CASE REPORT

Comprehensive Treatment of Upper Lip Arteriovenous Malformation

Mohammad Jafarian¹ · Nima Dehghani² · Shahin Shams³ · Mohammad Esmaeelinejad⁴ · Farzad Aghdashi⁴

Received: 6 January 2015 / Accepted: 30 July 2015 / Published online: 18 September 2015
© The Association of Oral and Maxillofacial Surgeons of India 2015

Abstract Arteriovenous malformations are uncommon congenital disorders in vascular development. They frequently involve craniofacial structures and result in a morphogenic abnormality with ominous arteriovenous shunting. We present a huge AVM of the upper lip in an 18-year-old patient who was successfully treated by the combination method of presurgical endovascular embolization and complete resection of the lesion. Subsequent surgical defect in upper lip, which involved more than two-third of the lip length, was reconstructed via Webster's modification of cheek advancement flap.

Keywords Arteriovenous malformations · Embolization · Resection · Reconstruction · Upper lip

Introduction

Arteriovenous malformations (AVMs) are structural vascular anomalies which occur as the result of errors in morphogenesis of the vessels between 4th and 6th weeks

of gestation [1, 2]. These developmental defects provide unwanted communications between arteries and veins, bypassing the capillary system. Histologically, AVMs are composed of numerous aberrant arteriovenous shunts without normal interconnecting capillary bed and these tangled vessels consist of multiple arterial and venous compartments without muscular support, endothelial proliferation and giant cells. They are usually present at birth but may not be clinically evident; they have a normal growth rate and endothelial turnover, showing proportionate growth in relation to body volume and present no signs of spontaneous involution. This growth is the result of vascular recruitment and collateralization with the acquisition of feeding arteries and draining veins, which conduce to increasing blood flow through the lesion. On occasions, rapid expansion can occur following trauma, infection, and hormonal changes including puberty or pregnancy [3–10].

Generally AVMs are rare lesions and approximately half of them are located in head and neck region. They can be asymptomatic or compress and destroy the surrounding tissue to cause functional and cosmetic problems like facial asymmetry, pain, bone destruction and unexpected hemorrhage. With this circumstance, treatment is inevitable and varies between nonsurgical and surgical modalities including: using sclerosing agent like ethanol or glue, ligation of feeding arteries, curettage and resection. Nowadays, surgical resection following endovascular embolization is the treatment of choice and immediate reconstruction of the remnant defect is necessary [1, 2, 6, 8, 11, 12].

Here, we present a huge AVM of the upper lip which was extended to nose tip and successfully managed by injection of sclerosing agent, embolization and surgery. In this case, upper lip was reconstructed through modified

✉ Nima Dehghani
nimadt2002@gmail.com

¹ Department of Oral and Maxillofacial Surgery, Dentofacial Deformity Research Center, Research Institute of Dental Sciences, Shahid Beheshti University of Medical Sciences, Tehran, Iran

² Department of Oral and Maxillofacial Surgery, Taleghani Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

³ Department of Oral and Maxillofacial Surgery, Qom University of Medical Sciences, Qom, Iran

⁴ Department of Oral and Maxillofacial Surgery, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Webster's cheek advancement flap and cosmetic and functional results were acceptable.

Clinical Report

An 18-year-old girl was referred to our department with huge swelling and malformation of upper lip with extension to nasal columella and tip (Fig. 1). The lesion was present at birth and developed gradually. She had good general health and did not mention any history of trauma to the lip or facial region. On clinical examination, the swelling was firm and painless. Overlying skin and oral mucosa were hyperemic and hyperpigmented with color change to red and bluish pigments. Intraoral evaluation showed anterior maxillary teeth crowding with mild anterior crossbite which was the consequence of compression exerted by this huge lip. In addition, thrill and pulsation were felt on palpation of the lip, so we were conceived to vascular nature of this lesion. Selective angiography for

bilateral carotid arteries showed an AVM of the upper lip with feeding vessels from both right and left facial arteries (Fig. 2). On two sessions with a 1-month interval, these two arteries were embolized and subsequently six intralesional sclerosing agent (TROMBOVAR[®], Therapex Sodium Tetradecyl Sulfate Sclerosing Agent) injections, once in a week, were given. Each injection contained 0.5–1 ml sodium tetradecyl sulfate 1 % solution. Secondary angiograms showed significant decrease in vascularization of the lesion and the patient underwent operation for resection and reconstruction of the upper lip (Fig. 3).

