Treatment of high-flow arteriovenous malformations (AVMs) of the head and neck with embolization and surgical resection

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AIM: *Arteriovenous malformations (AVMs) with a high flow of the head and neck are quite rare compared to their low-flow counterparts, but when they do occur they are often accompanied by massive bleeding or present with significant esthetic defects. The treatment of these high-flow vascular anomalies is often highly risky. The multidisciplinary approach required for the assessment and treatment of these lesions should include angioradiology with preoperative superselective embolization, followed by surgical resection of the lesion within 24 hours and esthetic reconstruction.*
MATERIALS OF STUDY: *We studied a series of 55 patients with AVMs of the head and neck that were treated surgically between 1999 and 2009.*
RESULTS: *Of the 55 patients with AVMs, 7 had hemangiomas and 48 had vascular malformations, of which 28 were low-flow lesions and 20 were high-flow lesions (AVMs). The high-flow lesions were most commonly located on the lip. All 48 AVMs underwent surgical resection for concerning symptoms, diagnostic purposes, or esthetic problems. Of the 20 high-flow AVMs, 20% were classified as Schöbinger stage I, 55% as stage II, and 25% as stage III. A combined treatment of embolization and resection was used to resolve 13 of the high-flow AVMs (stages II and III), of which 4 required a flap reconstruction.*
CONCLUSION: *Recent advances in microsurgery and interventional angioradiology have greatly improved the prognosis for patients with these malformations. Combined embolization-resection is the treatment of choice for high-flow AVMs, and esthetic reconstruction with flaps can prevent their recurrence.*

KEY WORDS: Arteriovenous malformation, Transcatheteric embolization.

Introduction

A common classification of vascular malformation pathologies is required, because in the literature there are currently more than 50 different types. The International Society of the Study of Vascular Anomalies (ISSVA) classification (ISSVA – 11th Workshop, Rome, 1996) is easy to use, particularly for vascular abnormalities of the head and neck. It is based on clinical criteria, evolution, histopathologic and hemodynamic properties, and comprises two large groups: hemangiomas and vascular malformations [1-2].
Vascular abnormalities of the head and neck can thus be classified as hemangiomas or vascular malformations.

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Hemangiomas are common in childhood; a conservative "wait and see" approach is recommended for this type because most of these lesions regress spontaneously [3]. Histologically, hemangiomas display endothelial hyperplasia and an increased number of mast cells in the proliferative phase [4], and phase involution is characterized by the fibrotic replacement type, reduced cellularity, and a normal mast cell count. Conversely, vascular malformations are labeled "mature" because although they sometimes occur at birth their incidence increases with the development and growth of the body, often manifesting in the adult; they do not resolve spontaneously [5]. Vascular malformations are not tumors, but rather an accumulation of abnormal vessels with endothelial proliferation and a normal mast cell count, representing morphogenetic errors with structural abnormalities but without endothelial cell proliferation [6].

Vascular malformations can be further classified according to the type of vessel involved, as capillary, venous, lymphatic, and arteriovenous. They are then further divided into low-flow lesions (capillary, venous, lymphatic, or combinations thereof) and high-flow lesions (blood component, usually arteriovenous) [7]. High-flow arteriovenous malformations (AVMs) of the head and neck are particularly problematic with regard to treatment because of their propensity for rapid growth. While surgical therapy alone often results in a significant reduction in most of the clinical features, it is not sufficient to resolve the problem; a multidisciplinary approach is considered the treatment of choice [8].

Transcatheteric embolization is particularly useful by itself in children with vascular malformations of the maxilla or jaw. However, in growing children, surgical removal of large segments of bone may lead to impaired facial growth, resulting in an abnormal appearance and dental occlusion, which itself requires orthodontic-prosthetic rehabilitation [9]. Surgery in this age group is confined to "debulking" of the soft tissues of the tumor that is infiltrated by the malformation.

