

# Fronto-temporal Scalp Arteriovenous Malformation: Case Report and Review of the Literature

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## Abstract

*Extracranial arteriovenous malformations (AVMs) are far less common than intracranial AVMs. Among them, scalp AVM's are rare vascular anomaly having a poorly reported incidence and natural history. They presented mostly with localized headache and a progressively enlarging pulsatile mass with or without features of overlying skin ulceration and hemorrhage. Complete surgical excision is the treatment of choice and considered curative.*

*We present a case of a scalp AVM of the right fronto-temporal region of a 28-years-old male in whom the mass had undergone progressive painless enlargement within six months. We also reviewed the relevant literature, discussed the pathogenesis, natural history, clinical features and treatment options for this rare vascular malformation.*

**Key words:** arteriovenous malformation; scalp AVM.

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## Introduction:

Arteriovenous malformations (AVMs) are high-flow lesions providing a direct communication between artery and vein without intervening capillary bed. AVMs are usually latent during infancy and childhood and may enter an active expanding phase in adolescence. Extracranial AVMs are rare vascular malformations<sup>1</sup>. These lesions often occur on the extremities or trunk where they may present as an enlarging soft tissue mass in the subcutaneous tissue, or may be located below the deep fascia and involve the musculoskeletal system. Although the mechanisms are poorly understood, infection,

trauma, or hormonal changes in puberty and pregnancy are known factors that can trigger the rapid expansion of AVMs. Schobinger described a useful clinical staging system that describes their progression<sup>1</sup>. AVMs initially present as warm pink-blue macules (stage I), proceed to enlarge with pulsations, thrills and bruits (stage II), subsequently can become painful, bleed or ulcerate (stage III) and finally can result in cardiac failure (stage IV).

Scalp AVM is a complex vascular network of feeding arteries and draining veins without an intervening capillary bed, having a 'nidus' located in the subgaleal plane. The draining veins are hugely dilated and tortuous because of direct shunting of high flow blood from the artery resulting in arterialization of these veins. Regarding its distribution, location of the occipital region is considered rarest among all. Usually, this lesion presents as palpable pulsatile scalp lump with or without audible bruit and overlying skin change. Other associated symptoms are headache, tinnitus and very rarely features of epilepsy<sup>2</sup>. Magnetic resonance imaging (MRI) is helpful in determining the extent of the lesion and any involvement of associated structures. Angiography is the ideal modality to determine the anatomy of the feeding and draining vessels including the extent of arteriovenous shunting, fistulae and vessel tortuosity. Angiography can also be used for therapeutic

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embolization of the AVM prior to excision to control peroperative hemorrhage<sup>3</sup>. Although different treatment modalities were mentioned in the previously reported literature, total surgical excision is considered curative among all.

#### Case Report:

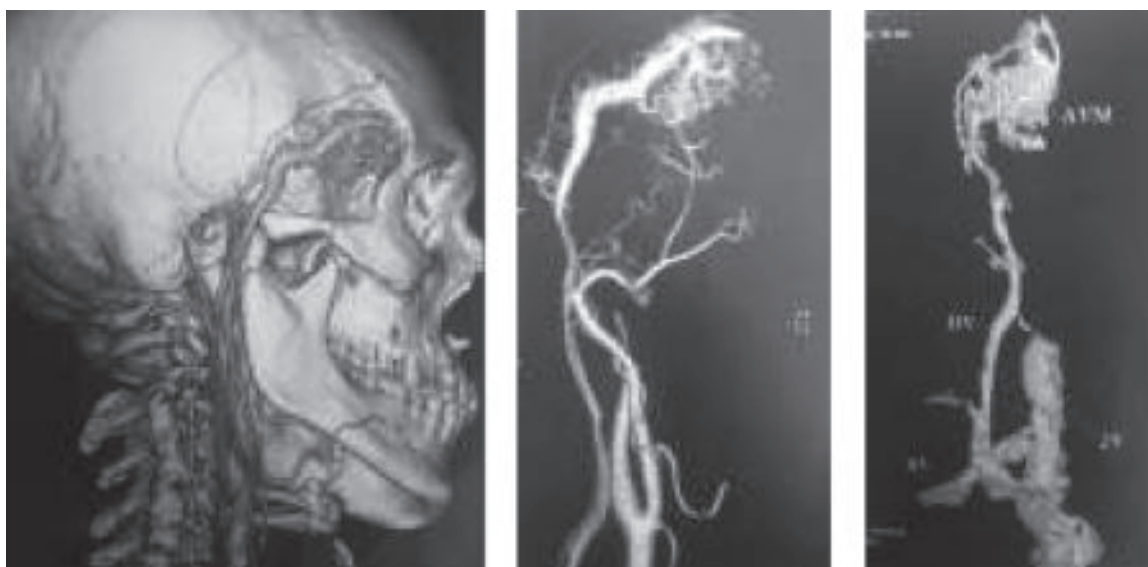
A 28 years old male presented with a painless swelling in his right fronto-temporal region. The mass had undergone progressive enlargement within six months. The swelling was ill defined, about 4 by 3 cm in diameter, pulsatile, non- tender, non-compressible (Figure 1A, 1B). After clenching of teeth, it became

