Surgical Treatment of Giant Frontonasal Infantile Haemangiomas

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Abstract
Infantile haemangiomas, also known as “haemangiomas of infancy,” (IHs) are common benign tumors of endothelial cells characterized by a unique pattern of rapid proliferation that occurs in the first months of life, followed by slow involution that may take years to complete. They reportedly occur in as many as 12% of children and are frequently brought to the attention of plastic surgeons, dermatologists, and paediatricians. Although most are ultimately of little significance, a portion have the potential concern to produce complications. Hemangiomas of the fronto-nasal region are frequent, and create scare to parents and doctors for reasons related to their considerable size, the critical reduction of the visual field, the ulceration, the risk of direct trauma and critical bleeding, and not least the activation of psychological aspects, if you opt to drug therapy for a conservative approach. The authors present some cases of giant hemangioma of the fronto-nasal region, surgically treated at an early stage, with scar contained in the aesthetic units where appropriate, with satisfactory clinical results appreciated by the parents of the young patients.

ABBREVIATION
IH: Haemangioma of Infancy

INTRODUCTION
Vascular tumors in newborns are largely made up of the Infantile hemangiomas (IH), characterized by a very fast development just a few weeks after birth [1-4]. After rapid growth phase, generally it follows a slow apoptosis of vascular tumor, which gradually turns in a soft fibro-adipose tissue and redundant skin [5]. The fibro adipose tissue may be more or less abundant, in relation to the onset of the lesion volume. In particular anatomical areas, such as the frontal and periorbital region, the size cannot be in itself significant, and binding in addressing the decision about surgical excision [6]. The reason is that even small lesions can cause extensive damage difficult to repair of noble structures such as the eye and the periorbital region, but also the nose and the lip. Early surgery is mandatory mainly when these lesion are huge [7,8] the involvement of the visual field is significant, and the will of the parents is to solve the problem as quickly as possible. Periorcular IHs warrant special consideration because failure to treat or inappropriate treatment may lead to permanent visual damage. This approach minimizes the risks associated with severe visual axis impairments such as amblyopia, or related late sequelae such as strabismus, belated refractive deficiency or dysmorphic eyelid cosmetically disfiguring [9]. In this context we frame massive infantile hemangiomas, called “giants” characteristically affecting the periorbital tissue, the frontal region and the root of the nose. The anatomical topographical distribution is not random, and follow the lines of embryological development as described by some authors [10,11].

MATERIAL AND METHODS

Some cases surgically treated from 2010 to 2016 at early stage (mean age 2.2 years) are presented below. They come all from monocentric series of the Vascular Anomalies Center at Bambino Gesù Children’s Hospital in Rome. Patients had never been subjected to drug therapy (propranolol, steroids, interferon), or interventional angioradiology (schleroembolization), nor to the use of beta blockers for topical use (Timolol). The patients did not undergo blood transfusions neither during nor after the surgical procedure. The images refer to an intermediate follow-up running 2-4 years after surgery. For none of them have been performed minor surgical procedures yet or ancillary techniques after the primary surgery.

Case 1
D.E: Mixed IH of the glabellar region, seemingly harmless in respect of visual function. Actually an ophthalmological
and orthoptic examination had highlighted already an initial convergent strabismus of the right eyeball, so it was decided to fulfill a surgical excision. The lesion is of small size, and although not framed as a giant hemangioma, shows that even small lesions can be considered to interfere with early visual function. The choice to a vertical scar is due mostly to the dislocation of the tumor on the paranasal right side. This approach allows a better distribution of redundant asymmetric tissue, and at the same time a complete removal of the subcutaneous and skin portion of the lesion (Figure 1,2).

Case 2

A.G.: Giant frontonasal IH of the frontal region and right eyebrow. Very rapid growth with slight push on the right eyeball outward, due to the extension of the lesion within the medial extraconal orbit site as shown from preoperative CT. The imaging also shows the integrity of the frontonasal bone, and allows the surgeon to exclude intracranial communications in the midline. Figures show normal reconstitution of the face volumes, symmetry of the eyebrows and minimum residual scar (Figure 3-8).

Case 3

D.S.M.: The haemangioma fills the entire nasal region, glabella and frontonasal area on the midline. Complete removal of the mass up to the periosteal plane. Reconstruction with advancement flaps on the median line, according to the aesthetic units. Two years follow-up shows a residual dystrophic scar on the back, to be reviewed in adolescence (Figure 9-16).

Case 4

C.E.: Giant IH of the nasal dorsum and glabellum. Complete

Figure 1 Mixed Glabellar Infantile Haemangioma. Asymmetrical distribution in relation to the midline.

Figure 2 Skin coverage obtained by advancement of free residual skin margins, after extensive dissection of subcutaneous tissue. Follow-up 7 days after surgery.

Figure 3 Frontonasal giant haemangioma with right eyeball dislocation toward the right side.

Figure 4 Volumetric enlargement of the lesion during crying (Spontaneous Valsalva maneuver).

Figure 5 CT scan shows normal frontal bone and no communications through the midline.

Figure 6 Follow-up two years after surgery. Frontal view.

Figure 7 Follow-up two years after surgery. Submental view.
Central

removal through a subperiosteal plane. Reconstruction using local skin flaps and Z-plasties (Figure 17-24).

