*Indian Journal of Medical Case Reports ISSN: 2319–3832*
*Online, International Journal, Available at http://www.cibtech.org/jcr.htm*
*2022 Vol.11, pp. 6-9/Pall et al.*
***Case Report (Open Access)***

# NEONATAL CONGENITAL SYMPTOMATIC SCALP ARTERIOVENOUS MALFORMATION-A CASE REPORT-IMAGING FEATURES AND FOLLOW UP AT TWO YEARS WITH REVIEW OF LITERATURE

**Mahander Pall, Gita Devi* and JP Singh**
*Neurosurgery Department and Radiodiagnosis & Imaging,*
*Sri Mata Vaishnu Devi Narayana Hospital Katra, Jammu and Kashmir, India*
**Author for Correspondence: gitabhagat@yahoo.com*

**ABSTRACT**
Congenital arteriovenous malformation (AVM) of the scalp is rare and is rarely asymptomatic at birth. These usually become symptomatic in adolescence and late adulthood or if complicated. We report a symptomatic neonatal scalp arteriovenous malformation who presented with gradually increasing scalp swelling associated with presence of thinning and discoloration on its surface suggestive of impending rupture. CTA showed complex vascular malformation with early draining veins draining into right scalp and facial veins and into right sigmoid sinus. Surgery was done with complete removal of the scalp AVM with no recurrence at two years of follow-up. Identification and treatment of the tumor in the early stage prevents complications with excellent outcomes.

***Abbreviation***
*AVM-Arteriovenous Malformation*
*CTA-Computed Tomograpgy Angiography*
*NCCT-Non Contrast Computed Tomography*
*CCA-Common Carotid Artery*
*ECA-External Carotid Artery*
*VRT-Volume Rendering Technique*

***Keywords:*** *Symptomatic Scalp AVM, Cirsoid Aneurysm, Congenital Arteriovenous Malformation, Scalp Swelling, Neonatal Scalp AVM, Arteriovenous Malformation*



## INRTODUCTION
Arteriovenous malformation (AVM) results from disturbances in angiogenesis that leads to abnormal shunting of blood between arteries and veins. These are high flow vascular malformations and can arise from any part of the body with wide range of presentations from an asymptomatic birthmark to life threatening impairment on vital organs (Lam *et al*., 2017).
Congenital AVM of the scalp are rare and commonly asymptomatic at birth (Feletti *et al*., 2018). They present as purple or red birthmarks and often mistaken as hemangiomas (Hussain *et al.,* 2017 and Komatsu *et al.,* 1989). Patients sometimes present with disfiguration of the scalp as the only sign (Khodadad, 1971). They usually become symptomatic in adolescence and late adulthood or after trauma with hemorrhage, infection or local tissue destruction (Jennifer *et al.,* 2005). Systemic manifestations like congestive cardiac failure and death can also occur. Only three case reports of symptomatic neonatal scalp AVMs exists in the literature (Hussain *et al.,* 2017, Vasconez, 1973 and Yoshie *et al.,* 1997). We will

report fourth case to the best of our knowledge. Treatment options are multiple. The best way of treatment is total removal of the tissue of AVM as early as possible (Schultz *et al.*, 1980).

**CASE**

The case is about a neonate patient 7 days old with a swelling in right scalp since birth (Fig 1A). It gradually increased in size since birth with presence of focal thinning and surface changes suggestive of impending rupture. It was soft pulsatile swelling. NCCT showed large hypodense scalp lesion with few small hyperdensities consistent with calcifications (Fig 1B). CTA of neck and cranial vessels showed presence of a large hypervascular scalp lesion measuring 6x5x2.5cm showing early draining veins draining predominantly into the right retromandibular vein and then into the right common facial vein and right IJV. The right retromandibular and common facial veins are dilated and tortuous (Fig 1D, F, G). Few thin early draining veins are also communicating with right sigmoid sinus with the result opacification of right transverse sinus and sigmoid sinus was present however no bony defects or extension of the lesion intracranially is noted (Fig. 1D, E, F,G). Other facial veins and left neck veins

