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Article

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Cirsoid Aneurysm of the Scalp: a case report

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Abstract: Cirsoid aneurysms (arteriovenous malformations [AVMs]) are Anomalous fistulous arteriovenous communications of scalp with ill-defined natural course that are rarely encountered in neurosurgery. Patients with AVM of the scalp present clinically with headache and either a small innocuous-looking subcutaneous scalp lump or a large, pulsatile mass with or without bruit, which has a propensity to massive hemorrhage. Complex vascular anatomy and interconnections and high shunt flow make their management difficult. We report a rare case of a 40-year-old man who presented with a swelling over his occipital region that progressively enlarged over the course of 6 years. Being high flow shunt surgical excision was attempted.

Key words: AVM- arteriovenous malformation, scalp, high flow shunt, occipital, cirsoid, Arterio venous fistula

Introduction

Aberrant persistence of primitive arteriovenous interconnections due to defective differentiation of the primary vascular complex leads to formation of arteriovenous malformations (AVM).AVM of scalp are rare occurrences among vascular lesions. Various names being used to describe the vascular malformations of the scalp include aneurysm cirsoid, aneurysma serpentinum, aneurysm racemosum, plexiform angioma, arteriovenous fistula and arteriovenous malformation [1]. Arteriovenous fistula was first described by Hunter in 1757. [2] The term cirsoid aneurysm was applied to vascular malformations of the scalp in 1833 by Brecht [2] and is used to describe a fistulous connection between the arterial feeding vessels of the scalp and the draining veins without an intervening capillary bed. AVM are composed of complex tangle of feeding arteries and draining veins, without an intervening capillary bed forming a 'nidus'located within the subcutaneous layer. The draining veins often are dilated owing to the high velocity of blood flow through the fistulae. The location of scalp arteriovenous fistulas is roughly evenly distributed among the frontal, temporal and parietal regions [1]. The incidence of cirsoid aneurysms of scalp is rare and infrequently encountered by the

neurosurgeon. [3] They are usually congenital in etiology however traumatic fistulas have also been described.. Whatever the cause,Clinically patient presents usually with an innocuous looking subcutaneous scalp lump or a visible large, pulsatile scalp mass associated with headache, tinnitus and hemorrhage. [4] A clear understanding of the diagnostic and treatment algorithms involved with AVM management is imperative, because AVMs are a cause of haemorrhage in young adults. Surgical treatment is primarily indicated in order to prevent bleeding and haemorrhagic complication along with resolution of cosmetic problems [1]. However the treatment of these lesions is difficult because of their complex vascular anatomy, high shunt flow, and cosmetic disfigurement. The treatment options of these lesions include endovascular treatment, direct intralesional injection of sclerosing agents, ligation of feeders, and surgical excision. [5–10]

In this case report we describe the clinical features and discuss the results of the surgical management of scalp vascular malformations.

Case report

A 40 year male complained of slowly progressive swelling localized over right occipital region of the scalp since around 5-7 years duration. It was associated with occasional headache and tinnitus. The swelling had been gradually increasing in size since 6 yrs. and was now pulsatile. There was no previous history of trauma or head injury. No history of recurrent massive bleeding, any visual disturbances or paresis. Neurological examination was normal. There was no other systemic abnormality detected. Local examination showed large swelling around 6cm-7cm in diameter, in midline extending over the right occipital region (Figure 1). The swelling was densely adherent to scalp. Local temperature was not raised. The swelling was pulsatile, nontender and soft in consistency. A bruit was also demonstrated over the swelling. Multiple tortuous & prominent, intensely enhancing vessels along occipital surface of the scalp was evident on Computerised tomography scan. MRI scan revealed a vascular lesion under the scalp in the occipital region containing tortuous dilated high contrast filled vessels. Digital subtraction angiography showed large occipital complex vascular network with simultaneous early filling of venous circulation in the scalp confirming the presence of an arteriovenous fistula (AVM) (Figure 2). The AVM had feeding arteries from bilateral occipital branches of external carotid arteries. Right occipital artery is more hypertrophied, dilated, and tortuous. Venous drainage appears from the communicating emissary veins to the internal jugular veins, and few tributaries to the external jugular veins. It appeared fairly high flow arterio-venous communication. There was no evidence of communication with intracranial circulation showed normal intracerebral circulation.

