ANNALI ITALIANI DI CHIRURGIA
Pubblicazione on-line
ISSN 2239-253X
Direttore Nicola Picardi

Disfiguring facial hemangioma compromising labial functionality

A case report

Ann. Ital. Chir.
Published online (EP) 9 April 2013
pii: S2239253X13020847
www.annitalchir.com

Simona Rinaldi, Antonio Rossi, Andrea Conversi, Marco Toscani, Mauro Tarallo, Pasquale Fino, Nicolò Scuderi

Department of Plastic, Reconstructive and Aesthetic Surgery, "Sapienza" University of Rome, Policlinico "Umberto I", Rome, Italy

Deturping facial hemangioma compromising functionality. A case report

Hemangiomas are proliferative lesions characterized by increased endothelial cell turnover.
Lip hemangiomas can distort lip anatomy and are at increased risk of ulceration and/or bleeding, which lead to impaired function and disfigurement. Surgery can provide active treatment but it can be problematic because vermilion tissue is unique and not found elsewhere on the body. Since there is no similar tissue that can easily be used for reconstructing missing vermilion, its preservation is essential for a satisfactory cosmetic result. We present the case of a 74 year old woman suffering from a large hemangioma of the mid-face and particulary with an angiomatous ulcerated lesion at the level of the lower lip. We describe clinical management and surgical treatment of this pathology.

KEY WORDS: Bleeding lip hemangioma; Facial hemangioma; Neoplasm embolization; V-shaped full-thickness lip excision

Introduction

According to the classification of Mulliken and Glowacki[1], hemangiomas are vascular lesions that show increased endothelial mitotic activity and cellular proliferation. They are the most common tumors of infancy, occurring in up to 12 percent of Caucasian children by the age of 1 year [2], and may occur in various anatomic sites, the most common being the head and neck [1-3]. Usually 80% of all hemangiomas are single lesions, but 20% of affected infants develop multiple tumors. Hemangiomas typically manifest themselves with a phase of rapid proliferation in the first year of life, followed by a slow, gradual involution over the next 5-7 years[1]. Based on this natural tendency to regress, conservative management with frequent observation is usually proposed. However, complications, including ulceration, that occurr in up to 15% of lesions[17], bleeding, infection, pain and functional impairment of adjacent structures, as well as psychological problems arising from the presence of large proliferative hemangiomas, are certainly indications for treatment. Moreover the presence of such tumours on the face, particularly when the eyelids, lips or nose are involved, may result in severe consequences, which cannot always be controlled by conservative therapy and may require active treatment.
Hemangiomas of the lip are of special concern, as they occur in a prominent location on the face. Lip hemangiomas can distort lip anatomy and are at increased risk of ulceration and/or bleeding, which lead to scarring, contour deformity, and disfigurement[14].

Pervenuto in Redazione Ottobre 2012. Accettato per la pubblicazione Gennaio 2013
Correspondence to: Pasquale Fino M.D., Department of Plastic, Reconstructive and Aesthetic Surgery, University of Rome " La Sapienza", Policlinico Umberto I, Viale Pantelleria, 35, Scala B, Interno 1/A, 00141, Rome, Italy. (e-mail: pasquale.fino@gmail.com, pasquale.fino@uniroma1.it)

This can be problematic because vermilion tissue is unique and not found elsewhere on the body. Since there is no similar tissue that can easily be used for reconstructing missing vermilion, its preservation is essential for a satisfactory cosmetic result. Hemangiomas must be differentiated from macular stains, vascular malformations, and other vascular tumors of infancy. This may be done in more than 93 percent of cases on a clinical basis, but radiographic studies and biopsy may be necessary in equivocal cases[15].
We present the case of a 74 year old woman suffering from a large hemangioma of the mid-face and particulary with an angiomatous ulcerated lesion at the level of the lower lip.
We describe clinical management and surgical treatment of this pathology.

