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AURICULAR EXTRA-CRANIAL ARTERIOVENOUS MALFORMATION CAUSING TORRENTIAL BLEEDING: A CASE REPORT.

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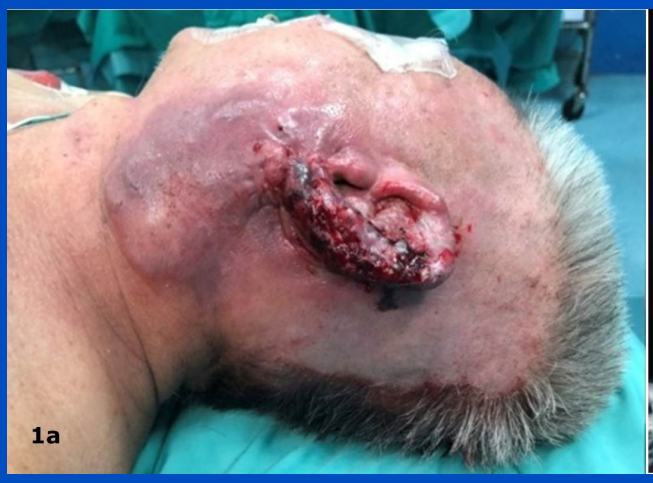

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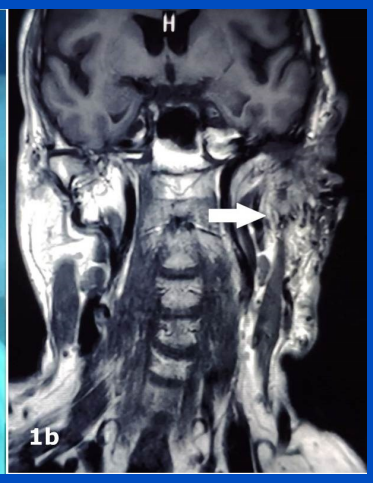

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ABSTRACT

Arteriovenous malformations in the extracranial region are uncommon. Most of these arteriovenous malformations are found in the paediatric age group, while some remain inert until adolescence or even adulthood. Radiological modalities play an important role in assessing these lesions and facilitate their management. When the head and neck region is involved, the goals of any treatment plans should evolve around both functional and aesthetic outcomes. Herein we report an unusual case of an arteriovenous malformation involving the left pinna causing torrential bleeding which was successfully managed surgically albeit without the aesthetic or functional components which may be revised in a future setting.

KEYWORDS: Arteriovenous Malformation, Angiography, Congenital Abnormalities, Ear Auricle, Haemorrhage

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ABSTRACT

Arteriovenous malformations in the extracranial region are uncommon. Most of these arteriovenous malformations are found in the paediatric age group, while some remain inert until adolescence or even adulthood. Radiological modalities play an important role in assessing these lesions and facilitate their management. When the head and neck region is involved, the goals of any treatment plans should evolve around both functional and aesthetic outcomes. Herein we report an unusual case of an arteriovenous malformation involving the left pinna causing torrential bleeding which was successfully managed surgically albeit without the aesthetic or functional components which may be revised in a future setting.

KEYWORDS: Arteriovenous Malformation, Angiography, Congenital Abnormalities, Ear Auricle, Haemorrhage

INTRODUCTION

Arteriovenous malformation (AVM) is a congenital anomaly characterized by an aberrant shunting between artery and vein from an incomplete involution of the foetal capillary bed.[1] It represents an uncommon entity with an estimated prevalence between 0.06% to 0.11%.[2] Approximately 40% of the lesions are clinically evident at birth.[3] AVM is reported to have a slight female predominance with a ratio of 1: 1.5.[4] Most lesions are identified intra-cranium.[5] The head and neck region constitute the most common site for occurrence of extra-cranial AVM, accounting for around 40% of all cases.[6] The rarity of the disease coupled with overlapping vernacular and histopathologic terms often contributed to diagnosis inaccuracy hence unsatisfactory treatment outcome.[3,7] The management of AVM of the head and neck region typically imposes therapeutic challenges. Herein, we present a rare case of an extensive long-standing AVM involving the left pinna, external acoustic meatus and parotid gland along with its management. We would like to highlight the challenges and dilemma encountered when dealing with patient presenting with acute life-threatening haemorrhage from the AVM, which failed initial control with angio-embolization and required urgent salvage surgery.

