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Successful En Bloc Resection with Acceptable Cosmesis of a Right Recurrent Giant Scalp-Orbital AVM Causing Severe Disfigurement: A Case Report

Anjan Singh Karki[1,*], Dipendra Kumar Shrestha[1], Gopal Sedain[1] and Sushil Krishna Shilpakar[1]

[1]Department of Neurosurgery, Tribhuvan University Teaching Hospital, Maharajgunj, Kathmandu

***Corresponding author:** Karki AS, Maharajgunj Medical Campus, Institute of Medicine, PO Box 3579, Maharajgunj, Kathmandu, Nepal, E-mail: dranjansinghkarki@gmail.com

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Abstract

Scalp Arterio Venous Malformation (AVM) is a rare congenital entity, accounting for only 8.1% of all AVM cases. These lesions are made up of an abnormal fistulous tangle of tortuous dysmorphic network of vessels "vascular nidus" directly connecting between the feeding arteries and draining veins, without capillary connection, located within the incision subcutaneous layer. With time, these congenital lesions may evolve and enlarge and clinically manifest with variable features. The only effective method of preventing evolution of these malformations is to exclude the lesion completely from the circulation. Generally, scalp AVM presents as a small pulsatile mass; but with time, if untreated, it will progressively enlarge and spread to the entire scalp, giving a grotesque appearance and involves the craniofacial structures and skull bone and even extends intracranially, leading to seizures and hemorrhages. Involvement of the orbit and face may cause severe facial disfigurement, proptosis, visual obstruction and even facial palsy. These lesions may be complicated by ulceration, infection and profuse bleeding. There are varieties of neuro-imaging modalities which help in characterizing the structural complexity, location and the extent of the lesion and assist in appropriate preoperative surgical planning. Among them, Three-Dimensional CT Angiography (3D-CTA) is regarded as the cornerstone for investigation. Management of these complex lesions may require multidisciplinary approach with preoperative embolization and surgical excision. As it is often difficult to cure scalp AVM with embolization alone, surgical resection is usually required. Generally, surgical excision is said to be curative; and recurrence is uncommon after complete resection. We present a rare case of a recurrent congenital scalp-orbital AVM causing proptosis, visual hindrance and severe facial disfigurement which was successfully excised with acceptable cosmesis along with a brief literature review.

Keywords: Disfigurement; En-blocresection; Proptosis; Scalp-orbital arteriovenous malformation

Case History

A 40-year-old housewife, from Western Nepal, presented to our Neurosurgical Out-patient Clinic with a large pulsatile grotesque mass over the right fronto-orbital region. She was very depressed; and she had visited numerous hospitals to have this lesion removed so that she could see with her right eye and get rid of the noticeable facial disfigurement. In past, she was born with a swelling in the right frontotemporal region, which over time increased in size progressively. The pulsatile lesion was excised; and the scalp defect was reconstructed with full-thickness skin graft in April of 1992 (Baisakh 2049 BS) in Palpa Mission Hospital, Nepal. Later, a small swelling recurred over the right supraorbital region, located anterior to the previous surgical site, which gradually increased in size over the last two years. This led to proptosis of the right eye as well as visual obstruction finally leading to severe facial disfigurement. However, there was no pain, discharge, ulceration or bleeding from the lesion.

On physical examination, she was fairly-built with darker skin complex. Vital signs were stable. Right frontotemporal region was bald with glistening skin due to previous skin graft. An oblong healthy incisional scar was noted surrounding the 10 × 6 cm^2 grafted hairless area. There was approximately 7 × 5 × 5 cm^3 soft to firm spherical non-tender pulsatile mass with engorged superficial vessels on overlying the coarse hyperpigmented skin arising from the right orbital region (Figure 1).

