CASE REPORT

Congenital neonatal scalp arteriovenous malformation: a very rare entity

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SUMMARY

Congenital arteriovenous malformations (AVMs) of scalp are rare congenital vascular malformations. They are usually not symptomatic at birth and are often misdiagnosed as haemangiomas. To date, only two cases of symptomatic neonatal scalp AVM have been reported in literature. Pathophysiology of congenital AVM is not completely understood but genetic and acquired causes are implicated. Diagnosis and management are often difficult and require multidisciplinary approach. We report a rare case of symptomatic congenital scalp AVM in a 10-day-old neonate who was successfully managed at our unit.

Figure 1 Lesion at presentation to our hospital.

BACKGROUND

Congenital arteriovenous malformations (AVMs) are high-flow vascular lesions that arise from persistent communication between blood vessels due to the failure of the embryonic vasculature to differentiate into arteries and veins.^{1,2} They appear as red or purple birthmarks and are often mistaken as capillary haemangiomas at births.^{1,2} AVMs are rarely symptomatic in the neonatal period and usually clinically manifest in adolescence and late adulthood or after trauma with haemorrhage, infection or local tissue destruction.³ They can also cause systemic manifestations like congestive cardiac failure and death. Several adult^{4–6} and few paediatric cases⁷ of scalp AVMs have been reported. Only two case reports of symptomatic neonatal scalp AVMs^{8,9} exist in the literature. This case report of ours highlights a 10-day-old neonate with symptomatic scalp AVM.

CASE PRESENTATION

A 10-day-old boy neonate presented with an episode of profuse bleeding from a scalp swelling on the left frontotemporal region. Mother received regular antenatal care at a local hospital and the baby was born via elective C-section.

Postnatal physical examination of the neonate revealed a small (2 cm) lesion over left frontotemporal area with denuded skin. Bluish in colour, the lesion had a flat surface with raised margins. The preliminary diagnosis on ultrasound was haemangioma for which oral propranolol was initiated at discharge. Baby remained stable at home but gradually developed swelling around the lesion. At 10th day of life, baby developed spontaneous bleeding from the scalp lesion for which he was brought to the emergency department

at our hospital (figure 1). On examination, baby was haemodynamically stable. A soft, warm and pulsatile swelling on the left temporal region was noticed with a bruit on auscultation. There was no active bleeding from the lesion. Rest of the systemic examination and initial evaluation done in the emergency department, including coagulation profile, was unremarkable.

A day after his admission, there was active, pulsatile bleeding from the lesion for which packed red blood cells were transfused and bleeding was controlled by manual compression. A CT cerebral angiogram (CTA) was performed which showed abnormal communication between left superficial temporal artery and tributaries of left internal jugular vein and supraorbital vein representing scalp AVM (figure 2A–C). After the episode of profuse bleeding and CTA showing AVM, digital subtraction angiography (DSA) followed by successful polyvinyl alcohol chemo embolisation and platinum coiling of the AVM was performed (figure 3A–D). Post-embolisation and coiling, contrast run showed minimisation of flow in AVM. Surgical excision of the AVM, without complications, was performed 48 hours after the embolisation. Ultrasound of liver and kidneys along with echocardiography was done to rule out any additional AVMs and its complications. Our patient did not develop any complication and was discharged in a stable condition.

Histopathology of the specimen showed a mixture of abnormally dilated small-sized and large-sized capillaries along with thick walled veins and a few arteries, an embolised vein along with a few arteries and multiple small and large calibre capillaries (figure 4A,B). These findings were consistent with AVMs.

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Figure 2 CT angiogram showing scalp arteriovenous malformation.

DIFFERENTIAL DIAGNOSIS

Haemangioma.

TREATMENT

Embolisation followed by complete excision.

OUTCOME AND FOLLOW-UP

Baby successfully managed and discharged home.

DISCUSSION

AVMs are vascular malformations leading to shunting of blood from arterial to the venous system.³ They are usually found

Figure 3 Digital subtraction angiography showing successful embolisation of scalp arteriovenous malformation.

Figure 4 (A) Histopathology slide showing embolised vein along with a few arteries and multiple small and large caliber capillaries. (B) Histopathology slide showing a thrombosed vessel with admixed multiple-sized capillaries filled with blood.