Materials and methods

In the 10 years between 1999 and 2009, we treated 55 patients surgically to repair vascular malformations of the head and neck. Data were collected regarding the patients' age at the time of surgery, sex, presenting signs and symptoms, anatomical location of the anomaly, size, mode of diagnosis, indications for surgery, surgery performed and other treatment modalities, histopathological data, and outcome after treatment. The vascular abnormalities were classified based on their histopathology as hemangiomas and AVMs.

TABLE I - *Clinical details of 20 patients with oral and maxillofacial high flow malformation.*

Case	Age	Sex	Location	Symptomatology	Istology/ Schobinger	Rx	Embolization	Type of surgery	Complications	Post surgery embolization	Follow up/m	Results
1	21	M	L	E	T II	–	EMB	Excision			60	VG
2	18	M	L	E	A III	–	EMB	Excision			60	G
3	6	M	F	S	Cg I	MR		Excision	REC	EMB Excision Tissue expansion scalp	56	S
4	8	F	C	S	Cg III	MR	EMB	Excision			48	VG
5	35	M	M	P	A II	CT/MR	EMB	Excision			45	VG
6	42	M	N	P	A II	CT/MR	EMB	Excision			43	G
7	29	F	L	HT	A I	–		Excision			38	G
8	40	M	L	S	A II	–		Excision			38	G
9	15	M	C	S	Cg II	MR	EMB	Excision			36	VG
10	23	M	C	P	A II	MR	EMB	Excision			36	VG
11	18	F	L	S	Pub I	CT/MR		Excision			29	G
12	18	M	Na	Pa	Cg II	CT		Excision			28	G
13	21	F	F	S	Pub II	CT/MR	EMB	Excision			28	VG
14	36	M	C	E	A I	CT/MR		Excision	REC	EMB Excision Free flap reconstruction	24	G
15	10	F	F	V	Cg II	MR	EMB	Excision			24	VG
16	36	M	L	S	A II	CT/MR	EMB	Excision			24	G
17	28	M	L	P	A I	–		Excision			19	S
18	14	F	F	S	Pub III	CT/MR		Excision			15	G
19	9	M	C	S	Cg II	MR		Excision			12	G
20	41	F	M	S	A III	CT/MR	EMB	Excision			6	G

L: lip; F: front; C: cheek; M: mandible; N: neck; Na: nasal dorsum; H: hemorrhage; HT: elevated local temperature; S: swelling; P: pulsatility; Pa: pain; V: sign of Valsalva; T: traumatic; A: adulthood; Pub: puberty; Cg: congenital; EX: surgical excision; S: sufficient; G: good; VG: very good.

Results

All 55 patients underwent surgical resection of vascular anomalies of the face or neck with or without the use of other modes of treatment. The vascular malformations of 7 the 55 treated patients were defined as hemangiomas, while those of the remaining 48 were AVMs. The series of hemangiomas comprised two males and five females (male:female ratio, 1:2.5) aged 5-42 years (mean, 22.4 years). The series of AVMs comprised 28 low-flow lesions and 20 high-flow lesions.

Of those with high-flow AVMs, 13 were men and 7 were women (Table I) with an age range of 6-42 years (mean, 29.2 years). The area most commonly involved was the lip (*n*=7), followed by cheek (*n*=5), forehead (*n*=4), mandible (*n*=2), neck (*n*=1), and nose (*n*=1). The lesion size ranged from 5x5x40 mm to 5x5x60 mm. Six of these patients submitted to surgery after recurrence or continuation of a previous treatment for a high-flow AVM. The 20 patients in this group presented with a bulge/swelling associated with a progressive enlargement thereof (*n*=10), along with an increase in the pulsatility index (*n*=4), bleeding (*n*=3), pain, and discomfort (*n*=1), a positive Valsalva sign (*n*=1), and a high local temperature (*n*=1). Six patients had a history of congenital disease, while the AVM was observed for the first time during puberty in three patients, immediately after a trauma in one patient, and in adulthood in the remaining ten patients.