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CASE REPORT

A 62-year-old gentleman was rushed to the emergency department with massive spontaneous bleeding from his left ear. He was known to have suffered from AVM of the left

pinna for the past ten years but refused surgical excision. Throughout the years, he had experienced eight episodes of spontaneous bleeding from the diseased ear which was controlled by embolization.

On examination, the bossellated left pinna appeared erythematous and disfigured with visible pulsation. The view of the left external acoustic canal was obscured by the overlying oedematous skin. There was profuse bleeding from the ulceration at the helical part of the ear which has temporarily stopped with compression dressing (Figure 1a). Magnetic resonance imaging (MRI) of the head and neck region showed multiple serpiginous flow void vessels involving the left pinna (Figure 1b). The malformation extended medially to involve the parotid gland. Inferiorly, it infiltrated the subcutaneous tissue overlying the left sternocleidomastoid muscle. The deep neck spaces were spared from disease. A subsequent angiogram revealed multiple feeding arteries originated from the left external carotid artery. As a result of vasoconstriction possibly secondary to haemorrhagic shock, an attempt to occlude the feeding arteries with embolization in the same setting was unsuccessful. Repeated compression with adrenaline dressing (1:10000) was the only avenue to reduce bleeding. The estimated blood loss over the span of an hour was one litre. Three units of red cell concentrates, and two units of fresh frozen plasma were transfused, and his vital signs were stabilized. The patient and his family were counselled thoroughly on the possible catastrophic outcomes should the bleeding recur. The option of surgery was again discussed. The possible risks and long-term sequel of wide local excision of the AVM with free flap reconstruction were explained in detail, of which he fully understood and finally consented for.

In view of the complexity of the disease, the patient was transferred to a tertiary care centre equipped with multidisciplinary subspecialties which include head and neck oncology, interventional radiology, and plastic and reconstructive surgery. In an attempt to reduce the risk of intraoperative haemorrhage, a repeated preoperative angiography was performed with the aim of embolization. The angiogram revealed multiple feeding vessels arising from the left posterior auricular and occipital arteries. Embolization was successfully performed using n-butyl cyanoacrylate glue (n- BCA) to occlude the nidus. A left

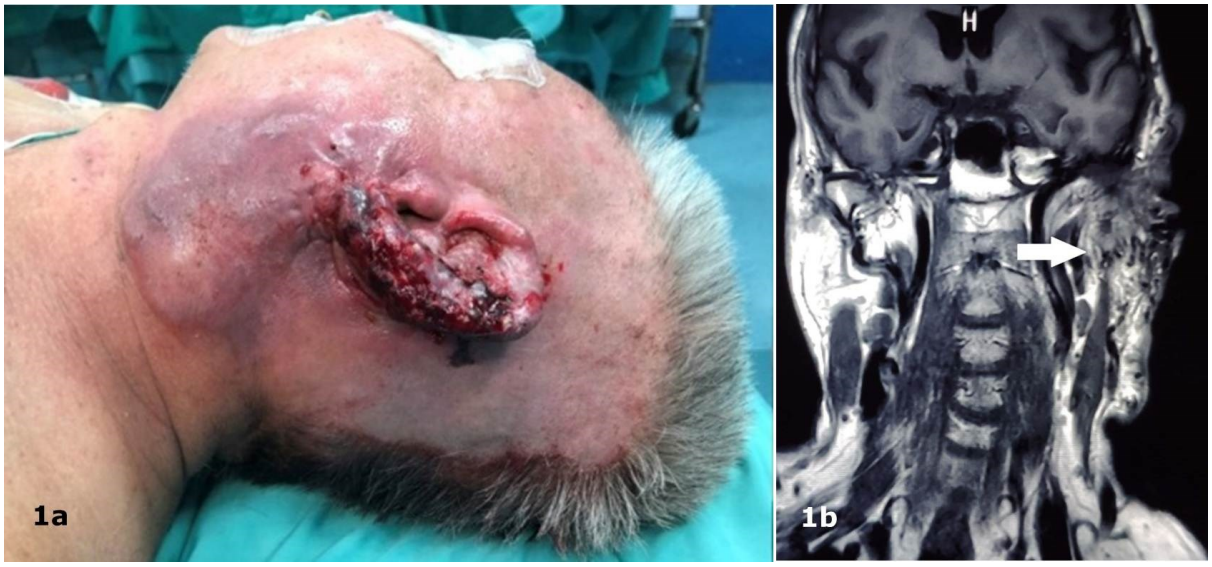

Figure 1: (a) Preoperative image of left pinna AVM; bleeding ulcer was seen over the helical part of pinna, (b) MRI of neck with T1 weighted coronal image showed multiple serpiginous flow void vessels involving the left auricle and parotid gland (white arrow) .

