

*Case Report***Extracranial Giant AVM of the Scalp with Plexiform Neurofibromatosis in a Juvenile Child: A Case Report**Vishwanath Sidram<sup>1</sup>, Chandrakumar P C<sup>1\*</sup><sup>1</sup>Assistant Professor, Dept of Neurosurgery, VIMS, Bellary, Karnataka

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*Received: 12/07/2012**Revised: 31/07/2012**Accepted: 04/08/2012***ABSTRACT**

We report a rare case of giant AVM with plexiform neurofibromatosis. A 7 year old girl presented with a 4 year history of large scalp mass on her right parieto-temporo-occipital scalp. She complained of headache due to its pulsatile nature. CT scan of head demonstrated a large extracranial tumour of soft tissue density. CT Angiography for the scalp vessels showed Arteriovenous Malformation (AVM) over right parieto-temporo-occipital region. Surgical excision of the tumour was performed. Histological diagnosis was AVM with plexiform neurofibromatosis. The rarity of scalp AVM in association with plexiform neurofibromatosis was highlighted.

**Key words:** Scalp AVM, Plexiform neurofibroma, Excision

**INTRODUCTION**

Scalp is the common site of soft tissue arteriovenous malformation. Their basic etiology is not clear. Aberrant differentiation of primitive vessel complex is considered as an etiological factor. Due to factors largely unknown, there is persistence of primitive arteriovenous interconnections which are normally replaced by an intervening capillary bed. [1]

Scalp AVM with plexiform neurofibromatosis is extremely rare, which may result in complex derangements of cerebral hemodynamics. They attain a certain size and may manifest as a visible pulsating swelling. [2] Their surgical treatment can be extremely difficult.

**CASE REPORT**

We report the case of seven years old female referred to our hospital with a four year history of right parieto-temporo-occipital scalp swelling. The swelling is manifested by headache and tinnitus in right ear. The swelling had been gradually increasing in size to attain a giant size and was pulsatile in nature. There was no previous history of trauma or head injury. There were also no visual disturbances or paresis or seizures.

On examination she was found to have a pulsatile soft tissue swelling over the right parieto-temporo-occipital region which was mobile over the underlying bone. Swelling extended through whole of the right parietal, temporal and occipital region

up to 2cm away from external occipital protuberance antero-posteriorly and was seen to extend towards the vertex vertically (Figure 01). A bruit was also demonstrated over the swelling. Examination of

cardiovascular system was essentially within normal limits. Fundoscopy showed no abnormality. Hematological and biochemical parameters were also normal. Chest X ray, ECG showed no abnormality.