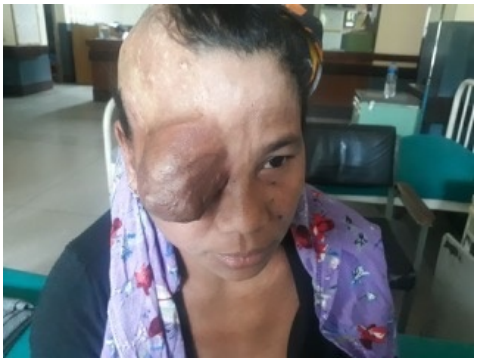

Figure 1: Preoperative picture showing the giant high-flow scalp-orbital AVM causing visual obstruction of the right eye and severe facial disfigurement. Note the right frontotemporal bald area due to previous skin graft and incisional scar.

On auscultation, bruit was heard clearly over the lesion. The patient was unable to open her right eye due to the large mass; and the right eyeball was displaced downward. However, the horizontal movement of the right eye was intact. Both pupils were round, regular and reactive to light. Visual acuity on both eyes was 6/9. Examination of all other systems was normal.

On neuroradiological evaluation, CT angiography of head demonstrated an extensive vascular nidus within the swollen soft tissue of right periorbital and inferior frontal region. The lesion was supplied by numerous feeding arteries, namely frontal, supratrochlear, supraorbital branches of the right superficial temporal artery, angular branches of both facial arteries; and the major feeder was from the intracranial circulation *via* right ophthalmic artery (Figure 2).

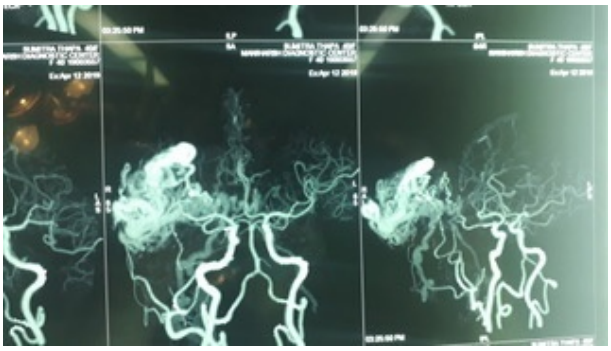

Figure 2: CT angiography demonstrating large vascular nidus with multiple arteries feeders, predominantly supplied by right ophthalmic artery (black arrow).

Three-dimensional images (3D-CTA) revealed large highly vascular nidus involving predominantly the right forehead on supraorbital region and extending into the orbital region without involvement of skull or intracranial structures (Figure 3).

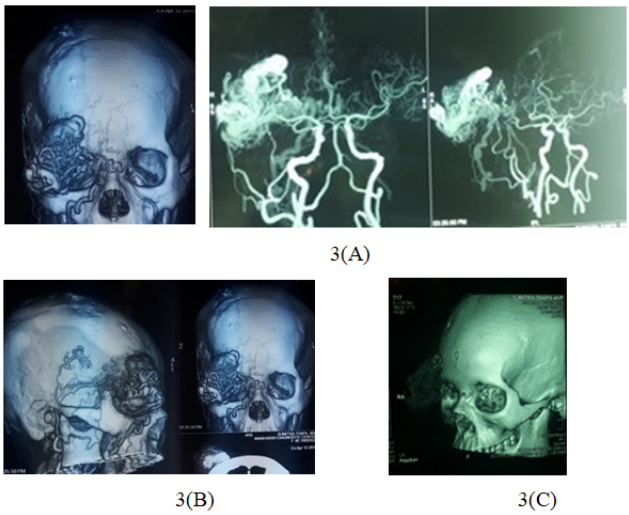

Figure 3: Three-dimensional CT angiography (3D-CTA) showing the giant right scalp-orbital AVM in relation to the skull and orbit (A) Anterior, (B) Lateral and (C) Oblique views. Note the extension of lesion into the orbit without involvement skull bone and without intracranial extension.