Case report

In June 2011, a woman of 74 years of age was brought to our attention in the Department of Plastic and Reconstructive Surgery of Policlinico Umberto I of Rome, affected by a voluminous hemangioma of the lower lip. This lesion was ulcerated and bleeding profusely (Fig. 1).
The patient reported that the angiomatous lesion has been present since birth, and involved much of the left half of the face. The ocular, nasal and oral region were extensively affected (Fig. 2).
At the age of 12 and 14 years she underwent surgery to remove the lesion and reconstruct the face with skin grafts taken from the thigh and left forearm.
The patient, due to a significant bleeding, performed embolization of the lesion on the lower lip about three years ago, with substantial resolution of the clinical problem.
She comes to our attention as the lesion of the lower lip was bleeding again.
Our team defined a surgical strategy to remove the lesion and use the procedure of embolization of vascular afferents to reduce intra-operative bleeding.
Selective arterial catheterization of the external maxillary artery was performed (Fig. 3) and embolized in order to obtain the exclusion blood of the lesion of the lower lip (Fig. 4).

Fig. 1: Voluminous hemangioma of the lower lip.

Fig. 2: Facial hemangioma.

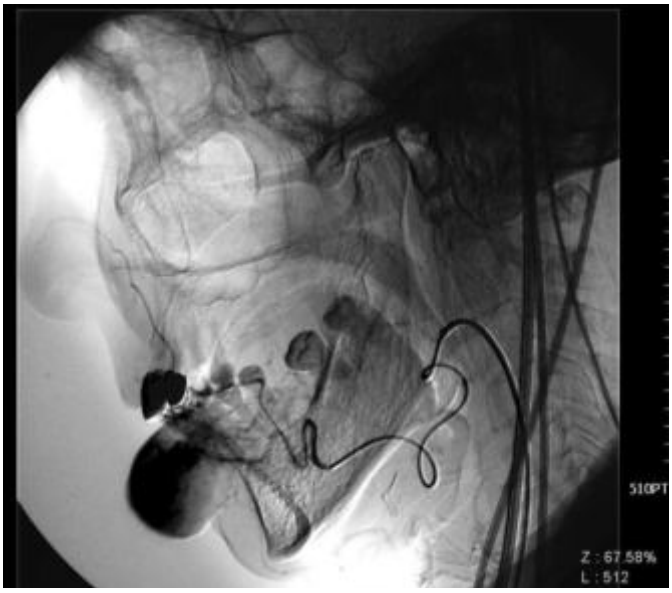

Fig. 3: Selective arterial catheterization of the external maxillary artery.

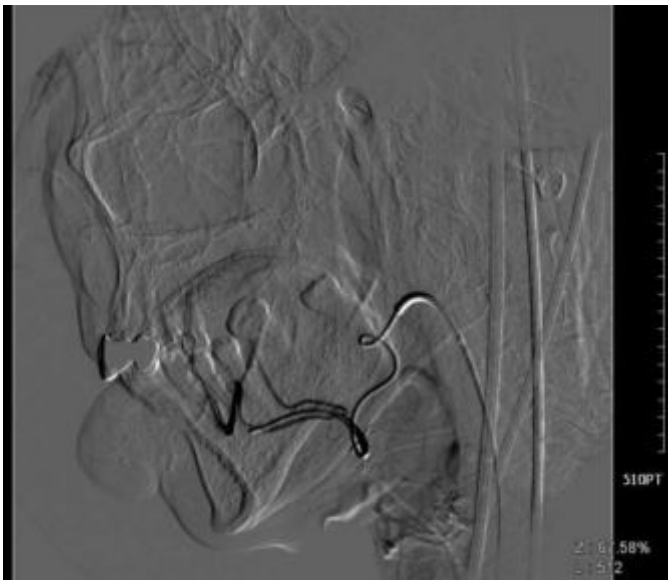

Fig. 4: Embolization of the lesion on the lower lip.