prominent (Figure 1C) No change was observed after coughing and Valsalva maneuver.

CT scan of brain with contrast with CT angiogram showed a serpiginously enhancing mass at the right fronto-temporal region. CT angiogram with 3D reconstruction showed a well-defined nidus, fed by both superficial and deep temporal artery. There was a single dilated tortuous draining vein at the postero- superior aspect of the lesion which ultimately draining into subclavian vein (Figure 2A, 2B, 2C).



**Fig. 1:** Patient presenting with an ill-defined palpable scalp lesion on the right fronto-temporal region which became prominent after clenching of teeth.



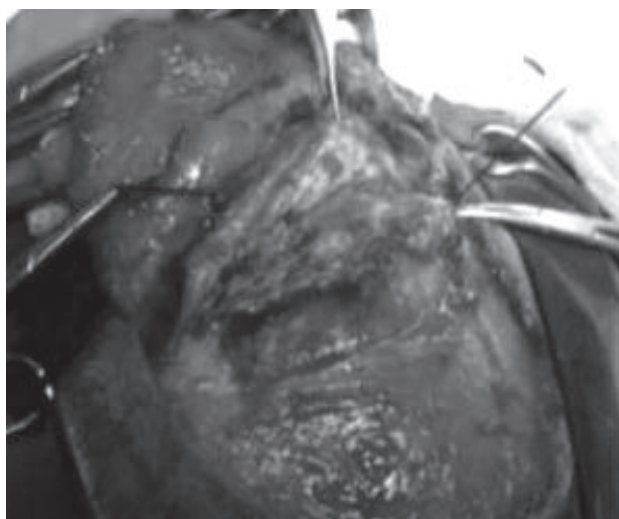
**Fig 2:** Right fronto-temporal scalp AVM, fed by right superficial temporal artery and deep temporal artery. There is a single dilated draining vein, draining blood into subclavian vein.

During operation, curvilinear incision was given 1 cm in front and above the tragus. After subgaleal dissection, temporal fascia incised