Surgical procedure for resection of the involved site was approached through normal tissue to bypass resection inside vascular tissues. Approximately more than two-thirds of the upper lip was resected and complete hemostasis was performed by ligation and cauterization of the supplying vessels. Reconstruction of such large defect in a young girl is challenging to achieve normal function and the most possible esthetic outcome. We decided to use Webster's modification of cheek advancement flap (Modified Bernard–Burrow's technique) in this case. For this approach, two crescent (Burrow's triangles) excisions in nasolabial region were performed. Cheek advancement flaps were mobilized by dissection near bucco-gingival junction to gantlet in midline. In addition, two other horizontal triangles from superior skin edge (in the junction of new lip and nose) were excised for reduction of excess lip height. Finally, skin, muscles and mucosa were sutured in three layers and columella skin edges were approximated together properly (Fig. 4). The patient's intraoperative and postoperative courses were uneventful and during 2 years follow-up, no recurrence was seen. Functional and esthetic result of this reconstruction method was good and satisfying for the patient (Fig. 5).

Fig. 1 An 18-year-old girl with a huge arteriovenous malformation (AVM) involving the upper lip. **a** Extraoral view. **b** Intraoral view

Discussion

Vascular anomalies are the most common soft tissue congenital abnormalities that affect up to 10 % of newborns. Current classification divides these lesions into two main categories including vascular tumors (predominantly hemangiomas) and vascular malformations. This classification is based on the cellular turnover, clinical behaviors and natural history. Hemangiomas are common vascular lesions with proliferative endothelial cell that produce incomplete vessels architecture. They have rapid growth and involution presented soon after birth and followed by spontaneous resolution afterward. In contrast, vascular malformations have normal cell turnover but comprise progressive dilated and ecstatic vessels. They are generally presented at birth and grow concomitantly with the child. Vascular malformations further subdivide into high-flow

Fig. 2 Radiographic examination of the upper lip AVM before endovascular embolization showing feeding arteries from facial arteries. **a** Three-dimensional computerized tomography (3D-CT) reconstruction with vascular enhancement. **b** Digital subtraction angiograph (DSA)

Fig. 3 Post embolization angiographs showing significant vascularization decrease after endovascular embolization. **a, b** Right external carotid DSA. **c, d** Left external carotid DSA

Fig. 4 Intra-operative photographs. **a** Upper lip AVM and plan for Webster's modification of Bernard-Burrow cheek advancement flap was outlined. **b** Upper lip defect after total resection of the AVM which involved more than two-thirds of the lip length. **c** Excision of

two Burrow's triangles from nasolabial region and mobilization of cheek flap. **d** Excision of another two Burrow's triangles for reduction of upper lip height. **e, f** Inset of the flap by three layers suturing

[arteriovenous malformations (AVMs), arteriovenous fistulas] and low-flow malformations (capillary, lymphatic, venous) based upon the velocity of fluid motion through their system [1, 4, 11–15].

AVMs (unlike hemangiomas) are uncommon lesions and most of them involve head and neck structures. These lesions are among the most serious vascular anomalies due to their high-flow hemodynamic nature which can cause fatal hemorrhage. Their signs and symptoms vary between asymptomatic lesions to pulsatile swelling, throbbing pain, uncontrollable hemorrhage following trauma or tooth extraction and cardiac failure. When the lesions are small and asymptomatic, close observation is sufficient. However, with continuing growth of AVMs, signs and symptoms outbreak and result in cosmetic and functional problems, which necessitate active treatment [1, 6, 7, 15].

Over the past years different treatment modalities were used for management of head and neck AVMs. Considering the benign cellular nature of AVMs, as less aggressive as possible methods were tried for management of these lesions, but unfortunately in many cases they were unsuccessful. Embolization and sclerotherapy techniques are based on using various materials to control blood flow of this obscurant vessels system. These materials consist of embolizing agent: Polyvinyl alcohol foam particles (PVA), Embosphers (like EmboGold), Gelfoam, NBCA-Histoacryl (Glue), OnyxTM liquid embolic system, Coils, Ballons, and sclerosing agents: Ethanol, Sodium tetradecyl, Doxycycline, Bleomycin. But this vascular system has special regrowth potentials including: (1) AVMs have high flow hemodynamic states which can washout sclerosing agents,

(2) AVMs have recruitment and recanalization potential and soon after embolization produce new vascular system, (3) AVMs are fed by more different vessels which are hard to control in a high vascular region such as head and neck. Also, these procedures can be accompanied with some complications like tissue necrosis, thromboembolism, stroke, nerve damage, blindness and severe pain. Nonetheless, their uses were accepted on preoperative adjuvant methods for reducing excessive hemorrhage in surgical operative time and palliative modalities for extensive, deep or inaccessible lesions [2, 6, 9, 10, 16–18].