Diagnostic magnetic resonance imaging (MRI) and/or computed tomography were performed in 15 cases, while superselective angiography with embolization was conducted in 11 cases. Surgical excision alone was used in 9 patients, while 11 cases (8 Schöbinger stage II or stage III) were treated with superselective embolization 24 hours after en bloc resection. Two of the patients required a free flap to be taken from the forearm for the reconstruction of a radial lip, with debulking in a second phase.

The follow-up period ranged from 6 to 60 months (mean 26 months). At follow-up, two of the patients who were initially treated by surgical resection only subsequently underwent preoperative embolization followed by excision of lesions that required reconstruction of the soft tissue using of scalp tissue (after its expansion). All of the other lesions were excised with direct closure. There were no recurrences in any of the other cases. At the last follow-up, the results were very good in 7 cases (35%), good in 11 (55%), and insufficient in 2 cases (10%).

Discussion

The correct management of vascular anomalies depends upon an accurate diagnosis. It is important to distinguish between hemangiomas and AVMs, and further, between high- and low-flow AVMs. The term "high flow" describes an arteriovenous anomaly comprising multiple, low-resistance vascular shunts [4]. These do not tend to exhibit expanded cell hyperplasia, but due to hemodynamic mechanisms leading to the formation of a collateral circulation they deflect the regional blood flow from the periphery. Of crucial importance is that primary pathological lesions often occur at the arteriole–capillary arteriovenous shunt, large arteries, with dilated draining veins due to recurrences.

Long-term success can only by guaranteed by complete resection of AVMs [3,4,10]. Proximal surgical ligation of arteries without resection is doomed to failure, and must be cajoled into what determines the formation of collateral circulation to the closure you can not use secondary therapeutic embolization. However, esthetic and functional considerations require minimization of tissue removal.

The classification of peripheral AVMs proposed by Allison and Kennedy [11] is based on both the clinical and angiographic characteristics of the flow. It comprises three groups with the following predominant characteristics:

Group 1: arterial and arteriovenous lesions.
Group 2: pathology involving small blood vessels.
Group 3: venous lesions.

More recently, the 11th Workshop of ISSVA presented a classification that is easy to use and provides for an initial distinction between hemangiomas and vascular malformations, and a second distinction between AVMs into low-flow (capillaries, venous, lymphatic, or combinations thereof) and high-flow (arteriovenous) types.

Most of our cases were identified as malformations of the lip (35%), cheeks (25%), and face (20%); there were no ear malformations. In contrast, Allison and Kennedy [9] and Kohout et al. [12,13] found that the most common locations of 81 AVMs were the cheek (31%) and the ear (16%).

AVMs may progress due to acute stimuli such as trauma, pregnancy, puberty, infection, or iatrogenic injury (biopsy, proximal ligation, or subtotal excision) [5,7,12]. Clinically, the overlying skin may appear normal, as a pulsatile mass, with redness and increased warmth; other clinical features may include pain and excessive growth, until bleeding heart failure occurs.

Using the classification of Schöbinger, 25% of patients were at stage I (skin redness or heat), 55% were at stage II (visible pulsation or expansion of the lesions), 20% were at stage III (pain, ulceration, hemorrhage, or infection), and none were at stage IV (heart failure). On MRI, the AVMs were characterized by enlarged blood vessels with vascular dilation, collateral arteries, and vascular lesions with considerable drainage (confirming their high-flow nature). MRI is an essential tool for the early diagnosis of AVMs that appear as irregular or globular masses and which may be located in the brain cortex, in the subcortical gray and white matters. They may

be observed as small, round, low-intensity signal spots in or around the mass on T1- or T2-weighted sequences. Low-signal-intensity hemosiderin may also be seen extracellularly or around the AVMs, indicating symptomatic or asymptomatic hemorrhage, respectively. Magnetic resonance angiography with contrast medium can identify masses of 1 cm in diameter, but it is not appropriate for studying the morphology of arteries and veins.