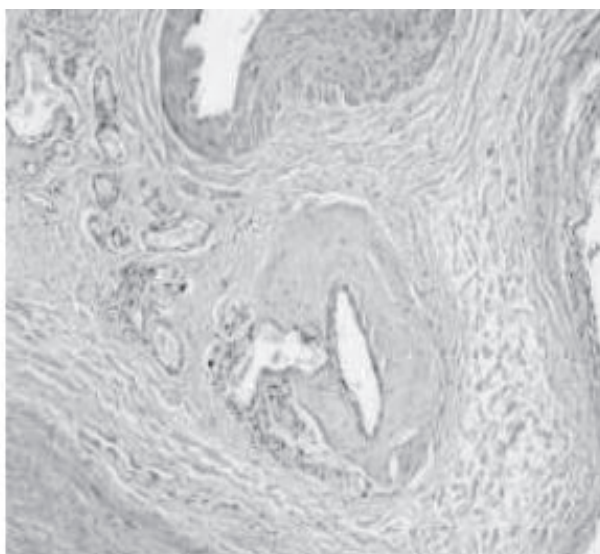
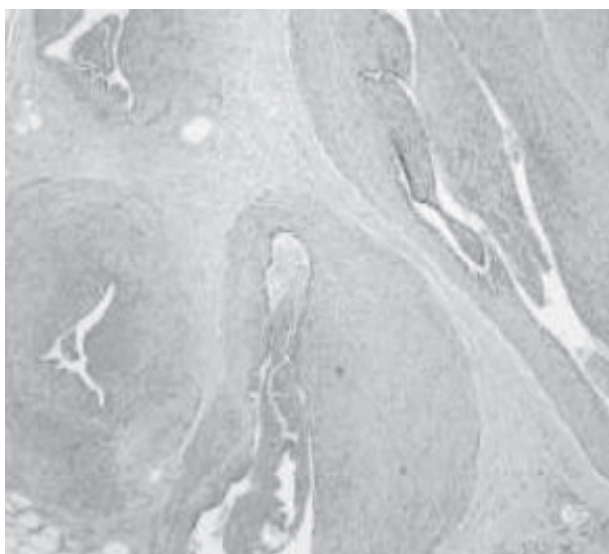
leaving a small cuff to facilitate future closure. Draining vein was identified first, following which nidus was reached. Superficial temporal artery was coagulated first; later on coagulation of deep temporal artery was done after slight retraction of nidus. Diminution of the nidus size was observed. Circumferential coagulation and dissection of nidus

was done. At the end of the procedure, draining vein was ligated and cut away. Ensuring hemostasis, layered closure was done. Postoperative period was uneventful (Figure 3A, 3B).

The pathology result demonstrated presence of multiple thin walled and thick walled blood vessels, some of which contained thrombi. Smaller vessels without muscle coat were also present and the diagnosis of arteriovenous malformation was confirmed (Figure 4A, 4B).



**Fig 3:** A: Dilated draining vein, marked by silk suture. The nidus was beneath the temporal fascia. B: showing closure of the fascia.



**Fig 4:** Haematoxylin and Eosin stained slides showing presence of multiple thin walled and thick walled blood vessels, some of which contained thrombi.

