Case Report

Giant arteriovenous malformation of scalp - A rare case report

PV Bhagwat, V Kuntoji, C Kudligi, GC Patil*, PV Nayak*

Department of Dermatology and Venereology, Karnataka Institute of Medical Sciences, Hubli, Karnataka

* Department of Radiology, Karnataka Institute of Medical Sciences, Hubli, Karnataka

Abstract Arteriovenous malformations (AVMs) are congenital lesions which result from abnormal communication between arterial and venous vessels directly without an intervening capillary bed. They may present as pulsatile subcutaneous mass which may bleed or ulcerate. We present a 28-year-old male with an unusually large AVM on scalp.

Key words

Arteriovenous malformation, fast-flow.

Introduction

Arteriovenous malformations (AVMs) are congenital, fast-flow vascular lesions composed of malformed arterial and venous vessels connected directly to one another without an intervening capillary bed.¹ They are more common in brain involving or supplied by intracranial vasculature than those formed by branches of external carotid arteries.² AVM of scalp is a rare lesion which has its abnormal arteriovenous communication within the subcutaneous fatty layer of scalp with the feeding arteries derived from vessels supplying the scalp. They present as a subcutaneous scalp lump or a large, pulsatile mass with a propensity for skin erosion and massive haemorrhage.³ We report a case of an asymptomatic giant arteriovenous malformation of scalp involving extracranial circulation.

Address for correspondence

Dr. PV Bhagwat Associate Professor, Dept of Skin & STD, Karnataka Institute of Medical Sciences, Hubli, Karnataka email: sharadapbhagwat@yahoo.com

Case Report

A 28-year-old male presented with gradually progressive asymptomatic swelling over scalp and forehead since birth. It was not associated with pain or bleeding. However, there was history of recent trauma (punch) in the right periorbital region 3 days prior to his presentation following which he had developed swelling after 1 day which was associated with mild pain. There was no history of headache and convulsions. On examination, soft compressible swelling of size 12x10cms was present over right side of forehead extending to the vertex and in right periorbital region (Figure 1). Mild tenderness was present in right periorbital region. Bruit was heard on auscultation. Systemic examination was unremarkable. Routine blood investigations were normal. Skull X-ray revealed prominent vascular markings of middle meningeal artery in right frontoparietal region. Ultrasonography showed multiple dilated tortuous hypoechoic linear lesions in the subcutaneous plane of right periorbital, maxillary, temporal and parietal regions. Colour Doppler revealed monomorphic blood flow suggestive of low resistance flow [Figure 2(a) 2(b) 2(c)]. Plain CT showed soft tissue lesion in

right periorbital region. There was also a bony erosion and hematoma in right temporal region. CT contrast showed few enlarged and tortuous blood vessels in frontoparietal region. CT Bone window showed sutural diastasis [Figure 3(a) 3(b)]. MRI brain showed tortuous multiple flow voids in temporal, frontal and parietal regions in subcutaneous plane. MRI neck angiography demonstrated the feeding arteries from superficial temporal artery, internal maxillary and middle meningeal arteries and also dilated tortuous veins [Figure 4(a) 4(b)].



Figure 1 Multiple nodules with pus discharging sinuses present over upper left chest and left shoulder region.



Figure 2a Colour Doppler showing monomorphic blood flow suggestive of low resistance flow.



Figure 2b X-Ray of the skull showing prominent vascular marking (middle meningeal artery) in the frontoparietal



Figure 2c CT Bone window showing sutural diastasis



Figure 3a Plane CT showing soft tissue lesion in right periorbital region.



Figure 3b Contrast enhanced CT showing tortuous dilated blood vessels in frontoparietal region.



Figure 4a MRI showing tortuous multiple flow voids in temporal, frontal region in s.c. plane in T1 sequence

Table 1	I	Schobinger	staging	for	AVM
I GOIC I		beneounger	beaging	101	

6	00
Stage	Clinical Symptoms
I (Quiescence)	Skin warmth, discolouration
II (Expansion)	Enlargement, pulsation, bruit
III (Destruction)	Pain, ulceration, bleeding
IV(Decompensation)	Cardiac failure due to volume
-	overload

Discussion

Vascular malformations are developmental abnormalities of the vascular system that occur during fourth and tenth weeks of intrauterine life. They are divided into slow-flow (capillary, lymphatic, venous and combined) and fast-flow (arterial, arteriovenous, and combined). They can be well-demarcated and localised or they can be the stigmata of deep lesions.⁴ 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.⁵

