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clinical practice

Extracranial association of arteriovenous and venous malformations. Case report

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SUMMARY: **Extracranial association of arteriovenous and venous malformations. Case report.**

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Association of multiple vascular malformations of the face is a rare condition. An arteriovenous malformation (AVM) with a venous malformation as the draining vein is also a rarity.

We report a case of extracranial mixed vascular malformations that deformed the normal architecture of the lower face. Removal of the AVM was followed by stability of the jaw and tongue malformation, indicating the AVM used the venous malformation as its draining vein. This approach spared the patient severe cosmetic and functional sequelae.

RIASSUNTO: **Associazione extracranica di una malformazione arterovenosa con una malformazione venosa. Case report.**

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Una malformazione arterovenosa (MAV) che ha come suo drenaggio venoso una malformazione venosa rappresenta una rarità. Riportiamo un caso di malformazioni vascolari miste extracraniche che hanno deformato la normale architettura inferiore del volto di un paziente. La rimozione della MAV è stata seguita dalla stabilizzazione della malformazione venosa della lingua e della mandibola, mostrando che la MAV ha utilizzato la malformazione venosa come suo drenaggio venoso. Tale approccio chirurgico ha risparmiato al paziente delle importanti sequele sia cosmetiche che funzionali.

KEY WORDS: Arteriovenous malformation - Venous malformation - Mixed malformation - Drainage vein.
Malformazione arterovenosa - Malformazione venosa - Malformazione mista - Drenaggio venoso.

Introduction

Association of multiple vascular malformations of the face is a rare condition. An arteriovenous malformation (AVM) with a venous malformation as the draining vein is also a rarity.

We report a case of extracranial mixed vascular malformations of the face in a young man.

Case report

At 14 year-old Caucasian boy was referred to our department with a long history of a painless, enlarged lower lip due to an extensive vascular malformation involving the lower lip, oral cavity and cheek. Malformation deforms the normal architecture of the lower face.

The patient suffered from dysfunctions in eating, speaking, inhaling and sleeping as well as severe psychological distress. Quality of life, as assessed on a Visual Analogue Scale (VAS), was strongly impaired (VAS=3). Clinical examination revealed a warm, pulsating mass on the lower lip. The surface was bleeding and covered in telangiectasias. Examination of the oral cavity, tongue and cheek showed a cold, bluish-coloured mass which was painless when touched. There was no recurrent spontaneous gingival bleeding (Fig. 1).

Angiography revealed an arteriovenous malformation (AVM) of the lower lip associated with venous malformations in the other districts. Facial magnetic resonance imaging (MRI) showed the mass extended to the pterygo-maxillary fossa and the mandible horizontalis branch, which was without visible deformation. Cerebral MRI did not reveal any additional vascular malformations.

The patient and his parents provided written, informed consent to treatment. As therapy we opted for a combination of embolization and surgery, postponing surgical demolition of the jaw and the tongue. Embolization was performed under sedation in the early morning 2 hours before reconstructive surgery. Microcatheters were inserted into the right facial artery and lower lip and microspheres (Embozene 250 micron) were injected through the catheter to occlude blood vessels. In the first operation (September 2008) the lower lip was removed, together with the AVM and part of the venous malformation. Histopathology confirmed arteriovenous and venous vascular malformations. The left facial artery and vein were isolated at the left

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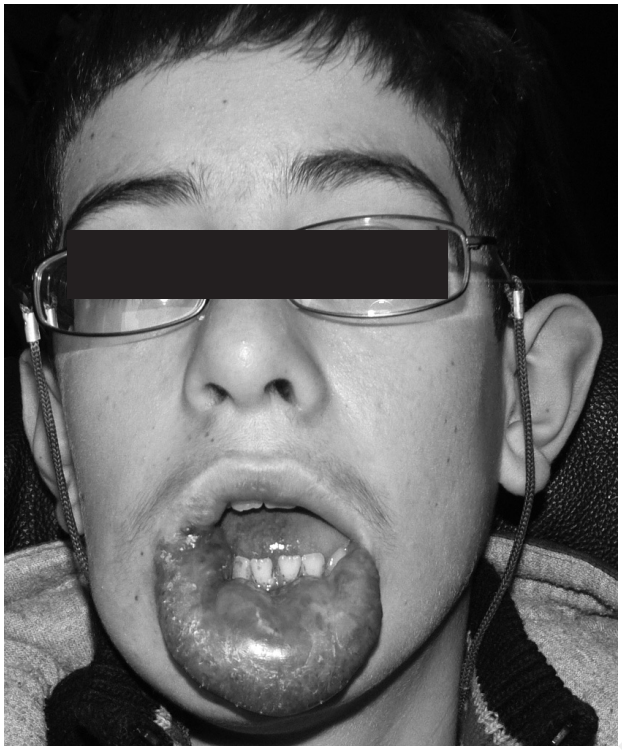

Fig. 1 - Arteriovenous malformation (AVM) on the lower lip associated with tongue and mandible venous malformation.

sub-mandibulary gland. A radial free flap of a 10x7 cm cutaneous paddle with approximately 13 cm vascular pedicle, which was harvested from the left forearm, was used to reconstruct the lower lip and cheek. The left facial artery and vein were anastomosized with the vascular pedicle by carefully tunnelling under the maxilla. A 7cm section of the long left palmar tendon was explanted for reconstruction of the orbicularis oris muscle.