Angiography, which was performed in 94.5% (52/55) of our cases, was necessary to evaluate the anomaly in more detail before excision surgery or embolization [14]. This revealed a large number of shunts with a high blood flow. One case involved the jaw in addition to the main focus of the hypothalamus and optic nerve, enabling a diagnosis of Wyburn-Mason syndrome [15]. Angiography also revealed involvement of the facial artery and facial vein in all cases (Table II).

The treatment of arteriovenous anomalies is difficult and the results are sometimes disappointing. In 1815, Bell [16] admitted that an AVM (aneurysm by anastomosis) is clearly different from an acquired fistula, and found that long-term success is only possible with total resection of the involved tissues. The treatment of choice is the selective embolization of vessels in combination with surgical resection and reconstruction of the soft tissues. The aim of preoperative embolization is mainly to reduce blood loss and facilitate the surgery, and must not be considered a method for reducing the extent of resection. Surgical resection should not be delayed beyond 48 hours after embolization because the resulting inflammation renders the aforementioned hemodynamic benefits useless and the surgery more difficult.

An AVM is often not diagnosed until a dramatic bleeding event, which may occur following dental treatment, for example [9]. A variety of maneuvers can be used to minimize intraoperative hemorrhage: hypotensive anesthesia [17], a temporary suture around the periphery of the excision margin, and cardiopulmonary bypass with hypodermic circulatory arrest [18].

Arterial embolization techniques have improved considerably since 1904 when Dawbarn [19] first injected hot liquid wax into the external carotid artery of a patient with a craniofacial tumor. Considerable progress has

TABLE II - *Detail of embolized oromaxillofacial high flow AVMs.*

Case	Location	Vessels	Embolization technique	Embolization results
1	L	Facial artery Venous drainage: facial vein	PVA	90% reduction of flow
2	L	Facial artery Venous drainage: facial vein	PVA and Gelfoam	70% reduction of flow
3	F	External carotid artery (internal maxillary and facial arteries) Venous drainage:internal maxillary vein, facial vein	Coils	40% reduction flow Residual AVM from facial artery
4	C	Internal maxillary artery, facial artery Venous drainage: superficial and deep facial veins	PVA	90% reduction of flow
5	M	External carotid artery (facial branches and internal maxillary) Venous drainage:internal maxillary vein, facial vein into jugular veins	PVA	90% reduction of flow
6	N	External carotid artery		70% reduction of flow
9	C	Facial artery Venous drainage: superficial and deep facial veins	Coils	90% reduction of flow
10	C	Facial artery Venous drainage: facial vein	Coils	90% reduction of flow
13	F	External carotid artery (internal maxillary and facial arteries) Venous drainage:internal maxillary vein,facial vein	PVA	90% reduction of flow
14	C	Facial artery Venous drainage: facial vein	PVA and Gelfoam	80% reduction of flow
15	F	External carotid artery(internal maxillary) Venous drainage:internal maxillary vein		90% reduction of flow
16	L	Facial artery Venous drainage: facial vein	Coils	80% reduction of flow
20	M	External carotid artery (facial branches and internal maxillary) Venous drainage:internal maxillary vein, facial vein into jugular veins	PVA	70% reduction of flow

been made in the last 10 years, both with respect to the improvement in embolic materials (quick-acting N-butyl cyanoacrylate glue, polyvinyl alcohol, thrombus-inducing coils, and Onyx liquid embolic fluid, or small balloons) and in knowledge of the anatomy of the microcirculation. Djindjian and Merland [14] were the first to use superselective angiography and described in detail the possible complications of this technique. The aim of embolization is to block the high-flow shunt between the arterial and venous systems. The reduction or closing of AVMs renders resection and radiosurgery more accurate, safer, and easier. The potential risks associated with extra-axial embolization include the neurological defects caused by the reflux of the material occluding the intra-axial vessels that supply blood to the brain and the possible paralysis of cranial nerves caused by the closure of the small vessels of the external carotid artery supplying the cranial peripheral nerves. However, these complications are rare and are minimized by the experience of the operator [20]. It is essential that these procedures are performed in specialized centers, where there are clinicians with specific appropriate skills. The introduction of the direct-flow embolization technique, and a better understanding of the principles of flow and occlusion have significantly lowered complication rates.

