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Original Article

Our Experience in the Surgical Management of Arterio-Venous Malformations of the head and neck

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ABSTRACT

Introduction: Vascular anomalies comprise a diverse group of abnormalities in blood vessel morphogenesis that usually occur prenatally. Arterio-venous malformations (AVMs) are rare congenital vascular lesions accounting for 1.5% of all vascular anomalies, with 50% of them occur in the oral and maxillo-facial regions. Treatment of large, complex vascular lesions is a serious challenge for patients and surgeons because it can cause disfigurement and massive haemorrhage, which may be spontaneous or the result of surgical intervention. Our study aimed to demonstrate surgical management of massive AVMs of the head and neck.

Method: This retrospective study shows the treatment outcomes of 28 patients with massive maxillo-facial vascular malformations, who presented to our department for treatment from 1 January 2015 to 31 July 2022.

Results: Twenty-eight patients with a mean age of 17.32 ± 12.21 years (women: 15, men: 13) were enrolled in the study. Diagnosis included extra cranial AVMs of the head and neck region. Treatment modalities, in isolation or combination, included angioembolisation procedure, sclerotherapy, and surgery.

Conclusion: Management of AVMs is challenging owing to the replacement of normal tissue by the diseased ones and the high rate

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of recurrence. Hence, multi-modal approaches are needed for the effective restoration of tissues.

Introduction

Vascular anomalies constitute some of the most difficult diagnostic and therapeutic problems encountered in medicine. Their clinical presentation is diverse, ranging from asymptomatic birthmarks to life-threatening haemorrhages.[1] The international society for the study of vascular anomalies categorises vascular malformations into high-flow and low-flow lesions.[2] This classification is useful in the correct diagnosis of the lesion and definitive management of the patient. Approximately 60% of malformations in children occur in the maxillo-facial region.[3] In the head and neck region, these malformations pose a functional threat and cause significant cosmetic disfigurement as well.

Arterio-venous malformations (AVMs) represent 1.5% of all vascular malformations, among which 90% are intracranial AVMs.[4,5] The complexity of AVM management increases when they are present in the head and neck region. Multiple treatment modalities have been postulated. These range from embolisation and sclerotherapy to surgery. Several authors have suggested a combination of embolisation followed by surgery.[6]

Multiple or large AVMs of the head and neck are associated with serious complications during their evolution, from recurrent bleeding, ulceration and infection.[2] Multiple emergency admissions may be required, which poses a burden on the health care system and the patient. Early diagnosis and treatment will aid in curtailing these issues.

This study was conducted in our department, and outlines the surgical management and presents the outcomes of patients with large AVMs of the maxillo-facial region.

Patients and methods

This study was approved by the Ethics Committee (IRB#0182-22).

This retrospective study was conducted on 28 patients who presented to our department from 1 January 2015 to 31 July 2022. Data were obtained from the medical records and all patients who met the inclusion criteria were included in the study.

Inclusion criteria

All patients with extracranial high-flow vascular malformations of the head and neck region, involving one or more than one facial subunit and or more than one-third area of the scalp.

Exclusion criteria

Vascular malformations in other parts of the body
Low-flow malformations of the head and neck
Vascular tumours such as haemangioma

Patient's demographics, gender and age at presentation were recorded and data is shown in Table 1. Other recorded information included area involved by the malformation, status of embolisation, treatment given, mode of reconstruction, postoperative complications, last follow-up, recurrence and the need for surgical revisions.

Radiological evaluation was carried out using either contrast-enhanced MRI or CT angiography. These cases were discussed with the interventional radiology team for assessing the possibility of

Table 1
Patient demographics and treatment.

Patient	0-10 years	11-20 years	21-30 years	31 years and above
No of patients (n)	**10**	**7**	**8**	**3**
Previous embolisation (other institutions)	**2**	**-**	**1**	**-**
Embolisation	**-**	**3**	**2**	**1**
Complete excision	**6**	**5**	**8**	**3**
Partial excision	**4**	**2**	**-**	**-**
Reconstructive modality				
Local flap	**3**	**-**	**1**	**-**
Free flap	**7**	**7**	**7**	**3**
Mean Follow-up period (months)	12.2	18.8	33.4	7.8
Secondary procedures	**1**	**1**	**5**	**-**
Recurrence	**2**	**2**	**1**	**-**

carrying out angioembolisation, which is usually performed 24 h prior to surgical debulking. Three patients in this study group were previously embolised in different centres, prior to presenting to our department. No records of their previous treatments were available.