Learning points

- Symptomatic congenital scalp arteriovenous malformations (AVMs) are rare in neonates and can be potentially life threatening if not identified and treated promptly.
- CT angiography/digital subtraction angiography are gold standard for the diagnosis
- Management requires multidisciplinary approach.
- Embolisation followed by complete excision of AVM is safe and effective in the neonatal period.

intracranially but extracranial sites are also not rare. Scalp AVMs may be present at birth, but clinically manifests itself in late childhood, adolescence or adulthood.¹⁰ Our patient presented at an unusually young age with spontaneous bleeding from the lesion.

Though the aetiology and pathogenesis of AVMs still remain unclear, inborn errors in embryological development, disruption among genetic, hormonal, biochemical factors and finally a positive vascular endothelial growth factor (VEGF) in histopathological milieu has been associated with its pathogenesis.¹⁰

Cerebral angiography is the gold standard for the diagnosis of AVMs. This can be performed using any cross sectional modality like CT or magnetic resonance angiography (MRA). Although CT and MRA both are complimentary to each other, CTA plays a vital role in the diagnosis of AVM as well as evaluation of surrounding tissue, bone, the feeding arteries and venous drainage especially in the neonatal age group. The management of AVMs requires multidisciplinary approach. Several treatment options have been described to treat these lesions including embolisation, surgical excision or combination of both.¹¹ Preoperative embolisation to control acute bleeding followed by excision has shown excellent results.^{12 13} Managed similarly, we controlled the bleeding by opting for embolisation followed by complete surgical resection of the lesion as a more controlled procedure within 48 hours of the embolisation.

CONCLUSION

Symptomatic neonatal scalp AVMs are extremely rare and possible cause of life threatening haemorrhage in neonate. Early identification and prompt multidisciplinary management lead to excellent outcomes.

Contributors This study has been performed by all the authors contributing equally in the conception, data collection and manuscript writing. SAA wrote the initial draft which was then edited by ASH who is the corresponding author. ASH, SRA and KA reviewed and finalised the manuscript.

Competing interests None declared.

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REFERENCES

- 1 Ennies JT, Bateson EM, Moule NJ. Uncommon arterio-venous fistulae. *Clin Radiol* 1972;23:392–8.
- 2 Schultz RC, Hermosillo CX. Congenital arteriovenous malformation of the face and scalp. *Plast Reconstr Surg* 1980;65:496–501.
- 3 Jennifer JM, John BM. Current management of hemangiomas and vascular malformation. *Clin Plasti Surg* 2005;32:99–116.
- 4 Hasturk AE, Erten F, Ayata T. Giant non-traumatic arteriovenous malformation of the scalp. *Asian J Neurosurg* 2012;7:39.
- 5 Mohamed WN, Abdullah NN, Muda AS. Scalp arteriovenous malformation: a case report. *Malays J Med Sci* 2008;15:55.
- 6 Kim ET, Lee Y-J, Park DW, *et al.* Arteriovenous fistula at Scalp: rapid Progression after embolization of Contralateral Facial Arteriovenous Malformation. *Neurointervention* 2010;5:36–9.
- 7 Gupta R, Kayal A. Scalp arteriovenous malformations in young. *J Pediatr Neurosci* 2014;9:263.
- 8 Vasconez LO. Congenital defect of the skull and scalp due to an arteriovenous malformation. Case report. *Plast Reconstr Surg* 1973;51:692–5.
- 9 Yoshie HA, Tamaki N, Kurata H, *et al.* KURODA R. A Neonatal Scalp Arteriovenous Malformation. *Neurosurgery* 1997;10:202–5.
- 10 Hage ZA, Few JW, Surdell DL, *et al.* Modern endovascular and aesthetic surgery techniques to treat arteriovenous malformations of the scalp: case illustration. *Surg Neurol* 2008;70:198–203.
- 11 Tao Q, Lv B, Bhatia KS, *et al.* Three-dimensional CT angiography for the diagnosis and assessment of arteriovenous malformations in the oral and maxillofacial region. *J Craniomaxillofac Surg* 2010;38:32–7.
- 12 Nagasaka S, Fukushima T, Goto K, *et al.* Treatment of scalp arteriovenous malformation. *Neurosurgery* 1996;38:671–7.
- 13 Kumar R, Sharma G, Sharma BS. Management of scalp arterio-venous malformation: case series and review of literature. *Br J Neurosurg* 2012;26:371–7.

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