Microshunts may appear after ligation of the main power supply of the vessels, or due to incomplete embolization, leading to a subtotal resection, after which the possibility of relapse is often greater than it was for the primary lesion. The recurrences observed in two of the present cases were due to incomplete and inadequate initial surgical resections and subsequent use of nonoptimal embolization. Appropriate patient selection and use of the right techniques are the main factors that contribute to successful embolization in the overall management of extra-axial lesions of the head and neck.

The embolic material used is commonly made of polyvinyl alcohol, which is not absorbed and is available in a series of particle sizes ranging from 0.15 to 1.0 mm in diameter. These particles expand by a factor of 10-15 a few seconds after contact with the blood.

The angiographic protocol that we use is as follows. First, diagnostic angiography is performed, followed by selective embolization. All superselective embolization procedures are performed within 24 hours before surgery. Access is via the femoral artery up to the carotid and vertebral vessels; the vessel nearest to the AVM is selected. Pressure measurements are then made with a coaxial catheter in order to identify the possible risks of subsequent bleeding. If intracranial involvement is expected, temporary anesthesia with sodium amytal can be applied to the vessels affected by the AVM; this is known as "superselective Wada testing." This test provides for the evaluation of particular parameters (language, memory, spatial vision, sensitivity and motor functions) within 5 minutes of general anesthesia to determine the possible involvement of the intracranial component and functions that arise from these locations that may in future lead to neurological disorders during embolization or resection.

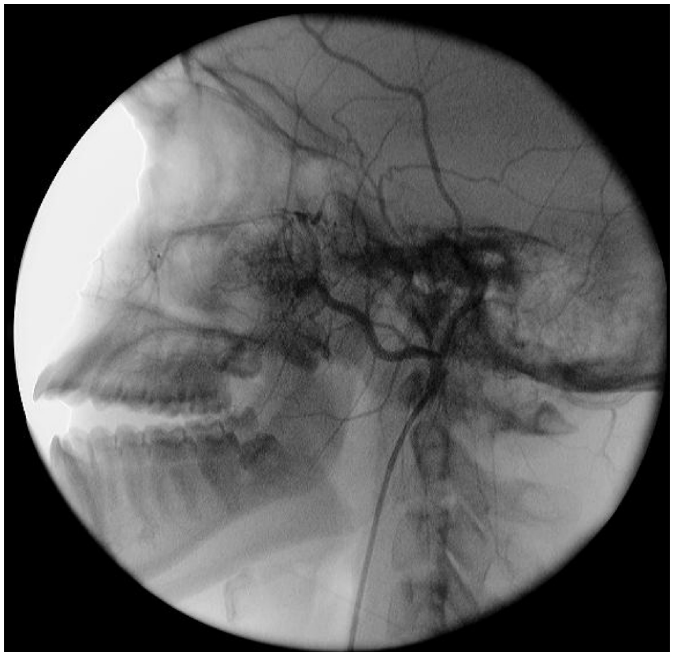

Fig. 1: External Carotid artery and superselective arteriography of internal maxillary artery demonstrating high flow AVM interesting maxillary and facial arteries.