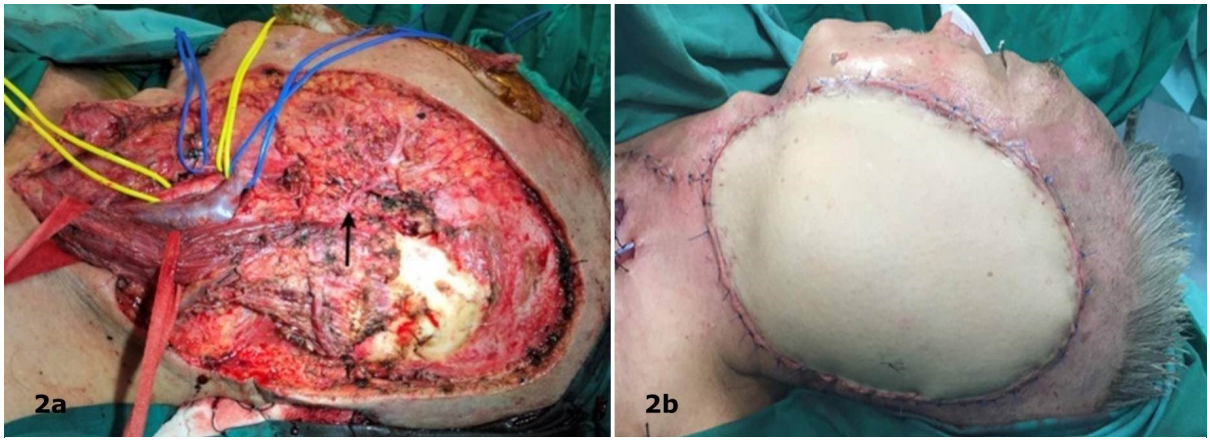

Figure 2: (a) Intra-operative image showing preservation of all vital structures. Facial nerve branches were preserved (black arrow). (b) Intra-operative image showing defect which was reconstructed with vastus lateralis myocutaneous free flap.

total auriculectomy, mastoidectomy, excision of arteriovenous malformation nidus and flap reconstruction were performed. The facial nerve branches were carefully isolated and preserved (Figure 2a). The left external acoustic canal, middle ear space and eustachian tube were obliterated with temporalis muscle and bone wax. The defect was later reconstructed with vastus lateralis myocutaneous free flap which measured 20cm x 15cm in size (Figure 2b). The vascular pedicle of the flap, which was from the descending branch of the right lateral circumflex femoral vessels were anastomosed to the facial artery and vein. Histopathological examination of the resected specimen showed multiple dilated and anastomotic arterial and venous components.

Post-operatively, the patient was commenced on systemic broad-spectrum antibiotics. Patient-controlled analgesic (PCA) morphine with antiemetics was given. A meticulous flap observation including its colour, texture, capillary refill along with the Doppler signals were monitored from time to time to ensure its viability. Chest physiotherapy was started on post-operative day three as well. Fortunately, he recovered well and was discharged after two weeks of hospitalisation. Upon follow-up in six months, no clinical evidence of recurrence was demonstrated (Figure 3). A further recommendation for an objective assessment with CT-angiogram was however declined by patient.

DISCUSSION

Vascular anomalies are broadly classified into two main categories, namely the proliferating vascular tumours and the vascular malformations.[3,8] In addition to their clinical behav-

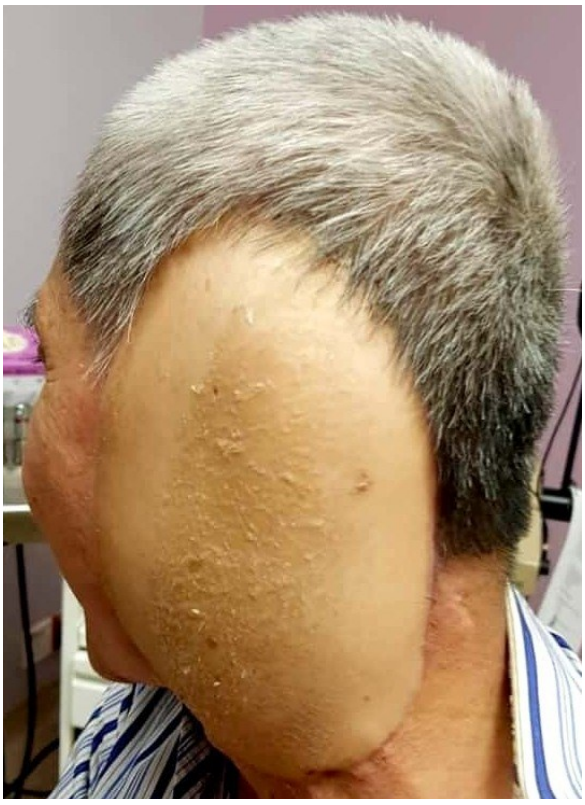

Figure 3: Flap remained viable with no clinical evidence of recurrence on post-operative 6 month.

