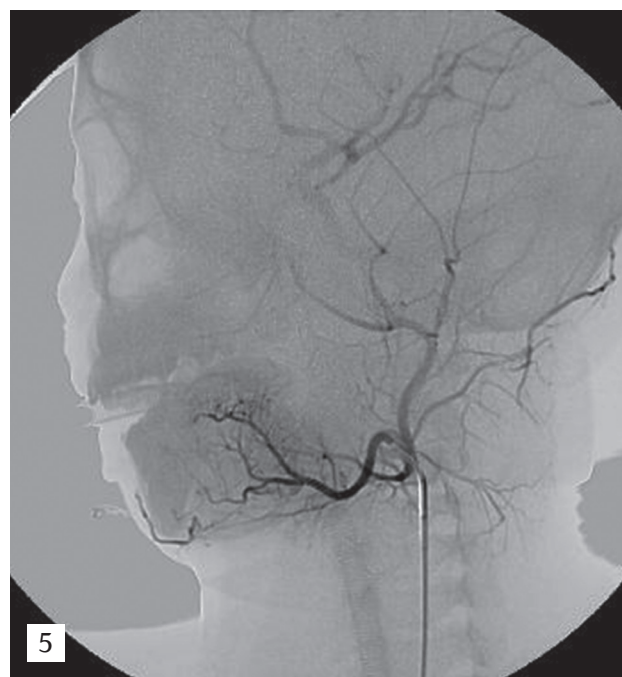
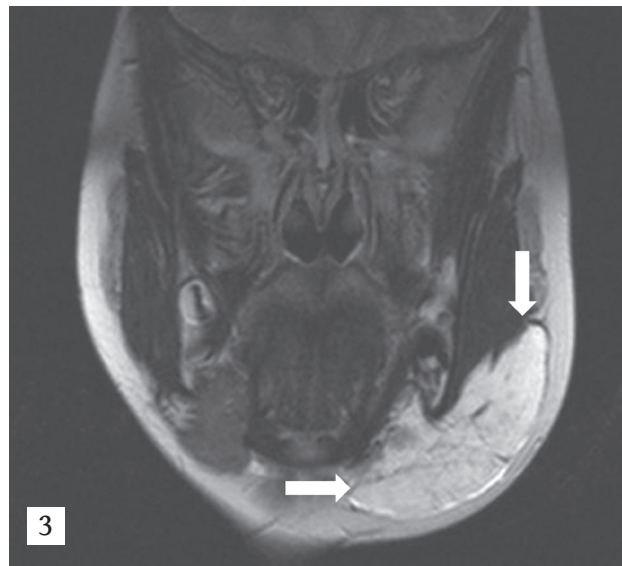
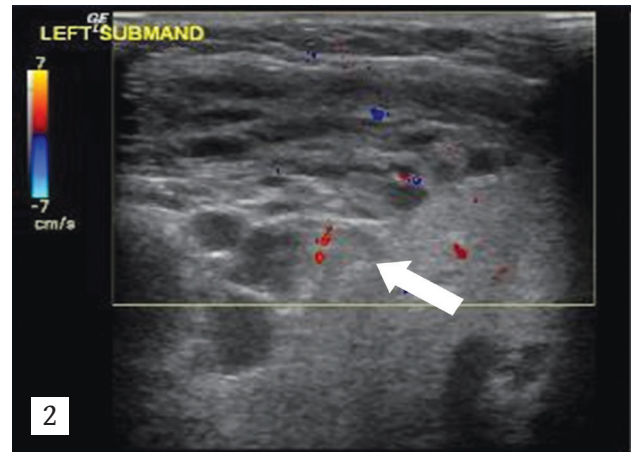


Fig. 1. Face photo shows extensive swelling of the left mandible with slight pink to red skin discoloration (a). Face photo 2 years later, after 3 sclerotherapy sessions shows significant clinical improvement (b)

Fig. 2. CDUS image of left mandible shows an irregular hypoechoic mass with only a few weak signals of flow within the mass. The majority of hypoechoic lesions were easily compressed by the probe pressure

Fig. 3. Coronal T2-weighted MR image shows a well-defined, multilobulated, septated mass with high signal intensity (arrows), characteristic of lymphatic and venous VMs

Fig. 4. Coronal, delayed gadolinium-enhanced fat-suppressed T1-weighted MR image shows partial nodular peripheral enhancement of the lesion (arrows), due to enhancement of the slow-flowing venous channels, characteristic of venous VMs

Fig. 5. Selective external carotid artery DSA excluded the presence of arterial flow in the lesion

rather aggressive sclerosant which causes strong endothelial damage with high response rate but it also results in the most serious side effects (nerve injury, deep vein thrombosis, skin necrosis, muscle contraction). STS solubilises endothelial proteins and induces thrombosis and fibrosis. It's effective and causes less complications in comparison to ethanol. Polidocanol induces rapid overhydration of endothelial cells, leading to vascular injury. The injection is painless and usually without complications. Finally, Ethanolamine Oleate has excellent thrombosing properties and has been used for the treatment of oesophageal and gastric varices. On the other hand, it may induce acute renal failure due to haemolytic effect. The evaluation of treatment effectiveness can be made with clinical examination for superficial lesions of the skin and mucosa or imaging studies such as US or MRI for deep lesions. Localised or limited VMs can be removed surgically whereas large lesions are suitable for partial excision after sclerotherapy decreases the difficulty of the dissection and minimises intraoperative blood loss, which is sometimes life threatening. Venous VMs and macrocystic lymphatic VMs (cysts ≥ 2 cm in diameter) respond well to percutaneous sclerotherapy in comparison to microcystic ones. Percutaneous sclerotherapy is generally a safe and reliable treatment. Skin blister or skin bruise are common complications but usually heal uneventfully with appropriate support care. Rare complications include allergic reactions and mucosal or skin necrosis usually at the site of injection.

In rare cases, nerve (sensory or motor) injuries can occur and can cause facial paralysis [9, 10].

Our patient was evaluated with clinical examination and imaging studies such as CDUS and MRI (Fig. 1-4). His medical history in combination with clinical examination which revealed a soft and compressible lesion with unclear boundaries and imaging studies (CDUS, MRI) suggested the presence of a combined low-flow lymphatic-venous malformation. In order to exclude the arterial component of the lesion, the patient also underwent selective external carotid artery DSA, through the right common femoral artery (Fig. 5). He was treated with percutaneous sclerotherapy using sclerosing foam from a mixture of polidocanol (aethoxysclerol 3%) and gas (liquid to gas ratio 1:4). The lesion was punctured with a 23 G butterfly needle under US guidance. We also used fluoroscopic guidance to prevent extravasation during the injection of sclerosing foam. Three sessions of sclerotherapy (10 ml foam in each session) were necessary to shrink the VM with high technical success and significant clinical improvement. The procedure was carried out under local anaesthesia (lidocaine) for pain control. There were no complications during and after the sclerotherapy sessions apart from bruising of the skin at the 1st postprocedural day, during the 1st sclerotherapy session. Two years later the lesion became clinically undetectable due to the significant reduction of its size. **R**

Conflict of interest:

The authors declared no conflicts of interest.



KEY WORDS

vascular malformation; percutaneous sclerotherapy; polidocanol; ultrasound; DSA

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