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Management of Extra-cranial Arteriovenous Malformations of the Scalp: A case report

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Abstract

Arteriovenous malformations (AVMs) are congenital fast-flow vascular malformations characterized by abnormally tangled arteries and veins connected by abnormal shunts without intervening capillaries. The growth of AVMs is usually initiated by trauma and hormonal changes in puberty. Compared to the intracranial AVMs, extracranial AVMs are rare. The treatment of extracranial AVMs, though poses a challenge to the surgeon, is debilitating to the patients.
We report a case of extracranial arteriovenous malformation of the head in a young man who noticed it from childhood. He was clinically and radiologically evaluated using CT angiography and had two-stage surgical interventions.

Keywords: Arteriovenous Malfornations, Extracranial, Surgical Intervention, Case Report.

Introduction

Arteriovenous malformations (AVMs) are congenital high-flow vascular malformations characterized by abnormally tangled arteries and veins connected by abnormal shunts with absence of normally intervening capillaries. These arise during intrauterine life and manifest during childhood or adulthood with occurrence of rapid growth following trauma or hormonal changes in puberty or pregnancy. The lesional enlargement arises from changes in pressure and flow, vascular dilation, shunting and collateral proliferation and not necessarily from cellular proliferation.[1] Arteriovenous malformations may be misdiagnosed as other vascular malformations like superficial haemangioma which has initial cutaneous blushing and proliferation in the first weeks after birth.[2] Apart from recurrence, AVMs can be complicated by life-threatening hemorrhage or congestive cardiac failure.[3,4]
The definitive diagnosis of AVMs can be made using Doppler ultrasonography, CT scan with contrast and/or magnetic resonance imaging.[5] Selective embolization under angiogram guidance followed by surgical excision is the treatment of choice.[1] Complications such as bleeding, ulceration and local gigantism are managed in a multidisciplinary manner.
The relevance of this study is to highlight the possible challenges which can be encountered in the management of AVMs in a resource poor setting like ours.

Case Illustration

A 24 year old male who electively presented via surgical outpatient department on account of pulsatile tortuous scalp swelling of over 20 years duration. The lesion was initially about the size of a coin and rapidly increased in size over the last 12 years, involving the entire scalp. He had one episode of bleeding 18 years ago following attempt at excision by the mother and another bleeding episode 5 months prior to presentation following scalp trauma which was managed by a patent medicine dealer. He sought care in our tertiary health facility because of intermittent

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bleeding from a scalp wound. No history of neurological deficit or features of heart failure.

Examination was unremarkable except for the ulcerated irregular scalp swelling involving the frontal, temporal, parietal and occipital regions. The swelling was pulsatile with thrill and bruit. The ulcer was irregularly shaped, measured 2 cm in its widest diameter with some sloughs in the floor.

Brain CT scan was unremarkable but CT angiography made impression of arteriovenous malformation of the right external carotid artery and the tributaries of the right internal jugular vein. Magnetic resonance angiography could not be done due to financial constraint.

Distal control was achieved by dissecting and looping the external carotid artery. A pre-auricular incision was made for ligation of the right superficial temporal artery followed by ligation of some dilated, tortuous venous channels and excision of some vascular masses. He had completion surgery by excision on the 9th day after the first surgery. He had unremarkable post-operative recovery and was discharged on the 11th day post-op. He received eight (8) units of blood which he paid for as altruistic blood donations are not routine in our West African subregion. Examination on follow up showed great resolution of the swelling and healed scalp wound. However, patient has been lost to follow up probably due to financial constraints.

A = Figure 1: Pre-operative Clinical Picture
B = Figure 2: Preoperative Clinical Photograph in the theater

First Stage Surgery: Intra-operative Pictures

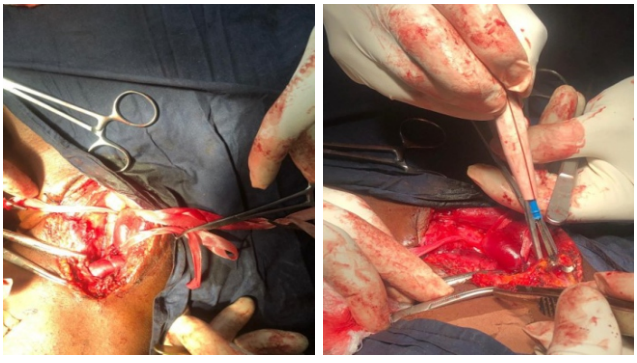

Figure 3: Intra-operative Photographs showing attempt to proximally control the bleeding by exposing both the common and external carotid arteries as well as the internal jugular vein