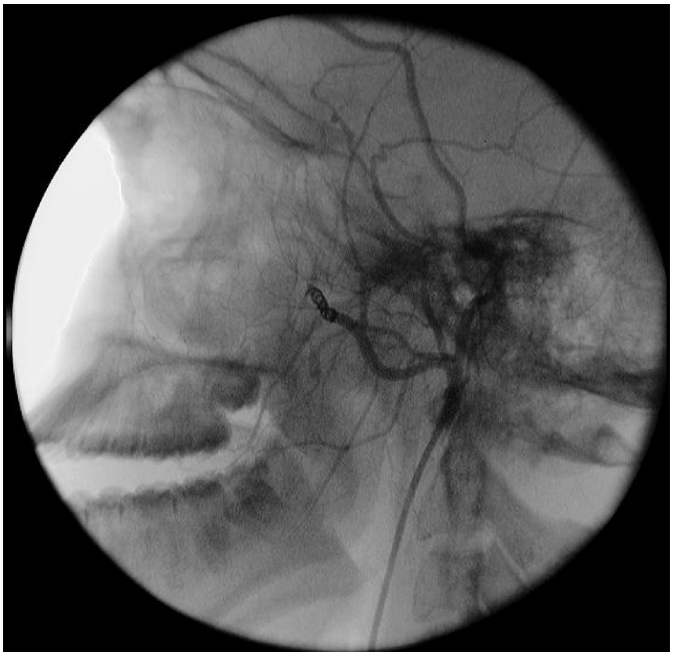

Fig. 2: Superselective embolization of maxillary artery with coils.

Conclusions

A multidisciplinary approach to the treatment of AVMs including interventional radiologists, surgeons, plastic surgeons, anesthetists, and the blood bank has been recommended by many authors (e.g., 8,21,22). The inevitable development of neovascularization, often as a result of intracranial surgery, may lead to further difficulties. Adequate amounts of blood also need to be transfused.

The total resection of large AVMs may not be possible, and so it is necessary to inform patients regarding the possibility of a second operation. The consensus among surgeons is that superselective angiography alone is not satisfactory as a treatment, unless it is performed within 24-48 hours before surgical resection.

We recommend careful evaluation of AVMs and a combined treatment protocol consisting of selective embolization followed by total resection, ideally within 24 hours [7,8].

Riassunto

Obiettivo: le malformazioni arterovenose (MAV) ad alto flusso della testa e del collo sono piuttosto rare rispetto a quelle a basso flusso, ma quando si verificano sono spesso accompagnate da emorragia massiva o si presentano con significativi difetti estetici. Il trattamento di queste malformazioni è ad alto rischio ed è spesso necessario un approccio multidisciplinare per la valutazione e il loro trattamento. Tale trattamento dovrebbe includere l'angioradiologia con l'embolizzazione preoperatoria superselettiva, seguita dalla resezione chirurgica della lesione nelle seguenti 24 ore e la successiva ricostruzione estetica.

Materiali di studio: Abbiamo studiato una serie di 55 pazienti con MAV della testa e del collo che sono stati trattati chirurgicamente tra il 1999 e il 2009.

Risultati: Dei 55 pazienti con MAV, 7 avevano emangiomi e 48 avevano malformazioni vascolari, di cui 28 sono state le lesioni a basso flusso e 20 sono state le lesioni ad alto flusso (MAV). La sede più frequente delle lesioni ad alto flusso sono state le labbra. Tutte le 48 MAV sono state sottoposte a resezione chirurgica legata ai sintomi, a propositi diagnostici, o per problemi estetici. Delle 20 MAV ad alto flusso, il 20% sono state classificate come stadio I di Schobinger, 55% di stadio II, e il 25% come stadio III. È stato utilizzato un trattamento combinato di embolizzazione e resezione chirurgica per risolvere 13 del MAV ad alto flusso (fasi II e III), di cui 4 hanno richiesto una ricostruzione estetica con lembo chirurgico.

Conclusione: I recenti progressi nella microchirurgia e nell'angioradiologia interventistica hanno notevolmente migliorato la prognosi per i pazienti affetti da queste malformazioni. Il trattamento combinato embolizzazione-resezione è il trattamento di scelta per MAV ad alto flusso, e la ricostruzione estetica con lembi può prevenire la loro recidiva.

References

1) Enjolras O: *Classification and management of the various superficial vascular anomalies: Haemangiomas and vascular malformations.* J Dermatol, 1997; 24:701-10.