Figure 01: Gross appearance of right parieto-temporo-occipital AVM

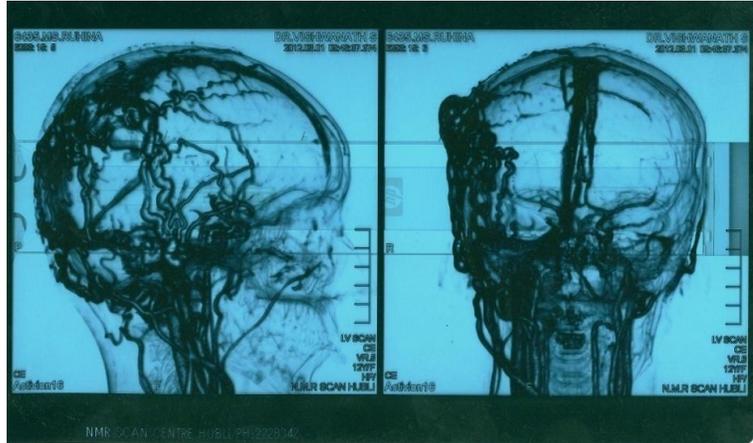


Figure 02: CT angiography showing AVM nidus with arterial feeders

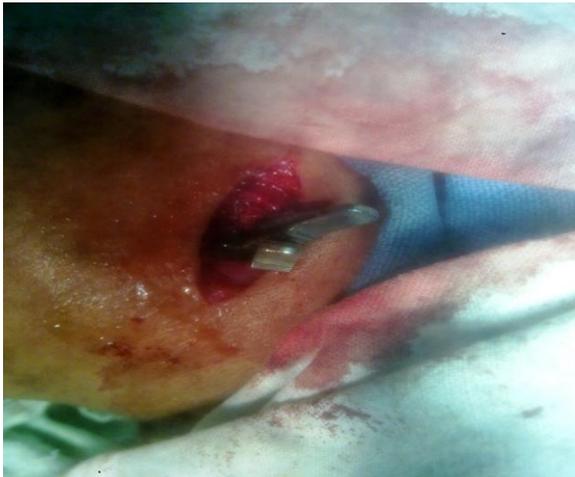


Figure 03: Bull dog clamp applied over right external carotid artery



Figure 04: Skin flap raised along with AVM nidus

A preliminary ultrasound and color doppler of scalp was done which suggested features of AVM over right parietal, temporal and occipital region with no intracranial extension with high velocity blood flow with arterial feeders noted from superficial temporal artery, posterior auricular artery and occipital artery.

CT angiography of neck vessel and circle of Willis showed evidence of abnormal tuft of contrast filled vessels involving the scalp on right side in temporo-parieto-occipital regions. The right facial artery, superficial temporal artery and occipital arteries were dilated suggestive of feeding arteries. The right retromandibular vein, posterior auricular vein and right

external jugular veins were dilated suggestive of draining veins (Figure 02). There was neither intracranial extension of the lesion nor any intracranial AVM noted.



Figure 05: Portion of excised AVM nidus

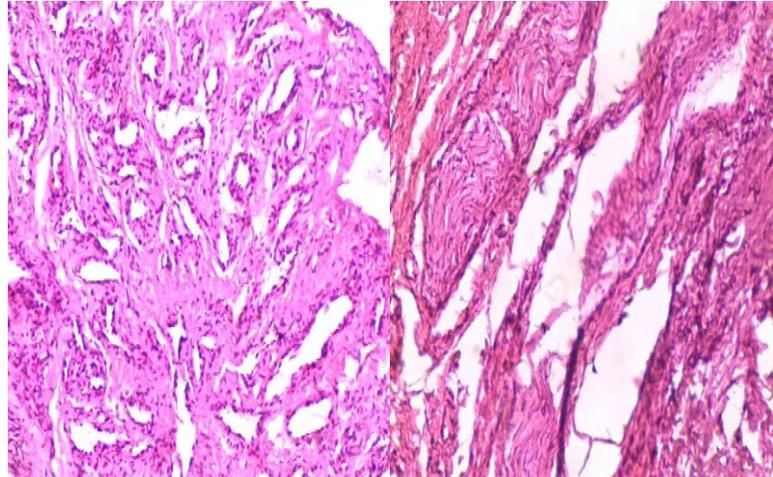


Figure 06 and 07: Histopathology revealing AV malformation with plexiform neurofibromatosis

Neck vessels and circle of Willis appeared normal in caliber. Brain parenchyma and ventricles did not reveal any significant abnormality.



Figure 08: Post operative CT angiography showing complete excision of AVM

Patient was advised surgery after explaining calculated risks. A bulldog clamp (Figure 03) was applied to right external carotid artery before performing excision of right AVM intoto. Right superficial temporal artery, posterior auricular artery and occipital arteries were identified and ligated. Excised soft tissue mass was sent to Department of pathology, Vijayanagara

Institute of Medical Sciences, Bellary, for histopathological examination(HPE) (Figure 04 and 05).