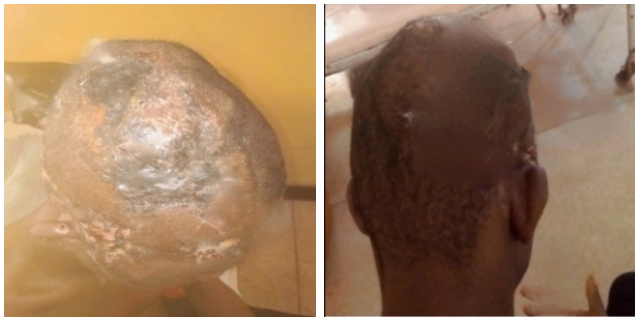

Figure 4: Post-operative Photographs

Discussion

Some AVMs of the head may not present with neurological symptoms because most of the draining veins are extracranial. In some rare situations, a patient may have neurological symptoms as well as an intracranial communication of the draining veins.[6] So, our patient who had extracranial AVM never had intracranial symptoms nor signs. Arteriovenous malformations are the most aggressive type of vascular malformations and can cause significant deformity and functional impairment.[7] Spontaneous regression of AVMs is rare and this occurs at the rate of 0.8% to 1.3%.[8] After some period of quiescence, a rapid growth and altered hemodynamics of the AVMs can be triggered by mechanical stimuli like trauma, incomplete surgery or hormonal surge as seen in puberty and pregnancy.[9] This was evident in our patient who had first growth episode following unsuccessful resection of the lesion by the mother and the second growth phase at puberty (12 years). The etiology of AVMs is unknown, but there are theories suggesting some genetic predispositions like loss-of-function mutations in endoglin and activin receptor-like tyrosine kinase which encodes a TGF-β family type I receptor, loss-of-function mutations in the RASA1 gene, and loss-of-function mutation in PTEN gene.[10,11,12,13] The clinical features of AVMs depend on the anatomical location and the Schobinger clinical stage.[14]

Stage I (quiescence phase): Local warmth mass, blush or red stain. Doppler ultrasound: increased vascularization with arteriovenous shunting.
Stage II (progression phase): stage I lesion + expanding tortuous draining veins, pulsation, thrill, and bruit. Doppler ultrasound: progressing arteriovenous shunting.
Stage III (destruction phase): stage II lesion + dystrophic skin changes, ulceration, and bleeding from increased total blood flow or tissue necrosis, and persisting distal ischemic pain from worsened arterial steal. Soft tissue and bone hypertrophy may occur.
Stage IV (decompensation phase): high output cardiac failure (only the largest AVMs reach this stage).
From this staging system, it can be deduced that our patient fit into stage III as he presented with extensive scalp swelling causing some dystrophic skin changes and ulceration following bleeding from trauma he had 5 months prior to presentation. He did not have pain possibly due to good blood supply of the scalp.
There are other classification systems for AVMs like the Mulliken classification, Hamburg classification, Yakes classification, Houdart classification and Do classification. The new Yakes AVM classification system has proven therapeutic implications such that it can be effectively applied in the treatment of complex AVMs in any anatomical region.[15] The Houdart classification of intracranial arteriovenous fistulae and malformation of high-flow lesions and the Do classification of AVMs of the peripheral arterial circulation share some similarities despite the difference in their anatomic locations (intracranial Vs peripheral vessels).[16,17] The two classification systems also share similar therapeutic measures.[16,17] The classification system of vascular anomalies adopted by the International Society for the Study of Vascular Anomalies helps in the standardization of diagnosis and treatment.[18]
The following imaging modalities can be of help in making a definitive diagnosis of AVMs: Doppler ultrasonography, CT scan with contrast and/or magnetic resonance imaging.[5] Contrast CT scan can demonstrate AVMs as an enhancing lesion with single or multiple dilated inflow and outflow vessels.[19] This was evident in our patient who had cranial CT angiography which showed arteriovenous malformation of the right external carotid artery and the tributaries of the right internal jugular vein. Magnetic Resonance Angiography (MRA) is considered an imaging modality of choice as it can delineate the extent of the lesion (the nidus), gives multiplanar images, and differentiates high-flow from low-flow lesions.[20] This was, however, not possible in the index patient, again due to financial constraints.
Treatment of AVMs can be challenging to the surgeon especially in centers where interventional radiological services are not readily available for pre-surgical lesional embolization. This was the challenge in our patient management, thus, leading to two-stage surgical resections. Isolated therapy like surgical ligation or embolization of the arterial supply alone should not be done as there is possibility of neovascularization from adjacent area to maintain supply to the nidus.[5] However, whenever the isolated artery ligation is done, every attempt should be made to completely resect the nidus to avoid the complications mentioned above.

Conclusion

Considering the progressive nature of the lesion and its rare chance of spontaneous obliteration, it is necessary for every clinician to be familiar with the clinical features to avoid wrong diagnosis. Also for the fact that trauma and hormonal surge trigger the growth of the lesion, early diagnosis and multidisciplinary based treatment modalities are of paramount importance to avoid challenges of managing Schobinger stage III or IV lesions. Combination of endovascular embolization, surgical resection and reconstruction, and stereotactic radiosurgery is advisable to reduce the chance of recurrence of the lesion. Close follow up is mandatory to ensure non-recurrence of the lesion. Health insurance and altruistic blood donations should be promoted in our subregion. This will help in the comprehensive and better management of patients.

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