The protocol for debulking was to mark the feeders using a pencil Doppler before draping the patient. Local adrenaline solution at a concentration of 1:400,000 was infiltrated in the operative field. All patients underwent controlled hypotensive anaesthesia to decrease the burden of blood loss during the resection phase. Control of the major feeding vessels was attained prior to excision. Surgical excision was performed with the help of vessel sealing devices (LigaSure or cautery).

Results

A total of 28 patients were included in the study, and 13 (46.4%) were men and 15 (53.6%) were women. The mean age was 17.32 ± 12.21 years. The most frequently involved area was the cheek. Two of the patients had bone involvement as well (the maxilla and the mandible).

Cosmetic disfigurement was the primary concern of all patients. Along with disfigurement, some patients also experienced functional abnormalities such as visual obstruction, drooling of saliva, occasional bleeding from the lesion and difficulty in mastication, articulation and ulceration.

Radiological imaging was performed for all patients. In 4 (14.3%) patients CT angiography was the sole investigation. Most patients (85.7%) underwent an MRI, followed by CT angiography to assess for preoperative embolisation. Most patients (75.0%) underwent a single session of debulking, 17.9% underwent 2 sessions and 7.1% underwent 3 sessions.

Six (21.4%) patients underwent angioembolisation of the AVM prior to surgical debulking. After 24 h, the patients underwent surgery. Polyvinyl alcohol was used for embolisation in our centre. No record was available for patients who underwent embolisation outside of our facility.

Reconstruction was planned in accordance with the defects (Figs. 1-4). In 85.7% of the patients, reconstruction was done using free flaps and local flap coverage was performed in 14.3% of patients. In our department, radial forearm flaps were used in 18 patients (62.3%), anterolateral thigh flaps in 5 patients (17.9%) and vascularised fibula in 1 patient (3.6%). Seven (25%) patients needed secondary procedures for aesthetic improvement, which included flap debulking, lateral canthopexy, oral commisuroplasty and tissue expansion for restoration of scalp hair. One patient (3.6%) requested for dental implants in the upper alveolus post-resection of her involved maxilla. She was a young patient and followed up after 4 years of primary debulking. She had undergone subtotal maxillectomy and reconstruction with a de-epithelised anterolateral thigh flap. Her upper alveolus was reconstructed using a vascularised fibular graft. The patient recovered uneventfully and is scheduled for dental implants.

Complications ranged from remnant scarring (38.6%), mild bleeding not requiring active surgical attention (29.7%), hematoma formation (20.1%) and suture line dehiscence (11.6%). No flap failure was observed in any of the patients and these complications were dealt on an outdoor basis. Follow-up ranged from 6 months to 4 years. In all patients the minimum follow-up time was 6 months. During this time, 1 patient developed recurrence. Recurrence was observed more frequently in the younger