In the second operation (March 2009), the forearm flap skin was shaved down. Subsequently the lower lip underwent tattooing (July 2009). Hypertrophic scars were the only post-operative complication.

The rehabilitation programme included weekly sessions with the psychologist and speech therapist for three months. At present, after 43 months follow-up, mouth anatomy and function are normal (Fig. 2). Computed tomography angiography revealed the venous malformation involving the tongue and mandible was stable with no local recurrence of arteriovenous malformation. Functional results are good, patient satisfaction is excellent and quality of life has markedly improved (VAS=8).

Discussion

This rare case is interesting because an AVM of the lower lip was associated with a venous malformation of the tongue and mandible which may have developed due to venous hypertension which was caused by pre-existing arteriovenous shunting. Preoperative embolization of the arteries has been shown to reduce hypervascularity and significantly blood flow within the vascular malformation. This consequently decreases operative blood loss and allows resection of the lesion (1). As the mandible and tongue vascular malformation was asymptomatic, we decided on a policy of careful observation over time because surgery on the mandible and tongue may be associated with cosmetic and functional sequelae, even though many different techniques have been proposed (2). Removal of the AVM was followed by stability of the jaw and tongue malformation, indicating the AVM used the venous malformation as its draining vein. We believe the venous malformation had developed to compensate for AVM-related disturbances in venous drainage. To date, similar cases have been reported in neurosurgery (3,4). Yanaka et al. described a cerebral venous malformation whi-

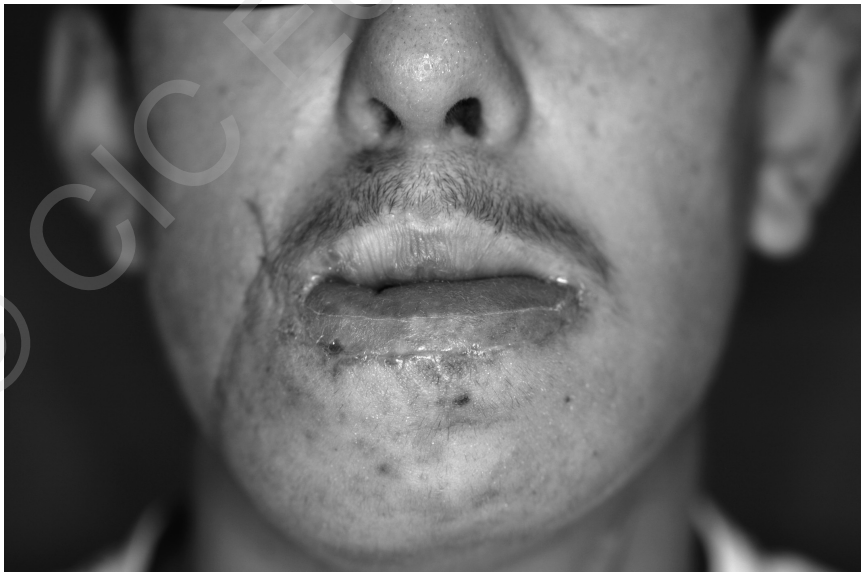

Fig. 2 - Post-operative outcome with lower lip tattoo.

ch served as the draining vein of an adjoining AVM but treated only the AVM. Follow-up magnetic resonance angiography at 6 years after diagnosis demonstrated complete obliteration of the AVM with the venous malformation remaining unchanged. Although most recurrences of AVM develop within the first year of surgery, a minimum of 5 years' follow-up is required to assess long-term control (5). At present with a 43 month follow-up our patient has not exhibited any AVM re-expansion.

Conclusion

This rare case of an extracranial malformation serving as the draining vein of an adjoining AVM was successfully treated by embolization and AVM removal with excellent cosmetic and functional outcome. In managing mixed vascular malformations we recommend careful assessments of options in order to ensure optimal outcome for the patient.

References

1. Kohout MP, Hansen M, Pribaz JJ, Mulliken JB. Arteriovenous malformations of the head and neck: Natural history and management. Plast Reconstr Surg 1998;24:1019-35.
2. Seccia A, Salgarello M, Farallo E, Falappa PG. Combined radiological and surgical treatment of arteriovenous malformations of the head and neck. Ann Plast Surg 1999;43:359-66.
3. Brusati R, Galioto S, Biglioli F, et al. Conservative treatment of arteriovenous malformations of the mandible. Int J Oral Maxillofac Surg 2001;30:397-401.
4. Yanaka E, Hyodo A, Nose T. Venous malformation serving as the draining vein of an adjoining arteriovenous malformation; Surg Neurol 2001;56:170-4.
5. Liu AS, Mulliken JB, Zurakowski D, et al. Extracranial arteriovenous malformations: natural progression and recurrence after treatment. Plastic Reconstr Surg 2010;125:1185-93.