Giant Hemangioma Presenting as a Scalp Mass Leading to a Craniofacial Deformity

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Abstract Hemangioma is the most common vascular tumor during childhood. However, cases of giant hemangiomas determining deformities in the craniofacial region are not reported frequently. Craniofacial giant hemangiomas are challenging for surgeons since they cause aesthetic and/or functional problems and may be associated with important complications such as ulcerations, infections and bleedings. The present report is aimed at describing a rare case of craniofacial asymmetry in a 30-year-old patient,

secondary to an extensive mass whose growth deformed the scalp in the frontal-parietal-temporal region. The lesion was completely surgically removed with good aesthetic results, and the anatomopathological examination revealed that it was a cavernous hemangioma.

Keywords Cavernous hemangiomas · Craniofacial abnormality · Hemangiomas · Surgery

Introduction

Hemangiomas are the most common benign tumors in childhood [1, 2]. Generally, they are not present at birth [1, 2], but they may develop at any time during the first year of life [1]. These tumors can occur in any tissue [1], mainly in areas of the head and neck (60% of cases) [2].

Most lesions tend to regress by 10 years of age [1, 2] and, in some cases, tumors can achieve large dimensions [3–11]. There is no clear definition about what size a lesion should achieve to be regarded as a giant hemangioma [6, 7]. Colebatch et al. [11] described the giant hemangioma as a tumor with a diameter of at least 5–6 cm.

Giant hemangiomas determining deforming masses in the scalp have been rarely reported in the literature [4–8, 10, 11]. In the craniofacial region, hemangiomas that reach large dimensions are challenging for the surgeon [3], because in addition to causing aesthetic changes [1–3] and psychosocial impact [1, 2], they can cause major complications such as heart failure [6], ulcerations and bleedings [4].

The present report is aimed at describing a rare case of craniofacial deformity in a 30-year-old patient, with a slow and insidious progression since childhood, due to a giant cavernous hemangioma of the frontal-parietal-temporal region that was successfully surgically removed.

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Case Report

A 30-year-old female, with fair skin (Caucasian) sought medical care with a history of a slowly growing painless mass, in the right frontal-parietal-temporal region, since her first year of life.

Family members reported that, initially, the mass was noticed only through palpation of the scalp in the right temporal region. Over the years, the mass showed a progressive growth, leading to a huge asymmetry with aesthetic damage in the craniofacial region, starting to interfere significantly on the daily activities of the patient, who has become isolated from social life. The patient had no other related symptoms, but complained of lack of hair in the area of lesion. The neuro-psycho-motor development during childhood was reported as normal. She denied a history of head trauma and reported no other family members with similar history.

The patient reported that 10 years ago, in another hospital, had undergone two embolizations of the vessels that irrigated the tumor. She indicated that the tumor exhibited growth stability for a few months after embolization, and then increased again. She denied having undergone other therapeutic approaches.

On admission, physical examination and neurological assessment of the patient were normal, except for the presence of a large bulge in the right frontal-parietal-temporal region, approximately 12 cm in diameter, which produced a significant change in the position of the right ear (Fig. 1a, b). On palpation, it was a firm painless mass, which had a slight pulsation. The evaluation of skin showed alopecia of the scalp in the affected area. The regional lymph nodes were not enlarged.

The cranial magnetic resonance imaging showed a semi-ovoid isodense lesion in the right frontal-parietal-temporal area, with approximately $12 \times 8 \times 6$ cm in dimension. The mass was located beneath the aponeurotica galea, but it was not seen adjacent bone destruction or invasion (Fig. 2a). No intracranial abnormalities were detected. Angioresonance of the head showed a vascularized lesion, with irrigation from the external carotid artery branches (Fig. 2b). No diagnostic preoperative biopsy was carried out due to the bleeding risk.

The option was the surgical resection of the mass and embolizations of vessels that irrigated the tumor before surgery in order to reduce bleeding during the surgical procedure. To further reduce the bleeding, before the skin incision, an adrenaline solution diluted at 1:100.000 of

Fig. 1 **a** Craniofacial deformity secondary to mass that bulges the scalp, **b** associated with change in the position of the right ear and alopecia

Fig. 2 **a** Magnetic resonance imaging of the brain showing extensive semi-ovoid isodense mass located beneath the aponeurotica galea in the right parietal region, compressing the temporalis muscle without infiltrating it and without bone involvement, **b** Head angioresonance showing tumor richly vascularized in the right parietal region

saline solution was infiltrated around the lesion. After the skin incision, a reddish circumscribed mass was observed; it was fibroelastic on palpation, with approximately 12×8 cm in length, and it was located below the temporalis muscle (Fig. 3). The mass was completely surgically removed through a pericranial cleavage plane (outside the lesion), with ligatures and electro-coagulation of the blood vessels that irrigated the lesion. The tumor was covered by a fibrous capsule, continuous with the pericranium. There was no evidence of involvement of the outer table of skull or regional musculature. The primary closure of the surgical wound was carried out. There were no complications in the postoperative period.