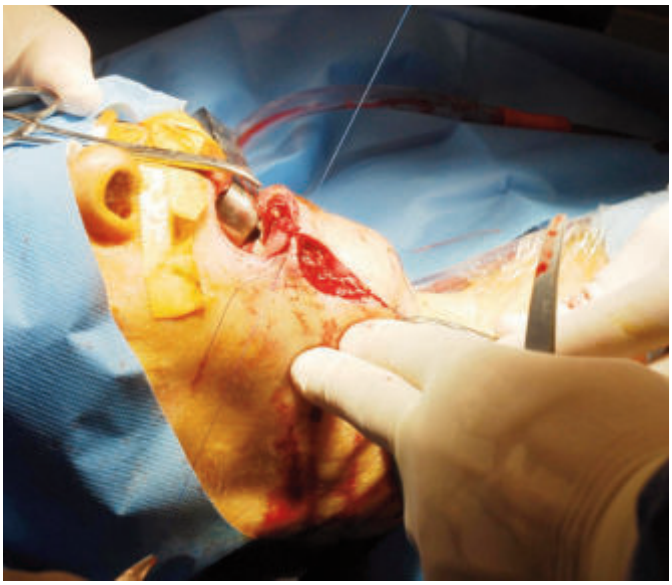

Fig. 6: Primary wound closure.

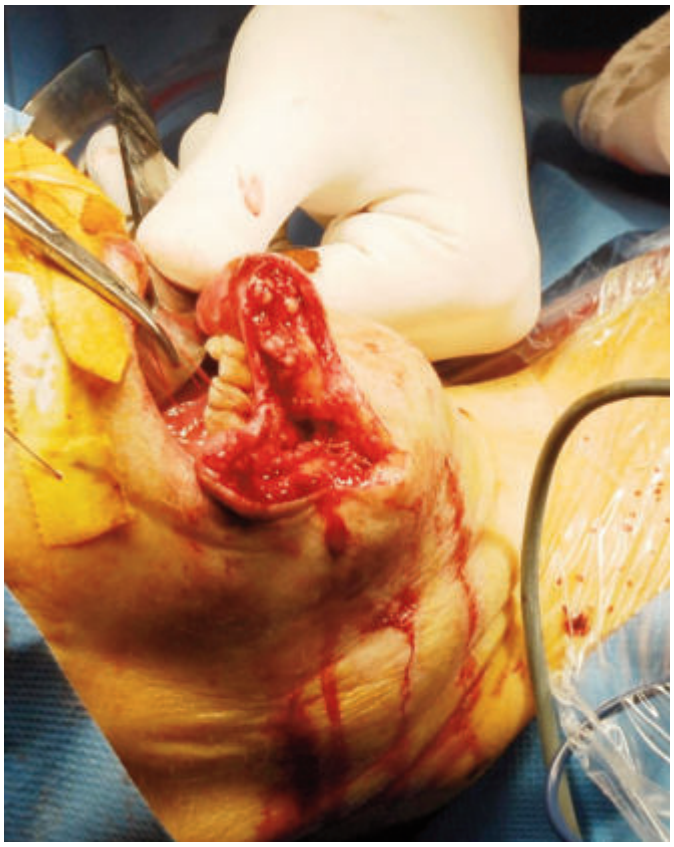

Fig. 5: V-shaped full-thickness lip excision.

Fig. 7: 12 month postoperative result.

The following day the patient had the lesion surgically removed. The excision of the lesion was carried out using full-thickness V-shape technique (Fig. 5), accurate hemostasis and suture with 2/0 - 3/0 Vycril and 4/0 Nylon (Fig. 6).

The postoperative course was uneventful and the patient had immediate and complete relief of her symptoms. The cosmetic result was good and the patient satisfied (Fig. 7).

Discussion

Hemangiomas are proliferative lesions characterized by increased endothelial cell turnover.

An understanding of vascular lesions has been greatly facilitated by the work of Mulliken and Glowacki, who developed a biological classification of vascular anomalies encompassing physical findings, clinical behavior and cellular kinetics. According to their classification there are two types of vascular lesions: hemangiomas and vascular malformations. Two main characteristics distinguish these two types. While hemangiomas are not usually present at birth but become apparent within the first weeks of life, vascular malformations are present at birth but may not be clinically evident. Furthermore, hemangiomas show rapid proliferation during the first two years of life, followed by a slow involution. In contrast to these characteristics, vascular malformations show proportionate growth in relation to body volume and show no signs of spontaneous involution.