The diagnosis was consistent with recurrent high-flow scalp-orbital AVM. She underwent surgical excision of AVM and superior eyelid repair under general anesthesia in May of 2019. Before making the incision, temporary occlusion of major dilated feeding arteries was performed by ligating them percutaneously with 3-0 silk suture, approximately 2 cm proximal to the planned excisional margin. Notably dilated anterior (frontal), supratrochlear and supraorbital branches of the right superficial temporal artery lying superiorly and right and left angular arteries located inferior-medially, supplying the pulsatile mass were ligated to reduce the blood flow to the lesion and decrease blood loss during dissection and excision of the vascular mass. A curvilinear incision was made along middle aspect of the globular swelling inferior to the presumed right eyebrow. Meticulous dissection and thorough hemostasis were maintained using monopolar and bipolar coagulation. The dissection was initiated from the normal-looking tissue over the superior aspect of vascular nidus. Using monopolarBovie cautery, the pericranium and frontalis muscle on the superior margin of the lesion were delineated and dissected off the frontal bone. The bony surface was thoroughly waxed and electrocauterized to stop the oozing blood. The large dilated vessels were dissected and individually ligated with 3-0 silk sutures. The lesion was circumferentially dissected and excess skin was also excised. Utmost care was taken to avoid injury to the lacrimal gland and levator and orbicularis muscles. Laterally, the zygomatic branch and the main stump and non-dilated normal branches of superficial temporal artery were carefully preserved so as not to jeopardize vascularity of the previously grafted scalp.

After circumferential excision, the lesion was still attached to the vascular pedicle involving the orbit. Within the pedicle, the distal part of right superior ophthalmic artery was identified and doubly-ligated separately. Finally, the lesion with the entire vascular nidus was completely removed *via* en bloc resection. Hemostasis was strictly secured using monopolar and bipolar cauterization. The excess skin over the superior eyelid was removed; and incision was reconstructed for acceptable aesthetic and cosmesis. The incision was then closed in layers: muscle-fascia with 3-0 vicryl interruptedly and skin with 4-0 prolene continuously. No drain was placed (Figure 4).

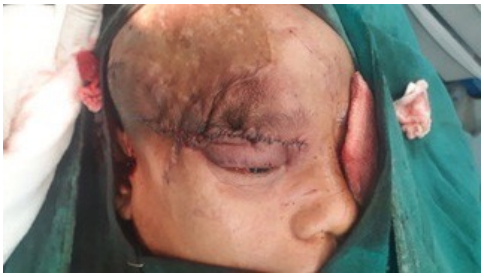

Figure 4: Postoperative picture showing curvilinear incision below the right eyebrow and acceptable cosmesis after excision of right scalp-orbital AVM.

Macroscopic examination demonstrated a single piece of grey-brown skin tissue grey, measuring 7.5 × 5 cm. The cut section showed multiple dilated vessels containing clot and pinpoint hemorrhagic areas. Microscopic view revealed multiple variable-sized interlacing vascular spaces in fat and soft tissues. Some of the vessels were lined by endothelial cells. However, other vascular spaces showed elastic and muscular layers as well as arteries and veins. Nerve bundles were also intermixed. The findings were consistent with AVM of supraorbital region.

Though there was mild periorbital swelling post-surgery which eventually subsided, the postoperative course was largely uneventful. The patient was discharged on Post-Operative Day four (POD#4). Sutures were removed on POD#10 in the Out-patient Clinic. On one-month follow up, the patient was high-spirited. She was finally able to see well, look up and close her right eye after two long agonizing years. The incision healed with secondary intention with patchy area of dark discolored indurated skin on the upper lateral aspect of the incision (Figure 5).

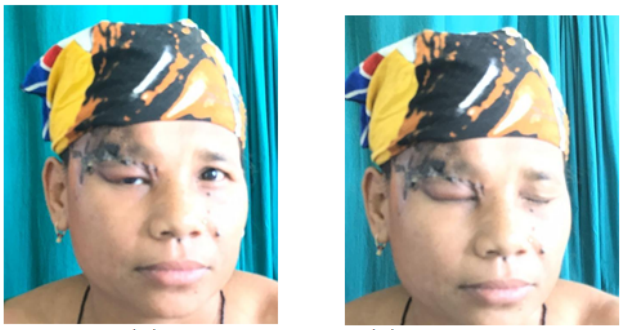

Figure 5: One-month follow-up pictures showing elated patient after complete excision of mass with right supraorbital incisional scar with patchy areas of hyperpigmentation and skin necrosis. She is able to (A) Open and (B) Close her right eye.

