Cirsoid aneurysm of scalp occipital region- A case report
Mahesh Mohanlal Pukar ¹, Ishaan Shrujal Patel ², Sohank G. Mewada ³

Department of Surgery, SBKS medical institute and research centre, Pipariya, Vadodara, Gujarat. 1- Professor, 2&3- Resident

Submission Date: 21-04-2014, Acceptance Date: 23-04-2014, Publication Date: 30-04-2014

How to cite this article:
Vancouver/ICMJE Style

Harvard style

Corresponding Author:
Dr. Mahesh Mohanlal Pukar, Professor, Department of Surgery, SBKS medical institute and research centre, Pipariya, Vadodara, Gujarat. Email: maheshreem11@gmail.com

Abstract:
Anomalous fistulous arteriovenous communications of scalp (cirsoid aneurysms) are infrequent occurrences of vascular origin with ill-defined natural course. Arteriovenous malformations (AVM) of the scalp presenting clinically at times with headache and a small innocuous looking subcutaneous scalp lump or a large, pulsatile mass with or without bruit which has a propensity to massive haemorrhage. These vascular lesions when present over scalp are seen mostly on the fronto-parietal and temporal regions. Complex vascular anatomy and interconnections, high shunt flow and possible cosmetic complications tend to make their management difficult. We report a rare case of high shunt flow scalp AVM of occipital region that progressively enlarged over the course of 6 years by capturing feeders from bilateral occipital arteries. 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 cirsoidae, aneurysma serpentinum, aneurysm racemosum, plexiform angioma, arteriovenous fistula and arteriovenous malformation. 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. Clinical picture presents usually as an innocuous
looking subcutaneous scalp lump or a visible large, pulsatile mass associated with headache, tinnitus. Complex aberrant vascular anatomy, high shunt flow tends to complicate Management of scalp arteriovenous malformation. 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 primary indicated in order to prevent bleeding and haemorrhagic complication along with resolution of cosmetic problems. In this case report we describe the clinical features and discuss the results of the surgical management of scalp vascular malformations.

Case report

A 19 year male complained of slowly progressive swelling localised over occipital region of the scalp since around 5-7 years duration. It was associated with occasional headache, 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 bilaterally over the occipital region. The swelling was densely adherent to scalp. Local temperature was not raised. The swelling was pulsatile, non-tender 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). 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 torcular herophili 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. Intraoperatively under Doppler imaging assistance both occipital arteries were identified. Both the occipital arteries were ligated. High flow was confirmed with Doppler. Subsequently, the dilated scalp vascular malformation was dissected all around and complete surgical excision was performed. AVM was densely adherent to scalp. Post operatively there was no neurological deficit and recovery was uneventful. Long term follow up of five years showed no recurrence. 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.