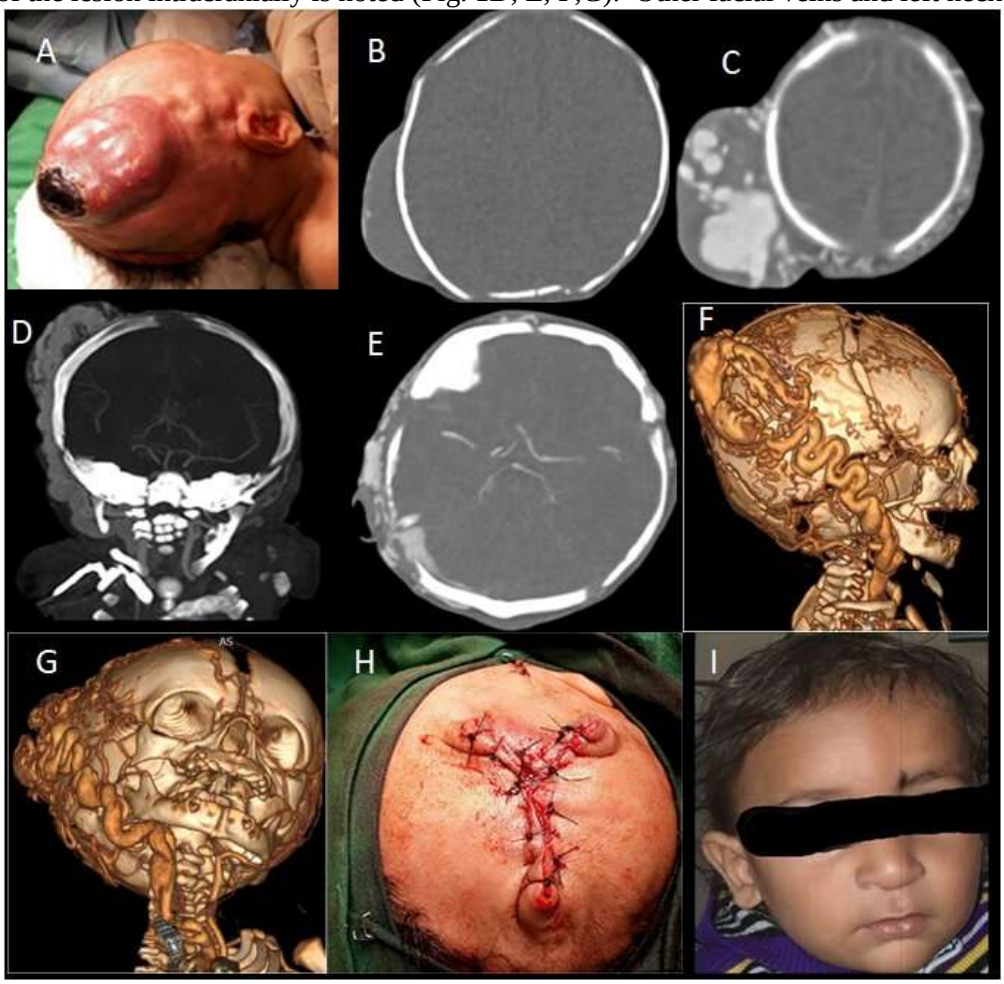


**Figure 1: (A) Showing Neonate with ~6x7cm right scalp swelling with focal discoloration and skin thinning. (B) NCCT showing few hyperdense foci suggestive of calcifications in the periphery of a large hypodense scalp lesion. (C, D) CTA showing scalp lesion with dilated and tortuous vascular channels, dilated and tortuous right retromandibular and common facial vein, prominent right CCA and superficial temporal artery with (E) early draining veins on the arterial phase causing opacification of right sigmoid sinus and right transverse sinus and (F and G) VRT images showing the same findings. (H) Post op picture showing single layer-interrupted sutures. (I) Follow up at two years of age. No recurrence**.

also showed early opacification. The right CCA and ECA are prominent. The right superficial temporal artery was also prominent. These features suggest of arteriovenous fistula in scalp. Immediate surgery was planned. A vertical incision was made in front of tragus up to the lower part of swelling. Superficial temporal artery was separated and ligated. Two vertical curvilinear incisions were made over the swelling joining at upper and lower ends. Frontal part of the scalp was separated from swelling coagulating and ligating frontal artery branches supplying the AVM. Posterior part of scalp was separated from swelling coagulating and ligating occipital branches supplying AVM. The lesion removed in toto. Veins were also ligated and hemostasis achieved. The wound closed in single layer with interrupted sutures (Fig 1H). Post-surgical course was uneventful and wound was healthy and sutures removed on 12$^{th}$ day. The patient was healthy at the time of discharge. After two years of follow up there was no evidence of recurrence (Fig 1I).