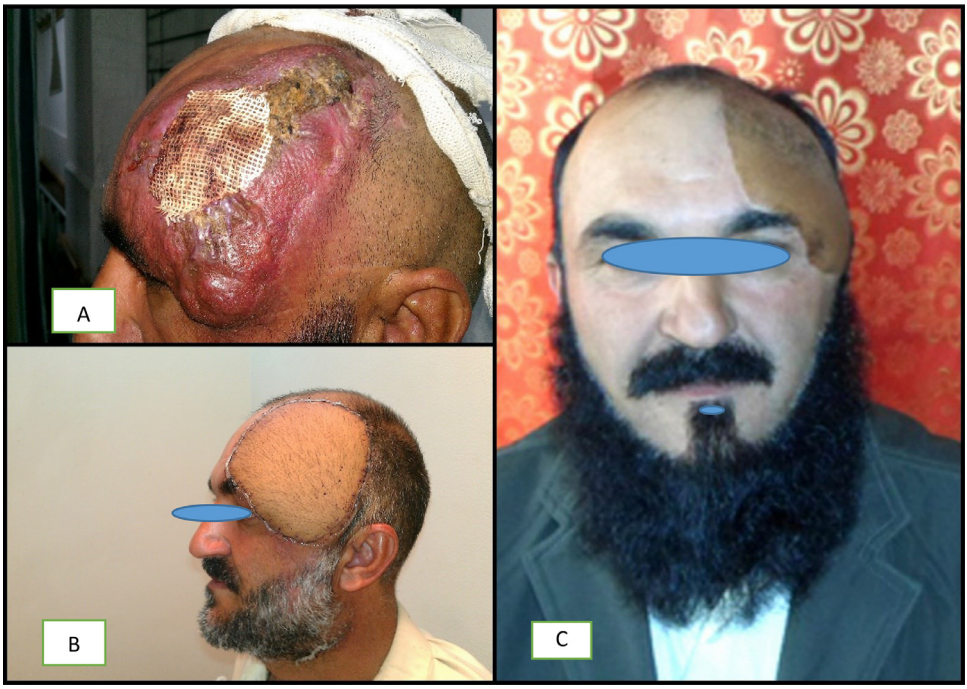

Figure 1. A: A 54-year-old patient with AV malformation over the left forehead, extending to the scalp; the lesion is ulcerative in nature. B: Post-resection image of the lesion and coverage with an anterolateral thigh flap, early postoperative period. C: The flap healed at the 9-month follow-up.

age group and in those who did not undergo complete excision of the malformation. Four patients developed recurrence after the initial 6-month period.

Discussion

Large AVMs of the maxillo-facial region are disfiguring owing to their location and nature. They cause significant morbidity and mortality in children and adults.[6] Most AVMs (90%) occur intracranially. In a series by Visser et al., extra cranial AVMs were present in only 4.7% of the 1131 patients with vascular anomalies who were referred to the centre of vascular anomalies over a 14-year period.[7] These lesions are diagnostically and surgically challenging. Most cases have delayed presentation due to misdiagnosis or lack of available treatment. Furthermore, these lesions may be associated with several different syndromes.

The onset of symptoms is usually before the age of 30 years,[8-10] our patients also presented in a similar age group. The course of vascular malformation correlates with the growth of the patient.[10] Hence, as the patient grows, severity of the symptoms worsens and the treatment plan becomes more complex. A study by Hong et al.[11] showed that patients usually present in the age group ranging from 22 to 43 years. Although these lesions occur congenitally, they tend to grow during adolescence.[12] The trigger for growth is still not understood completely. Hormonal changes (puberty, pregnancy and hormone therapy), trauma, infection or iatrogenic causes (biopsies, surgery and embolisation) are known to be possible factors that stimulate progression.[13] However, Liu et al.[14] did not find progression in pregnant mothers with quiescent lesions. We were also unable to delineate any specific factors causing the expansion in our patients.

There was no gender predilection with regard to AVMs. Our study had a slightly higher number of female patients, which could easily be considered as half the study population. A similar trend was observed in other studies.[12,13]