Though some patchy areas of skin necrosis, requiring no revision, were noted during one-month follow-up visit, there were no major postoperative complications, namely infection, dehiscence or bleeding. The final result was an acceptable cosmesis without visual obstruction and correction of severe facial disfigurement caused by the giant pulsatile mass.

Discussion

Scalp-orbital AVMs are rare congenital entities, which may have variable clinical pictures. Extensive lesions may cause ulceration, profuse bleeding and disfigurement and recurrence even after excision. In our case, the patient had undergone extensive excision with skin graft 10 years back; but the lesion had recurred and extended into the right orbital region causing visual obstruction and severe facial disfigurement. However, there was no skin ulceration or bleeding.

Over the last two decades, the reports of scalp AVM has increased significantly; and this is most likely due to improved diagnostic tools. In history, due to its unusual portly appearance, various nomenclature has been used to describe these lesions, including aneurysm cirsoides, racemose aneurysm, plexiformangioma, pulsating angioma, scalp arteriovenous fistula, arteriovenous aneurysm, aneurysmal varicose and arteriovenous malformations [2-4].

There is still ongoing controversy regarding clear-cut etiology of scalp AVM, it maybe congenital, traumatic, [5-12] infectious, inflammatory and idiopathic [13-19]. Most authors accept that it is caused by penetrating or blunt trauma; [20,21] some are congenital (spontaneous); [22,23] while few can be iatrogenic such as due to craniotomy, hair transplantation [24-27] or even intravenous scalp infusion. Congenital scalp AVM may present at birth; but, in majority, it remains asymptomatic until adulthood, [3,20,28] similar to our case.

Based on the extent of the blood flow, scalp AVM can be divided into low-flow or high-flow AVM. The low-flow lesions are usually well-demarcated, which include cavernoma, cavernous hemangioma, venous malformation and sinus pericranii, can easily be diagnosed with MRI and are treatable by sclera therapy or embolization alone. On the other hand, high-flow AVMs are large and extensive, producing loud bruit [6] heard on auscultation, containing multiple feeding and draining vessels and frequently requiring surgical excision. They may even reduce cerebral blood flow causing so-called “steal” phenomenon [29].

In their study, Shenoy and colleagues [20] classified these scalp AVM into two groups based their angiographic features: group I: The primary scalp AVM and group II: Secondary venous dilatations. In the primary scalp AVM, the arterial feeders arise from the calvarial branches of the external carotid arteries, ophthalmic arteries and vertebral arteries while the draining veins enter into the scalp venous system. In group II, the dilated channels are the main outflow vessels of the intracranial vascular malformations. Accordingly, our case belonged to group I.

Regarding anatomical location, scalp AVM are roughly evenly distributed located in the frontal, temporal and parietal regions [30]. Generally, the major feeding vessels are located in the subcutaneous layer of the scalp, often originating from external carotid, occipital and supraorbital arteries [31]. Frequently, STA is involved in traumatic cirsoid aneurysm due to its long unprotected course [31-33]. Inour case, being a high-flow AVM, it had multiple feeders, including from anterior, supratrochlear, supraorbital branches of STA and angular branches from both facial arteries. However, major feeder was from the right ophthalmic artery intraorbitally.

The clinical manifestation of AVM is highly variable, presenting with small pulsatile mass to extensive craniofacial disfigurement and grotesque appearance, producing throbbing headache, localized pain, proptosis, visual hindrance, epistaxis, tinnitus, numbness, necrosis, ulceration or hemorrhage [20,31,33,34].

In regards to the optimum management of such lesion, it may be quite complicated due to various factors such as the highly complex vascular anatomy, large collateral link, high-flow rate, massive blood loss, intracranial and facial involvement causing cosmetic and neurological problems after surgical intervention. The aim of treatment should be to completely eliminate the lesion from the normal scalp circulation. These lesions are generally diagnosed based on clinical observation as most are quite noticeable.