Discussion

Aberrant persistence of primitive arteriovenous interconnections is known as AVM. AVM of scalp are vascular lesions with rare occurrences. Aneurysm cirsoid, aneurysma serpentinum, aneurysm racemosum, plexiform angioma, arteriovenous fistula and arteriovenous malformation are various terminologies referred to describe the vascular malformations of the scalp. The abnormal vascular channel dilatation over scalp often results in deformity of the scalp that is usually not life threatening unless it cause haemorrhage but can lead to substantial cosmetic and social disturbances. According to Krayenbuhl and Yasargil’s review of 800 cases of AVMs from literature and their own clinical material, extra cranial AVMs to account for only 8.1% of the cases [1]. Pathological Autopsy data suggest an overall frequency of detection of AVMs to be 4.3% of the population [2]. Scalp arteriovenous fistulas are roughly evenly distributed among the frontal, temporal and parietal regions. Occipital region scalp AVM have been remotely reported which is found in our present report case. Feeder n drainers The AVM feeder vessels mainly arise from the subcutaneous tissue layer of the scalp. The feeder arterial system supplying an AVM frequently is multiple and complex, thus making a rich & extensive arterial network by branches. The source of feeder arteries includes, most frequently from the external carotid, occipital, and supraorbital arteries. In our case these vessels originated from occipital branches of external carotid artery. The aetiology of scalp AVMs may be congenital or traumatic. AVM of the scalp may present at birth, but in most patients, it is asymptomatic until adulthood. In a report by et al reported trauma, as cause of cirsoid aneurysms of the scalp, to account for 38% cases [3]. Defective
differentiation of the primary vascular complex lead to formation of arteriovenous malformations (AVM). 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 fistula so they become progressively dilated and tortuous. Various concepts have been proposed to explain the pathogenesis of formation of AVM. In congenital variant type, it is proposed to originate from the anomalous embryonic development of the vascular system. Developmental arrest of the scalp vascular system in the capillary network stage results in the formation of a haemangioma. The persistence of connections of the embryonal capillary network in the later stage leads to the formation of intercommunicating channels of varying forms between the mature arteries and veins. This may result in formation of mixed haemangioma, arteriovenous (A-V) fistula, or both. Blunt or penetrating injuries have been implicated for formation of Acquired AV fistula formation. These lesions undergo hypertrophy over long duration of months to years to become clinically significant. Two distinct mechanisms have been suggested for the formation of the traumatic A-V fistula of the scalp [4]. One of the theories is the disruption theory of the vasa vasorum of the arterial wall in which endothelial cells proliferation from the vasa vasorum into the hematoma around the disrupted vasa vasorum form endothelial buds and numerous small vessels. If these newly formed vessels make contact with the adjacent veins, blood will be shunted from the arterial system to the lower pressure venous system, and thus numerous A-V vascular channels will be created. The other theory is laceration theory in which simultaneous lacerations of the artery and of the accompanying vein result in a single fistula. Clinical features are associated with the size of the AVM. Most of the patients reported in the literature had a history of progressive increase in the size of the lesion and had become symptomatic in the third decade of life. Scalp AVM may present usually as an innocuous looking subcutaneous scalp lump or a visible large, pulsatile mass associated with headache, tinnitus, numbness, and/or haemorrhage. Others may present with severe symptoms such as scalp lesions. Haemorrhage is generally uncommon and may develop in the event of large vascular malformations. Recurrent haemorrhage rapidly deteriorating the neurological table is rarely seen in some of the patients. Investigative workup includes an array of options including MRI scan, CT angiography, Digital Subtraction Angiography scan. MRI is helpful to differentiate scalp AVMs from various other vascular lesions and aid in the correct diagnosis as well as to define any intracranial extension or involvement. MRI can also help to distinguish scalp AVMs which are high flow lesions from other low flow lesions such as venous or lymphatic malformations, and this will help with the treatment planning. However, CT angiography is still the gold standard modality to understand the angioarchitecture of the lesion and to exclude any intracranial component. Advantages of CT angiography (CTA) include shorter acquisition times, retrospective creation of thinner sections from source data, improved 3D rendering with diminished artefacts. CTA can also provide a very high resolution and the visualization of the related adjacent bony structures, which may be important in surgical planning. It is particularly employed for the determination of cranial feeders. Scalp AVMs are most frequently confused with haemangioma and cavernomas. No arteriovenous shunt is present in such pathologies, and they are seen as well-demarcated lesions. AVMs show flow void signs on MRI due to the rapid flow in the lesions. Selective angiography should be carried out for the differential diagnosis of the vascular lesions, such as aneurysms, sinus pericranii, venous malformation, and cavernous haemangioma. The case presented / in the presented case has features of Multiple tortuous & prominent; intensely enhancing vessels along occipital surface of the scalp was evident on Computerised tomography scan. MRI venography revealed extended sequences of all above findings indicative of the lesion to be abnormal arterio-venous malformation. OR 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. there is evidence suggestive of suggest a bilateral supply by the occipital branch of external carotid arteries on carotid angiogram 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 torcular herophili 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. A detailed study of the carotid circulation showed no abnormality in both carotids. The preoperative radiological evaluation should be used for the assessment of feeding arteries, drainage vessels, numbers of fistulas, connected vascular structures, and shunt flow volume in order to prevent any possible complications. Complex aberrant vascular anatomy, high shunt flow tends to complicate Management of scalp arteriovenous malformation. The various other treatment options include surgical excision, ligation of feeding vessels, transarterial and transvenous embolization, Injection of sclerosant into the nidus and electro thrombosis. Surgical treatment is primary indicated in order to prevent bleeding and haemorrhagic complication along with resolution of cosmetic problems. Basic and most preferred approach is complete surgical excision .Various techniques have been used to control the haemorrhage during surgery including percutaneous sutures of the feeding vessels, interlocking suture along the line of incision, and use of tourniquet and intestinal clamp over the base of the flap. Substantial complications during the operation are haemorrhage. Haemorrhage may be prevented with preoperative embolization, clamping, and suturing of feeding vessels. Incomplete surgical resection may also lead to recurrence or deterioration of the AVM. This may cause scalp haemorrhage and necrosis in elderly patients. Despite these treatments, recurrence due to feeding collaterals may develop. Recurrence of the lesion has been seen as late as 18 years after complete surgical resection of the malformation. To conclude, even though diagnosis of scalp AVM can be made easily, a complete angiographic study of the lesion has to be performed to differentiate primary scalp vascular malformations from secondary venous dilatations. 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. Metastatic deposits from follicular carcinoma of the thyroid have to be ruled out prior to the diagnosis of cirsoid aneurysm.[5]. The lesion was located in the occipital region in (33.3 %) cases, frontal region in (22.2 %) cases, temporoperital region in (22.2 %) cases, parietal region in (11.1 %) cases, vertex in (11.1 %). The superficial temporal artery was involved in (77.8 %) cases, the occipital artery was involved in (66.7 %) cases, the posterior auricular artery was involved in (55.6 %) cases, the supra orbital artery was involved in (22.2 %) cases and the middle meningeal artery was involved in (22.2 %) cases [6].

References
5. Leo francis Tauro, G Suhith, Prathvi shetty, and Divakar rao; J neurosci Rural Pract. 2012 Jan-April; 3(1); 95–96.

Source of funding: None

Conflict of Interest:
The authors declare that there are no conflict of interest

Acknowledgement:
Authors acknowledge the immense help received from the scholars whose articles are cited and included in references of this manuscript. The authors are also grateful to authors/editors/publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

Figure 1:
Figure 2:

[Image of an intraoperative picture of a partial lobe AVM]

Figure 3:

[Image of a CT picture of brain]

Figure 4:

[Image of a CT angiography picture]