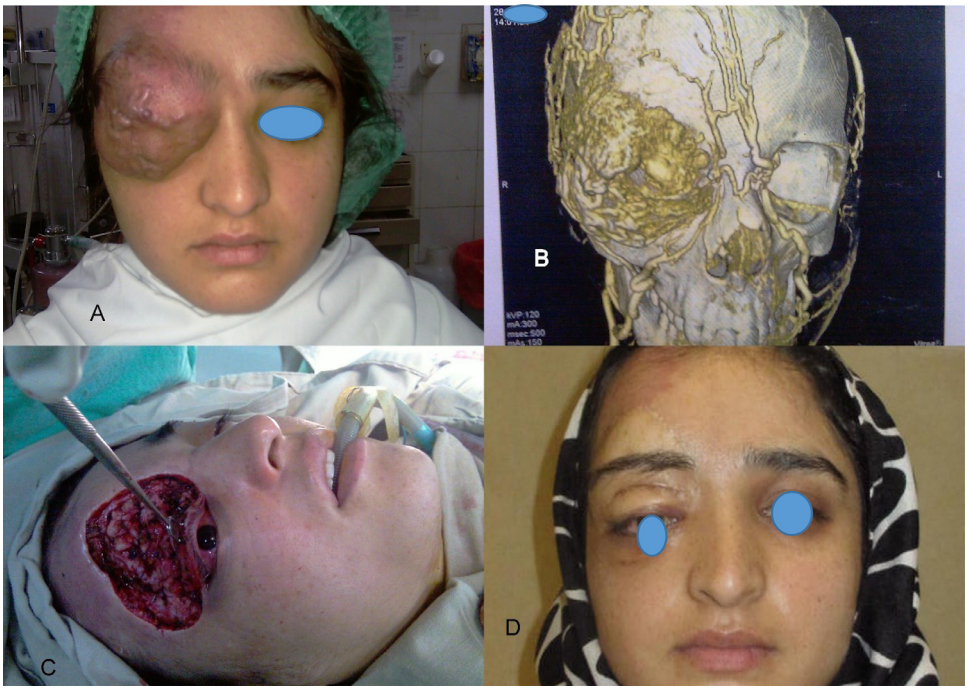

Figure 2. A: A young female with a large AV malformation involving the upper eyelid, which obstructed her vision. B: Angiogram showing the vasculature of the lesion. C: The post-debulking defect, without the involvement of the eye ball. The defect was reconstructed using a radial forearm free flap. D: The patient at the 20-month follow-up after flap debulking and eyebrow hair transplant

Maxillo-facial malformations can cause profuse bleeding, visual obstruction, airway obstruction, ulceration and pain.[15] All patients who presented to us were symptomatic. There were functional impairments and cosmetic disfigurements in these patients.

The diagnostic approach in our study included MRI with contrast and CT angiogram for these lesions. These modalities are proven to be far superior to others and can outline vascular pathology in much larger detail, which ultimately aids the management plan. CT angiography is essential, if preoperative embolisation is being considered. Most studies[10] diagnosed these lesions using colour Doppler and contrast-enhanced MRI, and CT was done in cases with skeletal involvement.

The management of AVMs continues to evolve with the advent of modern technology and medications. Several treatment options ranging from sclerotherapy, angioembolisation and surgical debulking have been employed.[16] In our study, we discussed our cases with an interventional radiologist and worked in liaison with their team. Twenty-four hours post-embolisation, the patients underwent surgical debulking. Literature suggests waiting for at least 1 to 3 days post-embolisation[7,17,18] before embarking on a surgical plan as the hypothesis suggests that longer waiting periods may result in the expansion of the lesion. Pedreira et al.[19] showed in their review that waiting for a long duration enabled better demarcation of the lesion and reduced oedema. They also concluded that the duration of the interim period did not affect the recurrence rates. In our study, no recurrence was noted in patients who were 31 years and above, probably owing to the completion of excisions. Recurrence rates were higher in the younger age groups, in which the patients had undergone partial excisions of the lesion. Goldenberg et al.[18] reported a 15.3% recurrence rate at the 3.5 years follow-up in 31 cases of complete resection using a protocol specifying resection within 72 h of embolisation in most patients.

Complete resection of the malformation provides the best chance of minimising the recurrence rate. However, areas that are technically difficult to reconstruct, such as the aesthetic units of the