Frequently, proximal ligation of feeding arteries to the AVMs results in recruitment of collateral vessels and recurrence. Furthermore, this ligation and subsequence collateralization (even more than previous) makes future embolization impossible and aggravates AVM's growth. Yet, ligation as an intraoperative or emergency accessory tool to control unwanted hemorrhage can be used. Also, other less aggressive surgical methods like curettage and partial resection cause reestablishment of the lesion and lead to rapid recurrence, too, and may be accompanied with dangerous intraoperative bleeding. Hence, complete resection is the most curative surgical method for AVMs and is often performed after presurgical selective embolization. This combination method has the greatest success rate and is nowadays considered as a gold standard treatment modality. However, this total resection particularly in head and neck region has some intrinsic drawbacks including remaining surgical defect that may produce significant esthetic and functional problems, and close proximity to vital structures that can restrict proper resection [2, 6, 8–11, 17].

Fig. 5 a, b 2 years follow up visit after surgery showing satisfactory esthetic and functional results

In the head and neck region, AVMs involve cheek, ear, nose, forehead, upper lip, lower lip, mandible, neck, scalp and maxilla by the order. It was shown that the majority of the AVMs occurred in the middle face (cheek, ear, nose, and upper lip) and each of them has specific treatment challenges. Upper lip AVMs have less surgical problems for resection because the lesions and feeding vessels are often accessible and control of hemorrhage is easier. On the other hand, reconstruction of the subsequent defect is critical because the lip has an important role in speech and mastication, and is one of the most important esthetic units in the face [1, 8, 16, 19].

Generally, reconstruction of the upper lip defects is approached by considering defect size and position. Small

defects up to one-third of the upper lip width are primarily closed properly. In some cases with more skin laxity like old patients, up to one-half of the lip can be repaired by primary closure. For defects ranging from one-third to two-thirds of the lip width, cross-lip transposition flaps are appropriate choices. Abbe flap for central and Estlander flap for lateral defects are two famous flaps of this category. Larger defects comprising greater than two-thirds of the upper lip are more challenging because they pose more degree of microstomia, esthetic and functional compromise. Viable options for these defects are cheek tissue advancement flap [Webster's modification of Bernard–Burrow's flap (Webster flap)] and lip flap from remaining lip tissue (Karapandzic flap). Successful result is provided by both techniques if proper case selection is done and advantages and limitations of each one considered. Cheek advancement flap should be used for the patient with enough cheek tissue that can be advanced medially or the laxity of the cheek permits tension free advancement. This flap has the benefit of providing additional tissues for bigger defects and has a mild microstomia. However, it does not provide vermilion tissue coverage, so oral mucosa should be rolled out to meet the external skin edge. Loss of cupid's bow is another disadvantage of this flap but on the other hand, remaining scar is rather smaller. Karapandzic flap has the main advantage of providing innervated tissues and vermilion. However, it can cause microstomia which may need secondary commissure surgery and distortion of commissures. Finally, defects of upper lip more than 80 % and total lip defects need extensive local flap such as bilateral nasolabial flaps or may need free flap transfer such as radial forearm free flap to reconstruct lips and surrounding structure if it is needed [19–24].

In this case we used a new modification of cheek advancement flap for lip reconstruction. In original Webster's modification of cheek advancement flap, just two Burrow's triangles were resected from nasolabial regions to permit flap advancement without any changes in lip height. In this new modification, via addition of two other Burrow's triangles to the horizontal segment of advanced lip or cheek tissues, we can change the vertical height of the lip. The base of this two added triangles were placed in midline of the lip to create natural curve of the lip.

Conclusion

Upper lip AVMs like most head and neck counterparts can be treated successfully by combination of presurgical embolization and complete resection. Up to now, this method is the most predictable treatment modality, but subsequent surgical defect needs precise reconstruction procedures. It is critical to preserve esthetic and functional

demand in the facial region which can greatly influence social and personal activities of the patient.

Compliance with Ethical Standards

Conflict of interest All authors declare that they have no conflict of interest.