AVM of the scalp is a rare lesion when compared with other subcutaneous or cervicofacial vascular anomalies such as the hemangioma or venous malformations.⁶ The feeder vessels of scalp AVM mainly arise from subcutaneous tissue of the scalp and the sources of the feeder arteries most frequently include the external carotid, occipital and supraorbital



Figure 4b MRI neck angiogram showing multiple arterial feeders and draining veins.

arteries. It usually involves frontal, temporal and parietal regions.⁷ It may be present at birth which will be usually asymptomatic until adulthood or it may be posttraumatic which is generally seen in patients over 30 years of age.8 While it may be associated with physical disfigurement or functional disturbances like throbbing sensations, tinnitus and headache in early stages, persistent pain, scalp necrosis, bone erosion. life threatening hemorrhage, cardiovascular failure, cerebral steal phenomenon and seizures are seen in late destructive and decompensated lesions.^{5,9} Our patient had none of these symptoms except for mild pain in the periorbital region following recent trauma. As per Schobinger staging for AVM^{10} , our patient was in stage 2 (**Table 1**).

Hemangioma or cavernoma often cause confusion in diagnosis especially in early stages, which unlike AVM are well-demarcated and do not have arteriovenous shunts.¹¹ AVMs show flow void signs on MRI due to the rapid flow in the lesions.8 The diagnosis of AVM in the present case was made by contrast enhanced CT and MRI brain and the feeding vessels viz., superficial temporal artery, internal maxillary middle meningeal and arteries were demonstrated by MRI neck angiography. The management AVM involves of a

multidisciplinary approach and the treatment options vary from conservative follow-up to aggressive treatment like embolization and/or surgical resection. The case was referred to neurosurgery department and is currently being followed up for interventional procedures. Since dermatologists rarely encounter such a giant asymptomatic arteriovenous malformation of scalp, we considered it apt to report the case to sensitise regarding this complex entity and stresses the importance of various radiological investigations in deciding further management.

References

- 1. Dompmartin A, Revencu N, Boon LM, Vikkula M. Disorders affecting cutaneous vasculature. In: Griffiths CEM, Barker J, Bleiker T, Chalmers R, Creamer D. editors. *Rook's Textbook of Dermatology. 9th edition.* Oxford: Wiley Blackwell. P.73.7.
- Weinzweig N, Chin G, Polley J, Chabrel F, Shownkeen H, Debrun G. Arteriovenous malformation of the forehead, anterior scalp and nasal dorsum. *Plast Reconstr Surg*. 2000;**105**:2433-9.
- Shenoy SN, Raja A. Scalp arteriovenous malformations. *Neurology India*. 2004;**52**:478-81.
- 4. Boon LM, Vikkula M. Vascular malformations. Goldsmith LA, Katz SI,

Gilchrest BA, Paller AS, Leffell DJ, Wolff K, editors. *Fitzpatrick's Dermatology in General Medicine*. *8th edition*. Phiadelphia: McGraw-Hill; 2008. p2077.

- 5. Khodadad G. Arteriovenous malformations of the scalp. *Ann Surg.* 1973;**177**:79-85.
- Mohamed WNZW, Abdullah NNLA, Muda AS. Scalp arteriovenous malformation: A case report. *Malaysian J Med Sci*. 2008;15(3):55-7.
- 7. Hasturk AE, Erten F, Ayuta T. Giant nontraumatic arteriovenous malformation of the scalp. *Asian J Neurosurg*. 2012;**7**:39-41.
- 8. Chowdhury FH, Haque MR, Kawsar KA, Sarker MH, Haque AFMM. Surgical management of scalp arterio-venous malformation and scalp venous malformation: An experience of eleven cases. *Indian J Plast Surg.* 2013;**46**:98-107.
- Schobinger MP, Hansen M, Pribaz JJ, Mulliken JB. Arteriovenous malformations of the head and neck: Natural history and management. *Plast Reconstr Surg.* 1998;102:643-54.
- Pekkola J, Lappalainen K, Vuola P, Klockars T, Salminen P, Pitkaranta A. Head and neck arteriovenous malformations: Results of ethanol sclerotherapy. *Am J Neuroradiol.* 2013;**34**:198-204.
- 11. Hasturk AE, Erten F, Ayata T. Giant nontraumatic arteriovenous malformation of the scalp. Asian J Neurosurgery. 2012; 7(1):39-41.