According to Mulliken[2], hemangiomas can be classified into "capillary", "cavernous" and "capillary cavernous" types. Capillary hemangiomas are superficial hemangiomas, originating from the papillary dermis, and were often called "strawberry" hemangiomas in the past. Cavernous hemangiomas are located within the reticular dermis or subcutaneous tissue and may appear bluish or colorless. Capillary cavernous hemangiomas originate from the papillary and reticular dermis or subcutaneous tissue as in the case of our patient.

Since all hemangiomas belong to a single entity, Waner and Suen[5] proposed a more practical classification of hemangiomas based on their skin depth. In this latter system, capillary hemangiomas are listed as "superficial hemangiomas", cavernous hemangiomas are replaced by "deep hemangiomas" and capillary cavernous hemangiomas are classified as "combined hemangiomas".

Histologically, hemangiomas are characterized by endothelial hyperplasia with or without vascular lumina and an increased number of mast cells during the proliferative phase; fibrosis, fatty infiltration, diminished cellularity, and a normal mast cell count are characteristics of the involutive phase [1-4].

In most instances the diagnosis of common hemangioma can be established on the basis of the patient history and physical examination. A history of early onset followed by rapid proliferation in early infancy is characteristic of hemangiomas. At times it will be difficult to distinguish a "field" hemangioma from a port-wine stain. Careful observation over several weeks will demonstrate that the hemangioma is proliferating. Subcutaneous hemangiomas are sometimes difficult to separate from lymphatic malformations of the cystic type. While lymphangiomas will not involute, sometimes the physician or parents would like to know prior to the age of resolution. Ultrasound, computed tomography (CT) scans, and magnetic resonance imaging (MRI) are helpful in separating these two entities [6-7]. In the proliferative phase, CT and MRI demonstrate hemangiomas as well-circumscribed, densely lobulated, uniformly enhancing lesions with dilated feeding and draining vessels in the center or at the periphery. The measurement of urinary levels of basic fibroblast growth factor may be helpful in distinguishing hemangiomas from structural malformations[8]. Measurement of this factor, which is elevated in hemangiomas, can also be used to monitor involution in select circumstances.

Histologic examination (biopsy) can be useful in select circumstances when the diagnosis of hemangioma is uncertain. Cutaneous rhabdomyosarcoma, nasal glioma, hemangiopericytoma, tufted angioma, KHE, dermoid cysts and some types of histiocytosis can all mimic the clinical appearance of common hemangiomas.

The natural history of these lesions is quite good; it is estimated, however, that up to 10% of hemangiomas can cause complications requiring active treatment. These include those patients for which involution has not afforded a satisfactory result.

Recommendations can be made even if the goal is to "wait and watch" for natural involution of the hemangioma. There are many treatments available, such as compression therapy, intralesional or systemic corticosteroids injection.

Interferon alpha-2a is a potent inhibitor of angiogenesis that has been used successfully in patients with aggressive hemangiomas that are refractory to systemic corticosteroid therapy [12] .

Various lasers have been used to treat hemangiomas. One of the strongest indications for laser use is the presence of ulceration. Ulcerative hemangiomas respond nicely to the pulsed dye laser if the ulcerative component of the hemangioma is limited and the rate of proliferating growth is mild [13]. Other approaches are the use of CO_2 laser or cryosurgery that enjoyed widespread use in the past and has been recently revived for the treatment of hemangiomas, particularly early superficial cutaneous lesions.

The most common complication of hemangioma is ulceration [15], which occurs in less than 5 percent of cases and often involves perioral and perineal lesions[16]. Treatment consists of wound care and topical antibiotic ointment. Excision of ulcerated lesions may be necessary. Indication for surgical excision can include hemangiomas with incomplete resolution, severe disfiguring facial lesions, those located in the periorbital area, nose, mouth, upper airway, ear canal or those lesions that pose a threat to the child's health. Patient with hemangiomas of the lip or nasal tip frequently undergo surgery prior to starting school because of the slow involution and high visibility of hemangiomas in these locations[10].