Surgical procedure

En block surgical excision of the lesion was planned. The patient was positioned with head elevated 30 degrees above the heart level. A question mark scalp flap was planned. After infiltrating 2% lignocaine with adrenaline, scalp incision was made in short

segments, ligating the feeding vessels and draining veins as they were encountered. Once the scalp flap was raised, the bleeding from the bone was controlled with bone wax and monopolar diathermy. After complete devascularization, the lesion along with pericranium, galea was excised circumferentially from the subcutaneous tissue with the use of bipolar diathermy and sharp dissection without breaching the skin (Figure 3 a,b). Intra-op, it was very difficult to get plane of dissection due to very large tortuous venous varix. There was no significant blood loss during surgery. On microscopic examinations, the histopathological specimen contained various well-developed arteries and dilated veins in the connecting tissue. Repeat computed tomography (CT) angiogram showed complete disappearance of AVM without any remnant. The patient was advised to continue follow-up. On microscopic examinations, the histopathological specimen contained various well-developed arteries and dilated veins in the connecting tissue. Endothelial cells and perivascular cells in arteries were positive with immunohistological staining for vascular endothelial growth factor (VEGF); the major feeding arteries and draining vein were VEGF-negative. There was no recurrence of fistula at the 3-month follow-up examination (Figure 3c).

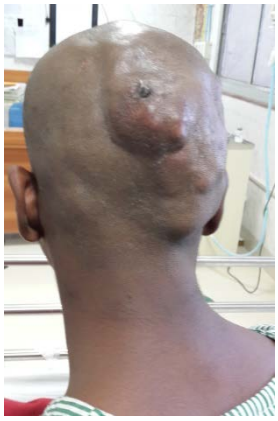


Figure 1 - Rear view pre-op photograph of the patient

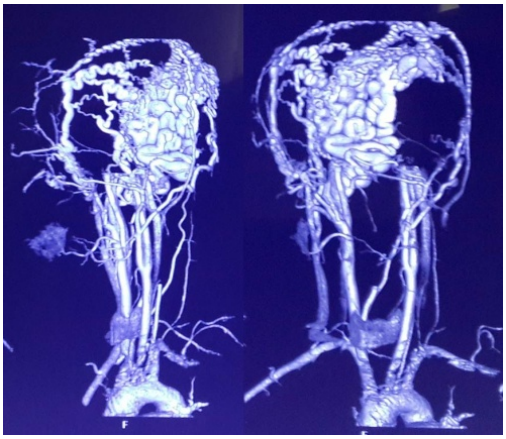
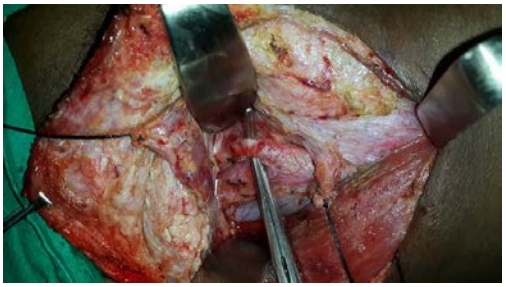
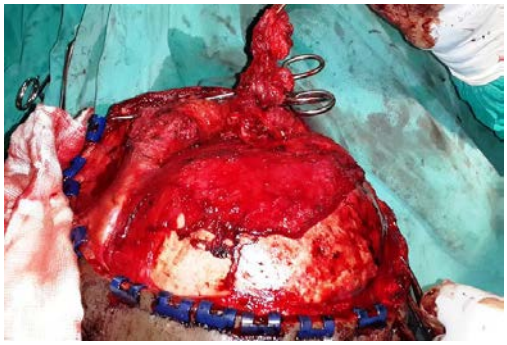