References

- Kohout MP, Hansen M, Pribaz JJ, Mulliken JB (1998) Arteriovenous malformations of the head and neck: natural history and management. *Plast Reconstr Surg* 102:643–654
- Lemound J, Brachvogel P, Götz F, Rücker M, Gellrich NC, Eckardt A (2011) Treatment of mandibular high-flow vascular malformations: report of 2 cases. *J Oral Maxillofac Surg* 69:1956–1966
- Motamedi MH, Behnia H, Motamedi MR (2000) Surgical technique for the treatment of high-flow arteriovenous malformations of the mandible. *J Craniomaxillofac Surg* 28:238–242
- Ethunandan M, Mellor TK (2006) Haemangiomas and vascular malformations of the maxillofacial region—a review. *Br J Oral Maxillofac Surg* 44:263–272
- Kademani D, Costello BJ, Ditty D, Quinn P (2004) An alternative approach to maxillofacial arteriovenous malformations with transosseous direct puncture embolization. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 97:701–706
- Behnia H, Jafarian M, Dehghani N, Dehghani S, Seyedan K (2014) Comprehensive treatment and rehabilitation of a patient with maxillary arteriovenous malformation. *J Craniofac Surg* 25:e463–e467
- Bhandari PS, Sadhotra LP, Bhargava P, Bath AS, Mukherjee MK, Maurya S (2008) Management strategy for facial arteriovenous malformations. *Indian J Plast Surg* 41:183–189
- Taskin U, Yigit O, Sunter VA, Albayram SM (2010) Intraoral excision of arteriovenous malformation of lower lip. *J Craniofac Surg* 21:268–270
- Oka H, Pogrel MA, Dowd CF, Lee JS (2010) Treatment of arteriovenous malformation of the mandible with resection and immediate reconstruction. *J Oral Maxillofac Surg* 68:658–663
- Qu X, Su L, Wang M, Fan X (2013) Two-year follow-up of osseointegration and rehabilitation in a patient with oral and maxillofacial arteriovenous malformations. *Int J Oral Maxillofac Surg* 42:1079–1082
- Chen WL, Ye JT, Xu LF, Huang ZQ, Zhang DM (2009) A multidisciplinary approach to treating maxillofacial arteriovenous malformations in children. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 108:41–47
- Richter GT, Suen JY (2010) Clinical course of arteriovenous malformations of the head and neck: a case series. *Otolaryngol Head Neck Surg* 142:184–190
- Connor SE, Flis C, Langdon JD (2005) Vascular masses of the head and neck. *Clin Radiol* 60:856–868
- Buckmiller LM, Richter GT, Suen JY (2010) Diagnosis and management of hemangiomas and vascular malformations of the head and neck. *Oral Dis* 16:405–418
- Elluru RG, Azizkhan RG (2006) Cervicofacial vascular anomalies. II. Vascular malformations. *Semin Pediatr Surg* 15:133–139
- Kiyokawa K, Takagi M, Fukushima J, Kizuka Y, Inoue Y, Tai Y (2005) Surgical treatment following huge arteriovenous malformation extending from the lower lip to the chin: combination of embolization, total resection, and a double cross lip flap. *J Craniofac Surg* 16:443–448
- Hohlweg-Majert B, Metzger MC, Schramm A, Berlis A, Schön R (2008) Complication of intraoral procedures in patients with vascular tumors. *J Craniofac Surg* 19:816–819
- Manganaro AM, Will MJ, Rafetto LK (2000) Rapidly enlarging lesion of the upper lip. *J Oral Maxillofac Surg* 58:883–887
- Anvar BA, Evans BC, Evans GR (2007) Lip reconstruction. *Plast Reconstr Surg* 120:e57–e64
- Harris L, Higgins K, Enepekides D (2012) Local flap reconstruction of acquired lip defects. *Curr Opin Otolaryngol Head Neck Surg* 20:254–261
- Mandrekas AD, Page R, O'Neill TJ (1994) Upper lip reconstruction with a radial forearm free flap. *J Craniofac Surg* 5:105–109
- Matin MB, Dillon J (2014) Lip reconstruction. *Oral Maxillofac Surg Clin North Am* 26:335–357
- Sun G, Lu M, Hu Q (2013) Reconstruction of extensive lip and perioral defects after tumor excision. *J Craniofac Surg* 24:360–362
- Brinca A, Andrade P, Vieira R, Figueiredo A (2011) Karapandzic flap and Bernard–Burrow–Webster flap for reconstruction of the lower lip. *An Bras Dermatol* 86:S156–S159