Case Report

Cirsoid aneurysm of the scalp

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Abstract

Cirsoid aneurysms are rare arteriovenous malformation of the scalp. They are usually congenital in etiology. The term "cirsoid aneurysm" is used because this lesion resembles a varix; the derivation of cirsoid is from the Greek word kirsos, or varix. The superficial temporal artery is the most commonly involved artery. Patients usually present with a pulsatile disfiguring scalp mass which can be extensive. Other presenting symptoms include headache, pulsatile tinnitus, and hemorrhage from the lesion following minor head trauma. Treatment options include surgical resection, endovascular occlusion, and direct percutaneous injection of sclerosing agents. The radiological findings are important for patient management. We report a rare case of high shunt flow scalp AVM of frontal region that capturing feeders from right angular artery.

Keywords: AVM- arteriovenous malformation; scalp; high flow shunt; occipital; cirsoid;

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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 cirsoide, aneurysma serpentinum, aneurysm racemosum, plexiform angioma, arteriovenous fistula and arteriovenous malformation[1-3]. 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[4]. 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 subcutaneous scalp lump or a visible large, pulsatile mass associated with headache, tinnitus. A clear understanding of the diagnostic and treatment algorithms involved with AVM management is imperative, because AVMs are a cause of hemorrhage in young adults. Surgical treatment is primary indicated in order to prevent bleeding and hemorrhagic complication along with resolution of cosmetic problems. In this case report we describe the clinical features, radiological findings and discuss the results of the surgical management of scalp vascular malformations.

CASE REPORT

A 27 year female complained of slowly progressive swelling localized over frontal 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 3 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 the frontal region. 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.

Radiological finding

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On CT angiography of face and brain there were multiple abnormally dilated and tortuous vessel were seen in subcutaneous fat of scalp in the frontal region in the midline. The arterial feeders arised from dilated right angular artery (branch of facial artery) and early filling draining veins were seen with preferential drainage into left external jugular vein through left angular vein. 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.

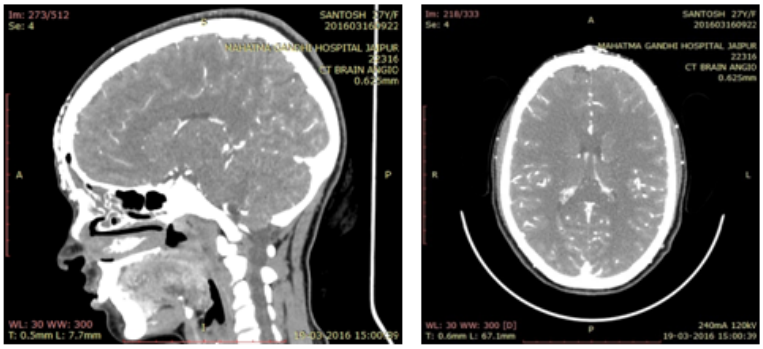

Figure 1: Cect saggital and axial scan showing multiple abnormally dilated and tortuous vessel were seen in subcutaneous fat of scalp in the frontal region in the midline

DISCUSSION

Aberrant persistence of primitive arteriovenous interconnections is known as AVM.AVM of scalp is vascular lesions with rare occurrences. Aneurysm cirsoide, 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 hemorrhage 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[4]. Pathological Autopsy data suggest an overall frequency of detection of AVMs to be 4.3% of the population[5]. Scalp arteriovenous fistulas are roughly evenly distributed among the frontal, temporal and parietal regions. 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 and 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 angular branches of facial artery. The etiology 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. 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 fistulae 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 hemangiomas. 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 hemangiomas, 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[6]. 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 hemorrhage. Others may present with severe symptoms such as scalp lesions. Hemorrhage is generally uncommon and may develop in the event of large vascular malformations. Recurrent hemorrhage 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 angio architecture 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 artifacts. 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 hemangiomas and cavernoma. 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 hemangioma. The case presented / in the presented case has features of Multiple tortuous and prominent; intensely enhancing vessels along frontal surface of the scalp was evident on Computerized tomography scan.

CONCLUSION

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 hemorrhage in young adults. Metastatic deposits from follicular carcinoma of the thyroid have to be ruled out prior to the diagnosis of cirsoid aneurysm.[7] 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[8]

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