On gross pathologic examination, the surgical piece was coated by a thin bright capsule consisted of soft reddish tissue and displays the cutting surface whitish-brown, shiny, with scattered hemorrhagic spots (Fig. 4a, b). Histological examination revealed large thin-walled vessels filled with erythrocytes, consistent with the cavernous

Fig. 3 Intra-operative aspect. Relatively circumscribed mass with a thin bright reddish capsule under the temporalis muscle. (Color figure online)

Fig. 4 Photograph of surgical specimen. **a** Lesion partially surrounded by fibrous tissue layer of indistinct boundaries. **b** Surface of section showing whitish-brown color, shiny, with scattered hemorrhagic spots. (Color figure online)

Fig. 5 Photomicrograph of surgical specimen showing vessel proliferation caliber/size varied, coated internally by endothelium with no signs of malignancy consistent with cavernous hemangioma (hematoxylin-eosin, original magnification $80\times$)

hemangioma, with no evidence of malignancy (Fig. 5). Surgical margins were tumor-free.

The patient was discharged 2 days after surgery, in good clinical conditions, without evident neurological lesions. Follow-up was carried out for 2 years after surgery, with no sign of local recurrence and with good aesthetic results (Fig. 6a, b).

Discussion

Historically, the development of the classification of vascular lesions of soft tissues was so varied and confused [1, 8]. The classification system of Mulliken and Glowacki organized these lesions according to their clinical and histological features [1, 8]. Hemangiomas affect mainly patients in the pediatric population [1, 2] with the incidence reaching 12.6% by the end of the first year of life [1]. The

Fig. 6 Patient showing good aesthetic results 2 years after surgery. **a** Anterior view. **b** Lateral view

main location of these vascular tumors is the head and neck (60% of cases), followed by the trunk (25%) and limbs (15%) [2]. Clinically hemangiomas may be superficial, deep (subcutaneous) or mixed [2] and approximately 80% of them appear as single and sporadic lesions during the postnatal period [1, 2]. In most cases they are characterized by a strawberry mark or port-wine stain [7]. Only a small percentage of tumors are installed in the subcutaneous tissue [7], and it may or may not be a slight bluish tone on the skin [2].

When originated in the calvaria, most hemangiomas are asymptomatic and some patients may complain of headaches, pain in the scalp and/or existence of a palpable mass [12]. In cases involving the scalp, the skin may be normal and there may be areas of ulceration, recurrent infections, painful thrombosis and/or bleedings [3, 4]. There may also be functional and/or aesthetic disability [3, 4], as large areas of alopecia, distortion of the hairline, or deformation of the ear [3]. In some cases of lesions in the temporal area, there may be neurological deficits, such as facial nerve paralysis and hearing loss [12].

Hemangiomas have a characteristic biphasic growth [4]. During the first years of life, lesions grow in size rapidly (proliferative phase) [1, 4]; proangiogenic factors (proliferating cell nuclear antigen, vascular endothelial growth factor, and type IV collagenase) are involved with this proliferating [1]. In about 70% of cases, after this stage, there is a growth arrest followed by a regression phase [2, 4] that can last up to 10–12 years of age [1, 4]; antiangiogenic factors (tissue inhibitor of metalloproteinase, mast cells, and scavenger macrophage) are involved with this regression [1]. In some cases, these lesions do not regress and they continue to grow and reach large dimensions [3–11]. Cases of giant hemangiomas forming masses that deform the scalp as the case reported here have rarely been reported in the literature [4–11], especially in adults [4].

When giant, hemangiomas may cause several clinical problems [5, 7], such as reduction of cerebral blood flow due to the tumoral “kidnapping”, which can cause mental retardation [7]. Other problems are congestive heart failure due to high cardiac output and thrombocytopenia known as the Kasabach–Merritt syndrome [5, 7]. They can also result in compression of vital structures, as the trachea and esophagus, and necrosis and desquamation of the skin overlying the lesion [6].

In histology, hemangiomas can be classified as cavernous or capillary [9, 12, 13]. The first type has no capsule and it is formed by large vascular cavernous spaces filled with blood and separated by connective tissue [9, 12, 13]. In contrast, capillary hemangiomas are composed of smaller vessels without fibrous septa [10, 13].