iour and histological variation, the International Society for the Study of Vascular Anomalies (ISSVA) incorporated blood flow characteristics into refining the classification of vascular anomalies. AVM is grouped under the umbrella of vascular malformation, with its distinctive characteristic of increased vascular flow.[1,3,8] Contrary to their counterparts, vascular malformations have normal rate of endothelial cell division.[3,4] They generally do not regress but grow with advancing age.[4,8] The contingency of vascular developmental error result in anomalous communication (central nidus) between the arterial and venous system thereby bypassing the highly resistance capillary bed.[1,3]

The pathogenesis for the formation of an AVM remains unclear with various hypotheses postulated. These include the possibility of genetic mutation and family inheritance.[8,9] Schobinger et al conventionally classified AVM into four stages from an indolent small lesion to a disfiguring expansile mass that is potentially life threatening which allows stratification of the severity of lesion and facilitate further treatment decision.[6,8]

The diagnosis of an AVM can be made based on clinical judgement with confirmation by radiological modalities, as shown in our patient. Its presence is evidenced by feeding arteries of corkscrew appearance with early opacification of dilated veins in angiogram.[3,9] MRI is invaluable in depicting the extent of the lesion within tissue plane.[8,9] The lack of enhancement of the adjacent soft tissue on T2 weighted post-contrast imaging differentiates AVM from other vascular malformation.[3,7]

Being notoriously known for its infiltrative nature with high tendency of recurrence, the management of peripheral AVM is complex and technically challenging.[10] Various factors that influence the outcome of treatment include the site of occurrence, size of lesion, clinical staging, numbers of feeding arteries, individual fitness and availability of expertise are considered during initial assessment. A multidisciplinary approach involving otorhinolaryngologists, interventional radiologists and reconstructive surgeons are advocated to treat head and neck extra-cranial AVM. An asymptotic or early stage AVM is generally treated conservatively with frequent follow up.[4,11] However, staged intervention consisting of pre-operative embolization in conjunction with surgical excision is the preferred treatment modality.[11]

Embolization serves to obliterate the feeders while preventing collateral formation.[6] Besides reducing the risk of intraoperative blood loss, embolization allows downstaging of the lesions and relieve symptoms. The ideal interval between pre-operative embolization and surgery is still debatable, with reported cases varied from one to fourteen days.[1] In our case, embolization was done 24 hours prior to surgical intervention. A variety of embolizing agents have been used in accordance to the availability and preference of the centre.[2] Endovascular intervention as a sole treatment for AVM is rarely employed and often reserved for unresectable or palliative cases.[7]

Conventional dogma in the cure of an AVM dictates a complete surgical eradication of the lesion.[4,11] Thorough evaluation of preoperative imaging to identify the limit and extent of the lesion is paramount to ensure success in surgery. Ligation or proximal embolization of feeding vessels must be avoided to prevent recruitment from nearby arteries and trigger further enlargement.[7,12] This was greatly reflected in our case. Despite repeated episode of bleeding with progressive enlargement of lesion, patient refused for surgical intervention in the initial stage and had undergone multiple embolization. Needless to say, the sequalae led to a catastrophic haemorrhagic event.

CONCLUSION

AVM represents a rare inborn oddity. A remarkable advancement in science and technology in recent decades has improved the understanding and appreciation of its definition and clinical behaviour. Multidisciplinary approaches have proven to be beneficial. Stratification on the stages of the disease should always be made diligently to provide guidance for the most appropriate management. An advanced stage AVM, as reflected in our case, is best treated with a combination of pre-operative embolization followed by surgical resection. For patient who refuses initial surgical intervention, it is of the utmost importance to explain thoroughly regarding the possible lifelong morbidities and even mortality. The patient should be followed up regularly to assess the risk of haemorrhage and counsel for the best outcome.

DECLARATION OF CONLICT OF INTEREST

The authors declare no conflict of interest. Patient was consented for the publication of images and report.

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