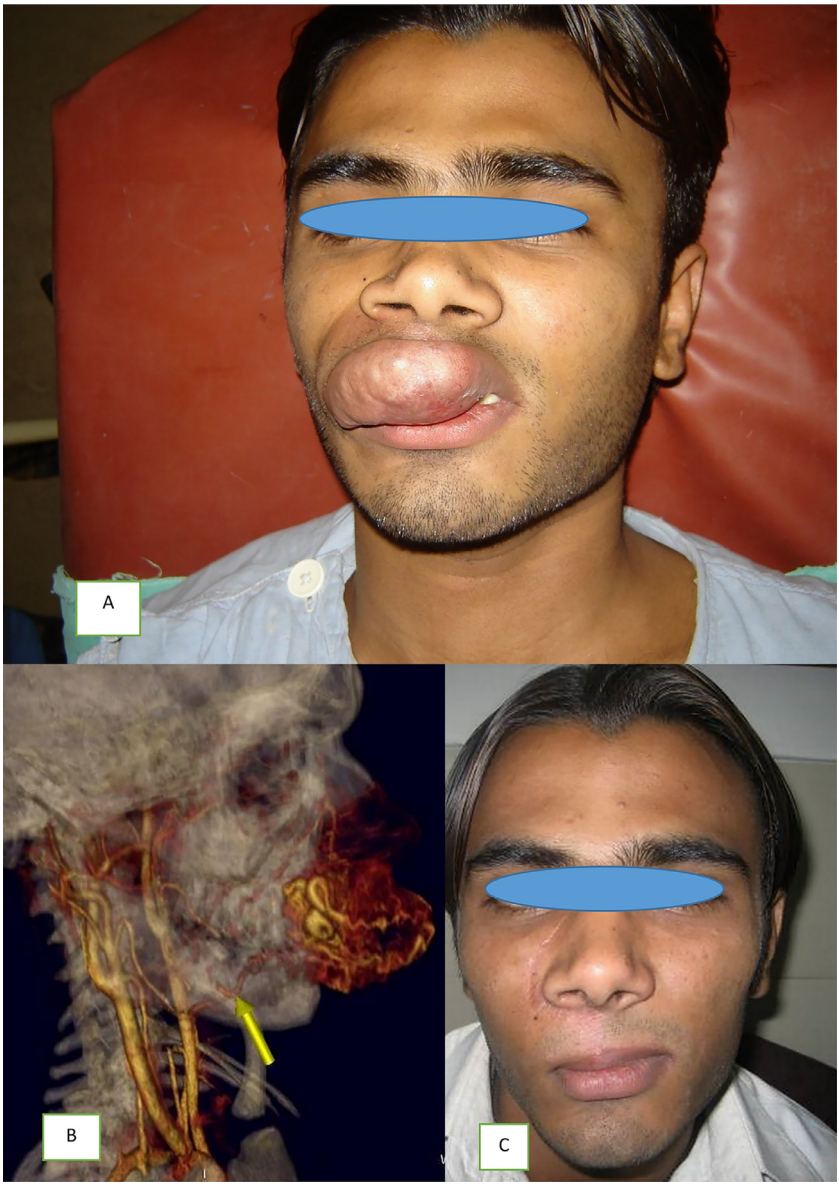

Figure 3. A: AVM of the upper lip. B: Radiological evidence of hyper vascularity. C: Post resection and reconstruction with local flap mobilization; photograph taken at the follow-up at 24 months.

face, pose a challenge to the surgeon. In our study, the primary importance was given to removing the nidus and completely excising the lesion where possible. Post-embolisation, identification of feeders was carried out using a pencil Doppler, and these were ligated prior to excision of the lesion. This technique provided good surgical control of the malformation and aided in controlling blood loss. Coverage was provided according to the defect, and secondary procedures were carried out later to improve facial symmetry and contour. In their case report, Wu et al.[20] highlighted the importance