The etiology of hemangiomas is still unknown [12]. Current theory suggests hemangiomas arise through hormonally driven vessel growth, but their origin as tumors of neogenesis or revival of dormant embryonic angioblasts remains controversial [1]. The cases characterized by a strawberry mark or port-wine stain are easily diagnosed [7] through previous history and physical examination of the lesion. In cases where there is little or no discoloration of the overlying skin, as in the case presented here, the distinction from other tumoriform lesions becomes difficult [7]. Differential diagnoses include epidermoid cysts, eosinophilic granulomas and sinus pericranii [7].

In cases of pericranial hemangiomas, most of the times, the diagnoses are obtained only after the surgery [13]. In the case described here, it was decided not carry out aspiration biopsy by fine needle or incisional diagnostic biopsy during the preoperative period due to the high risk of bleeding. The computerized tomography scan is the diagnostic choice since it confirms the plain skull findings showing detailed intracranial extension [12] and reveals a homogeneous mass with uniform enhancement after the application of intravenous contrast [1]. Magnetic resonance imaging findings for hemangiomas include a moderate degree of signal enhancement in T1-weighted images and a strong signal increase in T2-weighted images [14].

The tumor presented here probably has a bone origin, since the majority of hemangiomas arising from the calvaria develop from the frontal and parietal bones; they are usually cavernous [8, 9, 12] and supplied by branches of the external carotid arteries [9, 12]. However, differing from the case presented here, these vascular tumors originated from the calvaria generally determine erosion of the outer table and, in general, save the inner table [8, 9, 12].

This tumor could also have a muscular origin (temporalis muscle) [14]. However, during surgery, it was observed that the tumor compressed only the temporal muscle and that there was a clear boundary between the tumor capsule and the muscle. In addition, there were no

smooth muscle fibers in the pathological examination [14], which makes less likely the hypothesis of muscular origin.

The tumor capsule was closely in contact with the calvaria, which leads us to believe that the pericranium is the origin of the tumor reported here, similar to that presented earlier by Tokuda et al. [7]. There are situations where the exact determination of the hemangioma origin may not be possible [8].

The main targets for the treatment of hemangiomas include minimizing physical complications that can lead to morbidity and mortality, mitigate possible psychosocial conflicts, and avoid overly aggressive procedures, with toxic side effects or unnecessary scars in hemangiomas with a high probability of good prognosis without therapy [1]. Therapeutic options include intra-lesion or systemic steroids, embolization, interferon alfa-2a, cryosurgery, vincristine administration and surgical resection [2]. The choice of a conservative or aggressive therapeutic approach should be evaluated individually [2]. Factors such as location and depth of the lesion, presence or risk of complications are parameters that can serve as the basis for this decision [1].

Most cases of hemangiomas are treated conservatively in the expectation of spontaneous resolution [1–3, 7], since the majority of these tumors tend to regress [1–4, 7]. Surgical treatment is indicated for large hemangiomas that do not regress or that are functionally and/or aesthetically incapacitating, recurrent or hemorrhagic [4, 8]. In cases of giant hemangiomas, surgical resection continues to be the treatment of choice [4, 9, 12, 13].

In the case described here, surgical resection was considered as necessary due to the aesthetic and functional involvement, the size of the lesion, and because it was needed to differentiate the tumor from other lesions, particularly malignant ones. For cases of giant hemangiomas, the main goal of the surgical resection is the bleeding control [7, 8], since these masses pose risk of extensive bleeding in the intra-operative period [4, 7, 9, 12]. The preoperative embolization to reduce tumoral irrigation and bleeding during surgery facilitates the surgical procedure [8, 12].

Recently, Spector et al. [3] presented their experience in treating six cases of large scalp hemangiomas, and reported that the sequence for the treatment of lesions was important to reduce the bleeding risk during surgeries. They reported [3] that all excisions were carried out only after infiltration of the lesions and surrounding tissues, with a “tumescant” adrenaline solution, and they described hemostatic polypropylene sutures around hemangiomas to limit the blood inflow to the lesion; they used an avascular dissection plane in the galeal layer, outside the lesion, also aimed to minimize bleeding. Resection of hemangiomas by the endoscopic approach has some advantages over the conventional method [15, 16], as more precise evaluation of tumor extension, greater preservation of the local

anatomy and smaller scars [16]. Moreover, tumor size and a higher risk of bleeding may contraindicate endoscopic surgery in many cases [16].

Other treatment options are curettage [9, 13] and radiotherapy [6, 9, 13]. Curettage can determine an excessive blood loss with consequent disturbance of homeostasis, and higher risk of recurrence compared with surgical excision [9, 13]. Irradiation is dangerous due to damage to the underlying bone growth centers, tissue atrophy, scarring and risk of developing malignant lesions [6, 13].

Conclusion

Giant hemangioma should be considered in the differential diagnosis of tumoriform craniofacial lesions. Surgical resection is the main treatment option and it is usually effective. Special attention should be given to the risk of bleeding from trauma and during surgical resection.

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Conflict of Interest None

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