Recently, spectra of neuro-imaging modalities are available for diagnosing this pathology, to know the extent and complexity of the lesion and to understand the arterial feeders of the vascular nidus and to decide the best treatment strategy. These include plain and contrast Computerized Tomography (CT), Three-Dimensional CT Angiography (3D-CTA), conventional extracranial and intracranial angiography, Digital Subtraction Angiography (DSA), cranial doppler and Magnetic Resonance Imaging (MRI). Among them, angiography is considered as the gold standard of investigation to delineate the lesion, to understand its angioarchitecture and to exclude any intracranial component [8]. It is of great importance for diagnosis and treatment selection and particularly employed for the determining main cranial feeders. On the other hand, MRI of brain shows flow void signs due to rapid flow in the lesion [3,8,16,35] and is significant in establishing the diagnosis. In our case, we were able to obtain 3D-CTA which was able to clearly demonstrate the extent of the lesion and the multiple arterial feeders, making it possible for proper preoperative surgical planning.

Generally, this is a complex disease requiring multidisciplinary approach. In regards to treatment options for craniofacial AVM, there are numerous modalities, which include direct ligation of supplying vessels, extensive surgical excision [4,23,36-38], embolization *via* transarterial and transvenous routes [6,20,30,37], percutaneous intralesional injection of sclerosant [36,37] and electro-thrombosis [39,40]. Conventionally, the most common modality and treatment of choice that is curative is still the surgical excision [13,16,20].

However, with new advents, endovascular approach by itself or combination with surgery has been recommended for these complex malformations [41,42].

Variety of agents have been applied for pre or intraoperative embolization of these lesions, which include boiling water, absolute alcohol, gelfoampledgets, fibrin glue, thrombin, platinum coils, cyanoacrylate, sclerosant and onyx [43].

There are numerous innovative techniques to reduce bleeding during surgery, which includes percutaneous suturing of the feeding vessels [20], interlock-suturing along the incisional margin and tamponading with tourniquet, making stepwise incision with manual digital pressure and compressing the flap with intestinal clamp [31]. In our case, we applied the percutaneous technique of suturing to gain proximal vascular control prior to incision.

Due to the abnormal fistulous communication, AVM must be completely eliminated because recurrence or enlargement is reported after an incomplete resection [36]; so, emphasis should be given to enbloc resection of the vascular nidus and tight ligation of all the feeders to prevent recurrence. Recurrence has been reported as late as 18 years after surgical resection [5]. In our case, the lesion seems to have recurred after 8 years. After surgical excision, common postoperative complications include wound infection, sepsis, recurrence, hemorrhage and scalp necrosis [3,4,8,16]. Except for some patchy area of skin necrosis, none of other complications occurred in our case.

In our case, we did not apply preoperative embolization due to the involvement of right orbit rather we were able to attain proximal control by ligating the major arterial feeders percutaneously and eventually excising the lesion completely in to with minimal blood loss.

Conclusion

Scalp-orbital AVM is a rare entity which may cause proptosis, visual obstruction and severe long-standing facial disfigurement as in our case. With appropriate planning prior to surgery, proximal control of feeding arteries just before incision and complete removal of lesion in toto, large high-flow scalp-orbital AVM can be excised safely without any major complication and with acceptable cosmesis.