HPE revealed ambiguity of both arterial and venous blood vessels with abnormal muscularization of the arteries and arterioles with numerous cavernous like spaces suggesting prominent venous blood vessels. Interspersed within dilated vessels, features of extraneural plexiform neurofibromatosis like features with many prominent nerve bundles were seen. HPE confirmed diagnosis as AVM with plexiform neurofibromatosis (Figure 06 and 07). Post operatively the patient was followed for three months, where clinically the patient was relieved of symptoms which was evident by postoperative CT angiography, which showed complete excision of the lesion (Figure 08)

## DISCUSSION

An arteriovenous malformation of the scalp or face is an abnormal fistulous connection between the feeding arteries and

draining veins, without an intervening capillary bed in the subcutaneous layer. [3]

Various terminologies are in vogue; arteriovenous aneurysm, cirroid aneurysm, racemose aneurysm, aneurysm by anastomosis, plexiform angioma, aneurysmal varix, AV fistula, abnormal arteriovenous communication [4] and have been known for centuries. [5] They are normally supplied by the superficial temporal artery and occipital arteries. Occasionally they are supplied by dural arteries which penetrate the cranial vault. Venous blood drains mainly through scalp veins or via dural sinuses. [3]

Giant AVM with plexiform neurofibromatosis is a relatively rare entity with only few reported cases including the present case. [6,7,8] Soft tissue AVM occur more commonly in the scalp, Watson et al reported 50% of AVM in scalp region. [9]

AVM with plexiform neurofibroma is usually congenital in origin or may be followed by a trauma. However in our study either of origin could not be concluded. AVM with plexiform neurofibroma is characterized by very slow progression. [6] In our study the progression of AVM with plexiform neurofibroma is rapid, taking only four years to progress to a giant size.

In our study, the location of AVM with plexiform neurofibroma is parieto-temporo-occipital region in the scalp which is in accordance with Khodadad.G. [10] However, according to M.Tsutsumi et al, [7] majority of AVM with plexiform neurofibroma is occipital in location.

Rapid growth is due to intratumoral hemorrhage or malignancy, [6] neither of which was encountered in our case. Symptoms and signs of scalp AVM with plexiform neurofibroma range from a simple disfiguration to a life threatening hemorrhage. Tinnitus, occasional headaches and bruit are usually present, [4] which were also present in our case.

Fisher –Jeffes et al [11] in 24 patients with AVM of the scalp reported 38% of the lesions were preceded by trauma. Also no focal neurologic deficits or intracerebral involvement were noted in any of the patients. These clinical features were all seen in our patient who also had no history of trauma or neurological deficits.

In one study, congenital venous malformation with plexiform neurofibroma was associated with cranial defect, [12] however in our study, cranial defect was not present.

CT angiography is a very useful investigation in imaging neurovasculature. It delineates the lesion, excludes an intracranial component and demonstrates the related adjacent bony structures which may be important in surgical planning. [3,13]

The ideal treatment is complete excision of the AVM. It requires a complete knowledge of the feeding artery, the draining vein and the nidus of AVM.

It is best to identify the feeding arteries and ligate them first before exposing the AVM. Instead of ligating the external carotid artery, a temporary application of bulldog clamp is advisable. It is important to raise skin flap larger than the bulk of the AVM for easy dissection and to minimize bleeding. When possible, the AVM is removed in one piece, piece – meal excision increases the amount of bleeding and the operating time. [4] Total excision of the AVM lesion is essential for permanent cure.

## CONCLUSION

A rare case of extra cranial giant AVM of the scalp with plexiform neurofibromatosis in a juvenile child is presented. CT scan of head demonstrated soft tissue tumour over right parieto-occipital region. CT angiography revealed right parieto-temporal-occipital AVM with right facial artery, superficial temporal artery and occipital arteries as feeding

arteries & right retromandibular vein, posterior auricular and right external jugular veins as draining veins. HPE of the postoperative specimen revealed AVM with plexiform neurofibromatosis. Postoperative CT angiography showed complete excision of the lesion (Figure 08).

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