2) Chang MW: *Updated classification of haemangiomas and other vascular anomalies.* Lymphat Res Biol, 2003;1259-265.

3) McHeik JN, Renauld V, Duport G, Vergnes P, Levard G: *Surgical treatment of haemangioma in infants.* Br J Plast Surg, 2005; 58:1067-72.

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

5) Watzinger F, Gössweiner S, Wagner A, Richling B, Millesi-Schobel G, Hollmann K: E*xtensive facial vascular malformations and haemangiomas: A review of the literature and case reports.* J Craniomaxillofac Surg, 1997; 25:335-43.

6) Leon-Villapalos J, Wolfe K, Kangesu L: *GLUT-1: an extra diagnostic tool to differentiate between haemangiomas and vascular malformations.* Br J Plast Surg, 2005; 58:348-52.

7) Jackson IT, Carreño R, Potparic Z, Hussain K: *Hemangiomas, vascular malformations, and lymphovenous malformations: Classification and methods of treatment.* Plast Reconstr Surg, 1993; 91:1216-230.

8) Erdmann MW, Jackson JE, Davies DM, Allison DJ: *Multidisciplinary approach to the management of head and neck arteriovenous malformations.* Ann R Coll Surg Engl, 1995; 77:53-59.

9) Rodesch G, Soupre V, Vazquez MP, Alvarez H, Lasjaunias P: *Arteriovenous malformations of the dental arcades. The place of endovascular therapy: Results in 12 cases are presented.* J Craniomaxillofac Surg, 1998; 26:306-13.

10) Coleman CC: *Diagnosis and treatment of congenital arteriovenous fistulas of the head and neck.* Am J Surg, 1973; 126:557-65.

11) Allison DJ, Kennedy A: *ABC of vascular diseases. Peripheral arteriovenous malformations.* Br Med J, 1991; 303:1191-194.

12) Kohout MP, Hansen M, Pribaz JJ, Mulliken JB: *Arteriovenous malformations of the head and neck: Natural history and management.* Plast Reconstr Surg, 1998; 102:643-54.

13) Wu JK, Bisdorff A, Gelbert F, Enjolras O, Burrows PE, Mulliken JB: *Auricular arteriovenous malformation: Evaluation, management, and outcome.* Plast Reconstr Surg, 2005; 115:985-95.

14) Djindjian R, Merland JJ (eds): *Superselective Angiography of the External Carotid Artery.* Berlin: Springer-Verlag, 1978; 76-88.

15) Wynburn-Mason R: *Arteriovenous aneurysm of midbrain and retina, facial nevi and mental changes.* Brain, 1943; 66:163-203.

16) Bell J: *The Principles of Surgery.* London: Longman, Hurst, Rees, 1815: 456.

17) Munro IR, Martin RD: *The management of gigantic benign craniofacial tumours: The reverse facial osteotomy.* Plast Reconstr Surg 1980; 65:777-85.

18) Mulliken JB, Murray JE, Castaneda AR, Kuban LB: *Management of a vascular malformation of the face using total circulatory arrest.* J Gynecol Obstet, 1978; 146:168-72.

19) Dawbarn RHM: *The starvation operation for malignancy in the external carotid area.* JAMA, 1904; 17:792-95.

20) Riche MC, Merland JJ: *Embolization of vascular malformations.* In: Mulliken JB, Young AE (eds). *Vascular Birthmarks: Hemangiomas and Malformation*s. Philadelphia: WB Saunders, 1988; 436-53.

21) Apfelberg DB, Lane B, Marx MP: *Combined (team) approach to haemangioma management: Arteriography with superselective embolization plus YAG laser/sapphire tip resection.* Plast Reconstr Surg, 1991; 88:71-82.

22) Jackson IT, Jack CR, Aycock B, Dubin B, Irons GB: *The management of intraosseous arteriovenous malformations in the head and neck areas.* Plast Reconstr Surg, 1989; 84:47-54.