References

1. Huber P, Krayenbühl H, Yasargil MG (1982) Cerebral angiography (2nd edn). New York: Thieme.
2. Khodadad G (1973) Arteriovenous malformation of the scalp. Ann Surg 177: 79–85.
3. Fisher-Jeffes ND, Domingo Z, Madden M, de Villiers JC (1995) Arteriovenous malformations of the scalp. Neurosurgery 36:656–60.
4. Muthukumar N, Rajagopal V, Manoharan AV, Durairaj N (2002) Surgical management of cirsoid aneurysms. Acta Neurochir 144: 349–56.
5. Wilkinson HA (1971) Recurrence of vascular malformation of the scalp 18 years following excision. Case report. J Neurosurg 34: 435-7.
6. Barnwell SL, Halbach VV, Dowd CF, Higashida RT, Hieshima GB (1989) Endovascular treatment of scalp arteriovenous fistulas associated with a large varix. Radiology 173: 533–9.
7. Lanzino G, Passacantilli E, Lemole GM, McDougall C, Spetzler RF (2003) Scalp arteriovenous malformation draining into the superior sagittal sinus associated with an intracranial arteriovenous malformation: Just a coincidence? Case report. Neurosurgery 52: 440-443.
8. Matsushige T, Kiya K, Satoh H, Mizoue T, Kagawa K, Araki H (2004) Arteriovenous malformation of the scalp: Case report and review of the literature. Surg Neurol 62: 253–259.
9. El Shazly AA, Saoud KMF (2012) Results of surgical excision of cirsoid aneurysm of the scalp without preoperative interventions. Asian J Neurosurg 7: 191-196.
10. Nishijima I, Ikemura R, Gushiken M, Miyagi K, Iha K (2012) Nonsurgical treatment of scalp arteriovenous malformation using a combination of ultrasound-guided thrombin injection and transarterial coil embolization. J Vasc Surg pp: 833-836.
11. Dmytriw AA, terBrugge KG, Krings T, Agid R (2014) Endovascular treatment of head and neck arteriovenous malformations. Neuroradiology 56: 227-36.
12. Munakomi S, Bhattarai B, Cherian I (2015) Conquering the odds: Cirsoid aneurysm with holocranial feeders-staged embolization, excision and grafting. Asian J Neurosurg 10: 259-261.
13. Hendrix LE, Meyer GA, Erickson SJ (1996) Cirsoid aneurysm treatment by percutaneous injection of sodium tetradecyl sulfate. Surg Neurol 46: 557–560.
14. HageZA, Few JW, Surdell DL, Adel JG, Batjer HH, et al. (2008) Modern endovascular and aesthetic surgery techniques to treat arteriovenous malformations of the scalp: Case illustration. Surg Neurol 70: 198-203.
15. Godwin O, Ayotunde O, Millicent O, Yvonne O (2005) Extracranial arteriovenous malformation of the scalp: Value of computed tomographic angiography. The Internet Journal of Radiology 5.
16. Senoglu M, Yasim A, Gokce M, Senoglu N (2008) Nontraumatic scalp arteriovenous fistula in an adult: Technical report on an illustrative case. Surg Neurol 70: 194–197.
17. Chowdhury FH, Haque MR, Kawsar KA, Sarker MH, Haque AM (2013) Surgical management of scalp arteriovenous malformation and scalp venous malformation: An experience of eleven cases. Indian J Plastic Surg 46: 98-107.
18. Raheja A, Satyarthee GD, Sharma BS (2013) Single, small, spontaneous, accessory, closed type, frontal sinus pericranii in a child: Favorable outcome with surgical excision. Neurol India 61: 680-683.
19. Rane SY, Lamba S, Gohil AJ, Gupta AK (2017) Compendium of scalp Arterio Venous Malformation (AVM) cases—A retrospective study and review. Eur J Plast Surg 41: 223-228.
20. Shenoy SN, Raja A (2004) Scalp arteriovenous malformations. Neurol India 52: 478–481.
21. Massimi L, De Bonis P, Esposito G, Novegno F, Pettorini B, Tamburrini G, et al. (2009) Vertex scalp mass as presenting sign of a complex intracranial vascular malformation. J Neurosurg Pediatr 3: 307–310.
22. Schultz RC, Hermosillo CX (1980) Congenital arteriovenous malformation of the face and scalp. Plast Reconstr Surg 65: 496-501.
23. Domingo Z, Fisher-Jeffes ND, de Villiers JC(2000) Surgical management of arteriovenous malformations of the scalp. In: Shmidek HN, editor. Operative neurosurgical techniques:

Indications, methods and results (4th ed). Philadelphia: Saunders Company pp: 1331–1338.