Case 5

S.A.: Giant IH of the left frontal area, hairless eyebrow with serious deformity. Complete removal of the lesion lengthening the surgical incision toward glabellar region in order to get a
Central brow symmetry and distributing the redundant tissue (Figure 25-33).

DISCUSSION

Highly vascularized soft-tissue tumors can clinically mimic deep hemangiomas. Clinical clues against the diagnosis of IH include lesions that are firm, fully formed at birth, lack warmth despite a vascular appearance, or continue to progress beyond the expected timeframe for IH or despite treatment. In some cases, the vascular nature of the tumor also mimics IH on MRI [12,13]. In these cases, histologic confirmation of the diagnosis is warranted [14]. The diagnosis of IH is usually straightforward, but other vascular malformations and tumors, as well as soft tissue tumors must be considered as shown [15].

Congenital

- Encephalocele
- Sinus pericranii
- Nasal glioma

Figure 17 Preoperative frontal view.

Figure 18 Preoperative Submental view.

Figure 19 Preoperative drawings conducted to approximately 1 mm surrounding the red part of the skin, trying to save more skin as possible to cover.

Figure 20 Complete enucleation of vascular injury to the periosteum.

Figure 21 Surgical removal must be conducted following a natural cleavage between hemangioma and healthy tissue, thus minimizing intraoperative bleeding.

Figure 22 In this case flaps are not perfectly respectful of the aesthetic units of the nasal dorsum, but are sufficient to give a coverage to the loss of substance.

Figure 23 3 weeks postoperatively.

Figure 24 Follow-up 5 years after surgery frontal view.
Lipoma
Fibroma


From our point of view these giant vascular tumours must always be operated. Supporting the surgical approach, there are studies suggesting that centrally located facial infantile hemangiomas may present lower regression rates [16]. Considering that central lesions are associated with increased psychosocial prob-lems, the predominance of surgical treatment in the eyelids, lips, and nose is justified. Surgical excision should not cause complications that in themselves would be as or more amblyogenic than the hemangioma itself; namely, ocular movement anomalies, ocular deviation in primary position, ptosis, or visual loss (secondary to retrolbulbar hemorrhage).
Seven days after surgery check, still showing substantial periocular haemangiomas. The ophthalmologist, dermatologist/CONCLUSIONS

[17,18] There is a long-standing misconception that haemangioma excision carries a higher risk of blood loss compared with other lesions, adding to the reluctance of surgical treatment [19]. Unfortunately giant vascular tumours can bleed a lot during the surgery if surgery is not performed correctly. So it is advisable to carry out the removal to anatomical planes free from injury, coagulating or binding each time the feeding vessels. It should always apply firm traction of the mass during the disconnection from other tissues, thus minimizing blood loss [11,20].

Children not operated may develop amblyopia in three ways: refractive-error differences, visual deprivation (e.g., cataracts or severe ptosis) and strabismus. In children with a periocular haemangioma, all three amblyopiogenic causes could potentially lead to amblyopic vision. The most common cause of amblyopia associated with periocular haemangioma is astigmatism induced by the presence of the haemangioma distorting the cornea. Robb noted that in nearly all of his reported cases, the haemangioma seemed to exert pressure on the eye in a direction perpendicular to the axis of astigmatism. In addition, haemangiomas can affect the growth of the eye, potentially inducing myopia or hyperopia. As a result, anisometropia (a marked difference in refractive error between the two eyes) can also be an amblyopiogenic factor [21]. It has been shown that a difference of 2 diopters or more (hyperopia or myopia) is significant enough to produce amblyopia [22]. As the amblyopia is often multifactorial in children with haemangiomas, prompt multimodality treatment is often necessary to reverse the condition. Early surgical debulking to quickly alter lesions posing significant risk to the visual axis is an important component of this treatment [23-27].

CONCLUSIONS

Multidisciplinary care is central to effective management of periocular haemangiomas. The ophthalmologist, dermatologist/ pediatrician, and plastic surgeon all have central roles to play. Management is driven by functional issues that are detected and monitored by regular ophthalmologic assessment. Surgery in these huge lesions is mandatory for the risk of frequent trauma and unmanageable bleeding, in addition to problems linked to exclusion from social life as part of the primary school. Surgery is indicated for sight-threatening haemangiomas unresponsive to other forms of management, or for small discrete lesions that are amenable to total excision with minimal risk of morbidity. Cosmetic indications for school-aged children with disfiguring lesions must be taken into account. Extensive haemangiomas may be excised partially to protect sight with the aim of minimizing the morbidity of surgery and achieving the best final cosmetic result. Moreover early treatment will have appropriately averted any psychological distress that could potentially occur in school or amongst peers. In conclusion, treatment of giant haemangiomas to reduce their size is effective at reducing the amount of anisometropic astigmatism and mitigates both the duration and the incidence of deprivational amblyopia, ultimately maximizing good visual outcomes. Treatment of capillary haemangiomas should be initiated promptly in the presence of amblyogenic factors and treated quickly and definitively in the presence of occlusion.

CONSENT

All patients shown have given consent to the publication of the photos posted inside of this paper.

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