The operative technique depends on the location and size of the lesion, and focuses on resection of the tumour and reconstruction of the adjacent structures when necessary.

The lips form a distinct anatomic unit that is the principal feature of the lower face. Reconstruction of the lip can present a challenge to the reconstructive surgeon who seeks excellence in restoration of the aesthetics and functions of this unique structure.
Wedge or V-shaped full-thickness lip excision and primary wound closure has been the choice of treatment in our case.
The V-shaped excision is designed in such a way that it extends the complete height of the lower lip from the vermilion to the mental crease. In the lateral regions of the lip, the V is skewed to produce a more angulated wound closure line matching the natural relaxed skin tension lines of the lateral lip.
Optimal primary repair of full-thickness lip defects require approximation of at least four tissue layers: mucosa, muscle, subcutaneous tissue including deep dermis, and epithelium of the skin.
The anterior vermilion line is the principal landmark of the lip. To insure symmetry, its precise location on opposing edges of the wound should be identified early in the course of lip repair. Once identified, the line is marked in some way as a reference to help guide the rest of the lip repair. As the two borders of the lip defect are approximated, accomodation for the disparity of the width of the vermilion is accomplished by forward advancement of vermilion of the lateral lip segment and simultaneous slight posterior advancement of vermilion of the medical border of the wound.
The tendency toward development of a retracted and depressed lip scar is appreciably reduced when the wound is repaired with accurate approximation of the lip muscle.
Embolization of hemangiomas can be employed alone or, more commonly in conjunction with pharmacologic or surgical therapy[11]. A common indication is hepatic hemangiomas. After angiography, absorbable or permanent particles are introduced into feeding vessels under fluoroscopic guidance by an experienced surgeon or radiologist. Complications include cerebral vascular accidents and skin necrosis caused by accidental or overzealous obstruction of some arteries. Embolising materials can be substantially subdivided into liquid and non-liquid. The fluids polymerize in contact with blood, and become, after the addition of radio-opaque substances visible to X-rays; one advantage of these substances is, besides being a fluid which can be easily injected, the possibility of regulating polymerization time. The non-liquid or corpuscular materials are divided in re-absorptive and no re-absorptive according to their composition. The study of embolising materials is in continual evolution as are the techniques and new approaches. Embolization differs from sclerotherapy, in that ethanol is commonly used as a sclerosant. Injection of ethanol does not require angiography, and complications, including cerebral vascular accidents ,are unlikely.
It is important to remember that large cervicofacial lesions have been associated with other malformations in 10 percent of cases. These comprise the PHACES syndrome (P, posterior cranial fossa malformations; H, hemangiomas; A, arterial anomalies; C, cardiac anomalies; E, eye anomalies; S, sternal cleft)[9]. The causative event is unknown, but it occurs between 8 to 10 weeks of gestation. Anomalies include the Dandy-Walker malformation, absence of carotid/vertebral vessels, bifid or cleft sternum, and supraumbilical raphe. The vast majority of affected patients are girls (9:1 ratio), who are especially prone to occlusive cerebrovascular accidents at an early age [14].

Conclusion

In conclusion, surgery is appropriate in the management of facial haemangiomas causing functional disturbance or serious psychological distress. Proliferative labial tumours, which are prone to bleeding and cause difficulty while eating, require surgical correction. Treatment goals include strategic placement of the incision and restoration of normal lip contour. The procedure of embolization can be used to reduce intra-operative haemorrhage. Surgery can provide active treatment with satisfactory results and minimal morbidity. Early surgical treatment is also recommended for nasal-tip haemangiomas, which regress very slowly and may result in severe distortion of the cartilaginous framework, and large periocular haemangiomas, to prevent secondary amblyopia.