Figure 2 - Pre-op digital subtraction angiography image



A



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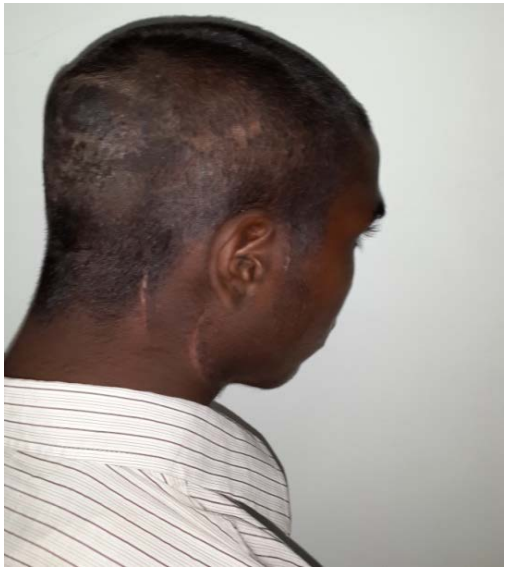

C

Figure 3 - Intra-op photograph before ligation of feeding arteries (a) and en block excision of the lesion (b). Post-op photograph of the patient after surgery (c)

Discussion

Aberrant persistence of primitive arteriovenous interconnections is known as AVM. Scalp AVM are rare despite the intense vascularity of scalp and relatively high frequency of trauma to this region. [3,10] The etiology of cirsoid aneurysm is still controversial. It is accepted that it may be either congenital or traumatic. Congenital was more common in large series published in the literature. [11,12]. In our case, traumatic etiology was the probable initiating factor. Due to the large size and complexity of the lesion, en block excision was planned. Ligation of feeding vessels followed by resection of the lesion has occasionally met with success. Multiple treatment schemes have been described and, as yet, no standard form of therapy exists. Some of the reported treatments include radiation therapy, electrothrombosis, scalp tourniquet, scalp compression with pads and springs, direct injection with alcohol, and embolization. [11,13]. Surgical resection of the fistula is usually successful, as was for this patient. Direct surgical excision risks severe intraoperative blood loss. The abnormal vascular channel dilatation over the scalp often results in deformity of the scalp that is usually not lifethreatening unless it causes hemorrhage but can lead to substantial cosmetic and social disturbances. [8,13] Scalp arteriovenous fistulas are roughly evenly distributed among the frontal, temporal, and parietal regions. Occipital region scalp AVMs have been remotely reported, which is found in our present case report. The AVM feeder vessels mainly arise from the subcutaneous tissue layer of the scalp. The source of feeder arteries most frequently includes the external carotid, occipital, and supraorbital arteries. Clinical manifestations relate to the size of fistula, and patient may present with loud bruit, hemorrhage, headache, and, in severe cases, scalp necrosis. [8]. The lesion usually begins as small subcutaneous lump, which, over a period of time, evolves into grotesque,

deforming mass. Digital subtraction angiography is the gold standard investigation for these lesions, which provides a road map of arterial supply and venous drainage. [3] Although the lesion lies in the subcutaneous layer of scalp, the pericranial component of the lesion is of surgical importance [2]. An important part of surgery is the excision of pericranial component with control of bone bleeding using bone wax. Endovascular and percutaneous occlusions of the fistulas have been described. The results of endovascular occlusion are dependent on the angioarchitecture of the fistula, the supplying arteries, and draining venous structures. Arterial approaches may not often be successful in occluding the entire fistula due to the problem of multiple feeding arteries. In our case, complex multiple feeding arteries were present, so endovascular approach was not considered. Recurrence has been reported as late as 18 years after complete surgical excision [14]. Hence, regular follow-up is recommended.

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