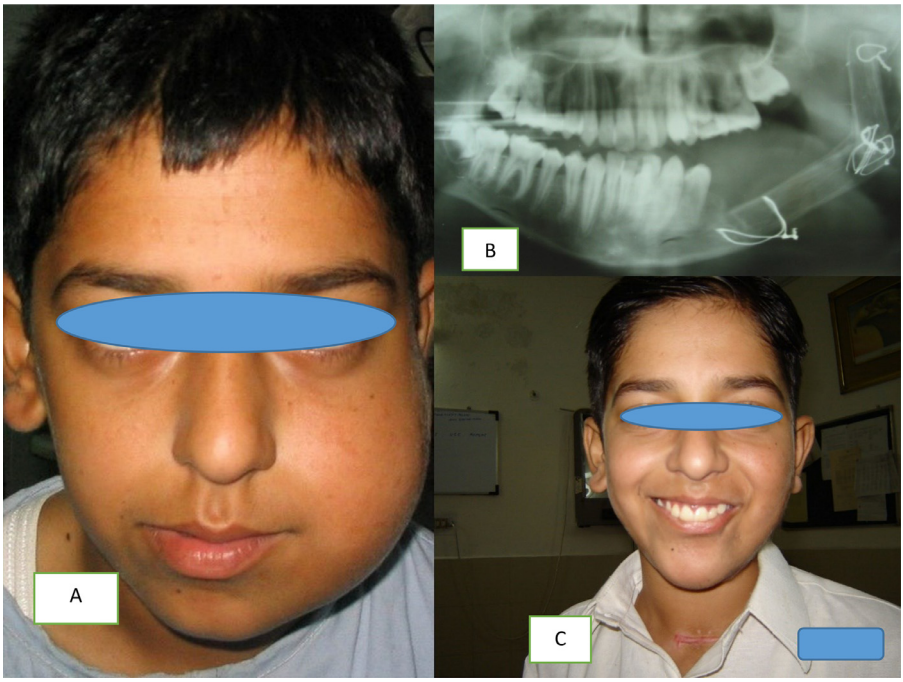

Figure 4. A: A young male with AV malformation of the mandible and overlying mucosa. B: Resection and reconstruction with vascularized free fibula as shown in the Orthopantomogram. C: Follow-up at 28 months showing excellent symmetry

of immediate reconstruction post-resection of facial AVM. Moreover, similar studies have endorsed immediate reconstruction after malformation excision.[7,21–23]

Free flaps have normal vascularity and are thought to suppress the hypoxic cycle responsible for AVM growth.[24,25] Some studies[11,15] suggest that free tissue transfer with their normal physiology acts as a "regulating flap" by suppressing residual AVM by not supporting collateral formation. Bradley et al. suggest that coverage using a free flap is a safe option, if the microvascular anastomosis is placed away from large pathological vessels.[26] On the contrary, some studies suggest using the feeding vessel as a recipient vessel for free tissue transfer, Yamamoto et al.[27] reported 12 cases in which they used the feeding vessel for anastomosis and reported no thrombus formation post-operatively. However, we avoided using pathological vessels for anastomosis and performed all anastomosis in the neck, except for one case where it was done with superficial temporal vessels. In this case, the flap was an anterolateral thigh flap, which had a short pedicle length and was inset over the forehead region; hence, it was feasible to anastomose with superficial temporal vessels.

Complications in such cases are numerous and range from simple wound infections to life-threatening on-table bleeding. No patient in our study experienced massive per-operative bleeding or flap failure. Wound complications were managed in the outpatient clinic. Secondary procedures were carried out for 7 (25%) patients. These procedures did not include re-excision of the remnant malformation or recurrence. These procedures were deemed necessary to enhance the aesthetic outcome of the patient. Another study[19] outlines the importance of secondary aesthetic procedures.

The recurrence rate (17.9%) in our study is comparable to those of few other studies.[18,19] Recurrence was observed more frequently in the younger population in our series, which could be attributed to the fact that these patients were growing and the remnant malformation grew with time. When compared with a similar study, the recurrence rate was dictated by the size of lesion, with the largest dimension being more than 6 cm. No such correlation was observed in our series. Kansy et al.[28] indicated that combination treatment (embolisation and surgery) had superior results in terms

of recurrence and disease control than monotherapy. They performed a meta-analysis, which also confirmed the above statement and reported lower success rates in non-resectable malformations that were managed with palliative treatment.

Conclusion

Management of vascular malformations has not been investigated in depth. Hence, no standard treatment algorithm is available. With the advent of technology and interventional angioradiology, the prognosis has improved immensely.[15] The management protocols are flexible and tailored individually to suit each case.

Conflict of interest

No conflict of interest.

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Statement of consent

Written consents were obtained from all patients for the publication of their photographs in the article.

This study was approved by the ethical review board (IRB# 0182-22)

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