Riassunto

Gli emangiomi sono lesioni proliferative caratterizzate da un aumento del ricambio delle cellule endoteliali.
Emangiomi delle labbra possono distorcere l'anatomia del labbro e sono ad alto rischio di ulcerazione e/o sanguinamento, che portano alla compromissione della funzionalità ed alla deturpazione. La chirurgia può fornire un trattamento efficace ma può essere un problema perché il tessuto del vermiglio è unico e non reperibile in nessun'altra parte del corpo. Poiché non vi è tessuto simile che possa essere facilmente utilizzato per la ricostruzione del vermiglio mancante, la sua conservazione è essenziale per un risultato cosmetico soddisfacente. Presentiamo il caso di una donna di 74 anni affetta da una grande emangioma della metà del viso ed in particolare con una lesione ulcerata angiomatosa a livello del labbro inferiore. Descriviamo la gestione clinica ed il trattamento chirurgico di questa patologia.

References

1. Mulliken JB, Glowacki J: *Hemangiomas and vascular malformations in infants and children: A classification based on endothelial characteristics.* Plast Reconstr Surg, 1982; 69:412-22.

2. Mulliken JB, Young, AE: *Vascular Birthmarks: Hemangiomas and Malformations.* Philadelphia: Saunders, 1988; 24-103.

3. Achauer BM, Chang CJ, Vander Kam VM: *Management of hemangioma of infancy: Review of 245 patients.* Plast Reconstr Surg, 1997; 99:1301-308.

4. Finn MC, Glowacki J: *Congenital vascular lesions: Clinical application of a new classification*. J Pediatr Surg, 1983; 18:894.

5. Waner M, Suen JY: *A classification of congenital vascular lesions. In: Waner M, Suen JY (eds) Hemangiomas and vascular malformations of the head and neck*. New York: Wiley-Liss, 1999; 1-12.

6. Burrows PE, Laor T, Paltiel H, Robertson RL: *Diagnostic imaging in the evaluation of vascular birthmarks.* Dermatol Clin, 1988; 16:455-88.

7. Greenspan A, McGahan JP, Vogelsang P, Szabo RM: *Imaging strategies in the evaluation of soft-tissue hemangiomas of the extremities:correlation of the findings of plain radiography in 12 histologically proven cases*. Skeletal Radiol, 21:11-8, 1992.

8. Chang E, Boyd A, Nelson CC, et al.: *Successful treatment of infantile hemangiomas with interferon-alpha.2b*. J Pediatr Hematol Oncol, 1997; 19:237-44.

9. Frieden IJ,reese V, Cohen D: *PHACE syndrome. The association of posterior fossa brain malformations,hemangiomas, arterial anomalies,coarctation of the aorta and cardiac defects, and eye abnormalities.* Arch Dermato, 1966; 132:307-11.

10. Pitanguy I, Machado BH, Radwanski HN, Amorim NF: *Surgical treatment of hemangiomas of the nose*. Ann Plast Surg, 1996; 36:586-92.

11. Argenta LC, Bishop E, Cho KJ, Andrews AF, Coran AG: *Complete resolution of life-threatening hemangioma by embolization and corticosteroids.* Plast Reconstr Surg, 70:739-44, 1982.

12. Blei F,Orlow SJ, Geronemus RG: *Interferon alfa-2a therapy for extensive perianal and lower extremity hemangioma.* J Am Acad Dermatol, 1993; 29:98-99.

13. Morelli JG, Tan OT, Yohn JJ, et al.: *Treatment of ulcerated hemangiomas with the pulsed tunable dye laser*. Am J Dis Child, 1991; 145:1062.

14. Gorlin RJ, Kantaputra P, Aughton, DJ, Mulliken, JB: *Marked female predilection in some syndromes associated with facial hemangiomas*. Am J Med Genet, 1994; 52:130.

15. Drolet BA, Esterly NB, Frieden JJ: *Hemangiomas in children*. N Engl J Med, 199; 341:173.

16. Mulliken JB: *Intralesional photocoagulation of periorbital hemangiomas (Discussion)*. Plast Reconstr Surg, 1999; 103:17.

17. Thomas RF, Hornung RL, Manning SC, et al.: *Hemangiomas of infancy: treatment of ulceration in the head and neck*. Arch Facial Plast Surg, 2005; (5):312e5.

18. Demiri EC, Pelissier P, Genin-Etcheberry T, et al:. *Treatment of facial haemangiomas: The present status of surgery*. Br J Plast Surg, 2001; 54(8):665e74.