## DISCUSSION

Scalp arteriovenous malformations are rare lesions and symptomatic neonatal scalp AVM are extremely rare. These can be congenital, traumatic or post infectious (Hussain *et al.,* 2017, Li *et al.*,2018 and Heifeman *et al.,* 2019). These are also called cirsoid aneurysm, serpentine aneurysm, recemose aneurysm, arteriovenous fistula or plexiform aneurysm (Ahmad *et al.,* 2017). Khodadad (1971) has mentioned four major etiologies of scalp arteriovenous fistulas, Congenital, traumatic, infections and inflammation and familial. Familial AVM is extremely rare. Khodadad (1973) has mentioned its observation in a Persian family. Congenital AVM can develop at birthmark site. It can also be present without obvious birthmark and sometimes only birthmark may be present without manifestations. Its development can be due to the persistence of primitive arteriovenous communications or from vascular hemartomas or fistulous formation at the site of crossing of arteries and veins during embryonic period (Khodadad, 1973).

Embryological pattern shows that origins of scalp, dural and cerebral congenital AVMs are different. Communicating intra and extracranial aneurysms are due to secondary communications.

Treatment options for scalp AVM are multiple. These include endovascular, ligation of the feeding vessels, electrothrombosis, use of sclerosing agents and surgical. Surgery is most commonly used and successful method. Endovascular treatment can be used as a definitive method of treatment or as an adjunct to surgical treatment (Shenoy *et al.,* 2004). Recurrence is more common after endovascular treatment (Reddy *et al.,* 2015).

## CONCLUSION

Congenital arteriovenous malformation of the scalp is rare and symptomatic cases are even extremely rare. Identification of early stage to avoid drastic complications like hemorrhage is important. Treatment of the tumor in early stage prevents complications with excellent outcomes. Surgical resection is most common and successful method of treatment.

## REFERENCES

**Ahmad F, Singh DK, Prajapati HP and Chhabra A(2017).** Cirsoid Aneurysm of the scalp: a case report. *Romanian Neurosurgery,* **31(3)** 426-430.

**Feletti A, Dimitriadis S, Vallone S and Pavesi G (2018).** Congenital Arteriovenous Malformation of the Scalp Involving the Orbit. *Journal of Neurological Surgery Part A: Central European Neurosurgery,* **79(6)** 541-9.

**Heifeman DM, Syed HR , Li D, Rothstein BD, Shaibani A, Tomita T(2019).** Resection of an Embolized Cirsoid Aneurysm with intracranial venous drainage: 2-Dimensional operative video. *Operative Neurosurgery*, **16(3)** E94-E94.

**Hussain AS, Ahmed SA, Ali SR and Ahmed K (2017**). Congenital neonatal scalp arteriovenous malformation: a very rare entity. *BMJ Case Report. Available at:* doi:10.1136/bcr-2016-218756.

**Jennifer JM, John BM (2005).** Current management of hemangiomas and vascular malformation. *Clinical Plastic Surgery,* **32** 99-116.

**Khodadad G(1973).** Arteriovenous malformation of the scalp. *Annals of Surgery,* **177** 79-85.
**Khodadad G (1971).** Familial cirsoid aneurysm of the scalp. *Neurologia Medico-Chirurgica,* **34** 664-667.
**Komatsu Y, Narsushima K, Kobayashi E, Nose T and Maki Y (1989**). Congenital arteriovenous malformation of the scalp—case report. *Neurologia Medico-Chirurgica,* **29(3)** 230-4.
**Lam K, Pillai A and Reddick M (2017).** Peripheral arteriovenous malformations: Classification and endovascular treatment. *Applied Radiology,* **46** 15-21.
**Li D, Heiferman DM, Rothstein BD, Syed HR, Shaibani A, Tomita T (2018).** Scalp Arteriovenous malformation (cirsoid aneurysm) in adolescence: Report of 2 cases and review of the literature. *World Neurosurgery,* **116** e1042-e1046.
**Reddy V R, Harishal P. N, Hegde K V, Agrawall A, Kandukuri K (2015).** Cirsoid aneurysm of scalp: Value of multidetector CT scan in pre-operative evaluation. *Medical Journal of Dr. D. Y. Patil University,* **8** 804-806.
**Schultz RC, Hermosillo CX (1980**). Congenital arteriovenous malformations of the face and scalp. *Plastic Reconstruction Surgery,* **65(4**) 496-501.
**Shenoy SN, Raja A(2004).** Scalp arteriovenous malformations. *Neurology India,* **52** 478-81.
**Vasconez LO (1973).** Congenital defect of the skull and scalp due to an arteriovenous malformation. Case report. *Plastic Reconstructing Surgery,* **51** 692-5.
**Yoshie HA, Tamaki N, Kuroda R (1997).** A Neonatal Scalp Arteriovenous Malformation. *Neurosonology,* **10** 202-5.