24. Souder DE, Bercaw BL (1970) Arteriovenous fistula secondary to hair transplantation. N Engl J Med 283: 473-474.
25. Barros d'Sa AA, Heard CE (1978) Arteriovenous fistula after hair transplantation. BMJ 1: 340.
26. Lanzieri CF, Sacher M, Som PM, Haimov M (1985) Arteriovenous fistula after hair transplantation. AJNR 6: 111-112.
27. Liounakos JI, Urakov T, Snelling B, Peterson EC (2018) Scalp arteriovenous fistula following hair transplantation: A case Report and review of the literature. Clin Med Rev Case Rep 5: 200.
28. Li F, Zhu S, Liu Y, Chen Y, Chi L, Chen G, et al. (2007) Traumatic arteriovenous fistula of the superficial temporal artery. J Clin Neurosci 14: 595–600.
29. Mast H, Mohr JP, Osipov A, Pile-Spellman J, Marshall RS, Lazar RM, et al. (1995) 'Steal' is an un-established mechanism for the clinical presentation of cerebral arteriovenous malformations. Stroke 26: 1215-1220.
30. Kasdon DL, Altemus LR, Stein BM (1976) Embolization of the traumatic arteriovenous fistula of the scalp with radiopaque gelfoampledgets. Case report and technical note. J Neurosurg 44: 753–756.
31. Hochberg J, Ardenghy M, Pait TG (1994) Scalp tourniquet. Br J Plast Surg 47: 194–198.
32. Gupta AK, Purkayastha S, Bodhey NK, Kapilamoorthy TR, Krishnamoorthy T, et al. (2008) Endovascular treatment of scalp cirsoid aneurysms. Neurol India 56: 167–172.
33. Burrus TM, Miller GM, Flynn LP, Fulgham JR, Lanzino GN (2009) Symptomatic left temporal arteriovenous traumatic fistula. Neurology 73: 570.
34. Manuel AM, Kalimuthu S, Pathmanathan SS, Narayanan P, Abidin ZZ, et al. (2017) Challenges in the management of massive intraorbital and hemifacial arteriovenous malformation as causing life-threatening epistaxis. Asian Journal of Surgery 40: 158-162.
35. Hasturk AE, Erten F, Ayata T (2012) Giant non-traumatic arteriovenous malformation of the scalp. Asian J Neurosurg 7: 39–41.
36. Shepard RH (1975) Proceedings: Cirsoidarteriovenous malformations of the scalp. J Neurol Neurosurg Psychiatry 38: 827–8.
37. Nagasaka S, Fukushima T, Goto K, Ohjimi H, Iwabuchi S, et al. (1996) Treatment of scalp arteriovenous malformation. Neurosurgery 38: 671–677.
38. Weinzweig N, Chin G, Polley J, Charbel F, Shownkeen H, et al. (2000) Arteriovenous malformation of the forehead, anterior scalp and nasal dorsum. Plast Reconstr Surg 105: 2433–2439.
39. Gardner AM, Stewart IA (1972) Treatment of arteriovenous malformation by endarterial electrocoagulation. Br J Surg 59: 146–148.
40. Heilman CB, Kwan ES, Klucznik RP, Cohen AR (1990) Elimination of a cirsoid aneurysm of the scalp by direct percutaneous embolization with thrombogenic coils. Case report. J Neurosurg 73: 296–300.
41. Komiyama M, Nishikawa M, Kitano S, Sakamoto H, Imai K, et al. (1996) Non-traumatic arteriovenous fistulas of the scalp treated by a combination of embolization and surgical removal. Neurol Med Chir 36: 162-165.
42. Tanaka T, Hasegawa Y, Kanki T, Abe T (2002) Combination of intravascular surgery and surgical operation for occipital subcutaneous arteriovenous fistula in a patient with neurofibromatosis type I. No Shinkei Geka 30: 309-13.
43. Cognard C, Januel AC, Silva NA, Tall P (2008) Endovascular treatment of intracranial dural arteriovenous fistulas with cortical venous drainage: New management using Onyx. Am J Neuroradiol 29: 235-41.