**Table-1**  
*Summary of reported cases of scalp AVM*

| Case | Author     | Year | Age (yrs) | Sex | Size (cm) | Side   | Feeding arteries                                    | Treatment                                     |
|------|------------|------|-----------|-----|-----------|--------|---|---|
| 01   | Vasconez   | 1973 | 2         | F   | 10. 12    | center | bilateral STA & OA                                  | Total excision                                |
| 02   | Stucker    | 1974 | 59        | M   | ?         | left   | left OA   | ligation and electrocautery                   |
| 03   | Mohanty    | 1976 | 1.5       | F   | 10. 6     | right  | bilateral STA                                       | ligation                                      |
| 04   | Takahashi  | 1977 | 29        | F   | 10. 8     | left   | left IMA & STA                                      | total excision                                |
| 05   | Kaufman    | 1980 | 17        | F   | ?         | right  | right STA & OA                                      | embolization and total excision               |
| 06   | Goya       | 1981 | 52        | M   | 8. 8      | center | bilateral STA, bilateral MMA & bilateral OA         | embolization and total excision               |
| 07   | Goya       | 1981 | 17        | F   | 3. 3      | right  | right STA   | total excision                                |
| 08   | Ohno       | 1981 | 32        | M   | 4.7       | left   | left OA   | embolization and total excision               |
| 09   | Konishi    | 1982 | 44        | M   | 4.5       | right  | right OA  | total excision                                |
| 10   | Yamaki     | 1982 | 31        | M   | 5.5       | left   | left STA & DTA                                      | ligation, embolization and total excision     |
| 11   | Tani       | 1983 | 11        | F   | small     | right  | right STA   | total excision                                |
| 12   | Shimoda    | 1985 | 25        | M   | ?         | center | bilateral STA & OA                                  | total excision                                |
| 13   | Komatsu    | 1989 | 43        | M   | 4. 4      | right  | right STA   | total excision                                |
| 14   | Nishimura  | 1996 | 47        | F   | 3. 3      | right  | right STA   | total excision with ligation of aneurysm neck |
| 15   | Kuroki     | 1999 | 23        | M   | 4. 3      | left   | left STA OA PAA & MMA                               | total excision                                |
| 16   | Nishimura  | 1999 | 27        | F   | 3.5 . 3.5 | center | bilateral OA MMA pial artery                        | embolization and total excision               |
| 17   | Matsushige | 2003 | 21        | F   | 7. 7      | right  | right STA OA PAA bilateral MMA right pial artery    | embolization and total excision               |
| 18   | Mohamed    | 2008 | 30        | M   | ?         | center | right and left STA right and left ophthalmic artery | total excision                                |
| 19   | Tabuchi    | 2013 | 75        | M   | 5         | right  | right STA   | total excision                                |
| 20   | Worm       | 2015 | 33        | M   | ?         | left   | left STA OA   | total excision                                |
| 21   | Kannath    | 2017 | ?         | M   | ?         | left   | left STA OA   | percutaneous direct puncture embolization     |
| 22   | Khan       | 2018 | 28        | M   | 3. 2      | right  | right STA DTA                                       | total excision                                |

The patient reported mild headache at postoperative period. However, his headache subsided with the course of time.

#### **Discussion:**

Hemangiomas and vascular malformations are benign lesions of blood vessels. To date, many different terms

and classifications have been suggested for these types of lesions. Today, most reports use the classification scheme suggested by Mulliken and Glowacki in 1982, based on the histological and clinical features of the lesion. This system divides vascular anomalies into hemangiomas, which are



neoplastic lesions with endothelial hyperplasia, and vascular malformations, which are congenital lesions with normal endothelial turnover<sup>4</sup>. From Krayenbuhl and Yasargil's review of 800 cases, 8.1% cases were extracranial AVM<sup>5</sup>. Khodadad reviewed 148 cases of scalp AVM in which 55% were thought to be congenital because of having no preceding history of trauma<sup>6</sup>. Our case is considered to be congenital because this patient also had no history of trauma. The natural history of these lesions is poorly understood. Matsushige was first credited for serial angiographic documentation of scalp

AVM which depict the progressive growth of scalp AVM, acquisition of new feeder and de novo aneurysm formation<sup>7</sup>.

We have reviewed 22 cases of scalp AVM<sup>2,7-10,20,22-32</sup>, including our own (Table 1). After analyzing location, size of nidus, feeders and treatment modalities, it has been showed that larger the size of the nidus-greater number of feeders supplied the nidus and not only from branches of ECA but also branches from ICA. With the course of time, expansion of nidus evolves 3 stages (Table 2). In stage 1, nidus fed by branches from extracranial carotid artery. In stage 2, additional feeder comes from meningeal arteries. In stage 3, nidus is supplied by branches of both ECA and ICA<sup>7</sup>. According to this staging, our reported case is in stage 1b.

In cerebral AVM, VEGF is a known angiogenic factor to promote its growth. Matsushige showed strong expression of VEGF in his reported case<sup>7</sup>. However, in our case, VEGF positivity was not analyzed due to lack of the availability of this test in our country.

**Table-2**  
Stages of scalp AVM

| Stages | Source of feeders   |
|--------|---|
| 1a     | single feeder arises from subcutaneous artery                   |
| 1b     | multiple feeders from subcutaneous arteries                     |
| 2      | multiple feeders from subcutaneous and meningeal arteries       |
| 3      | multiple feeders from subcutaneous, meningeal and pial arteries |

Regarding clinical feature, scalp AVM most commonly presents with a palpable pulsatile mass with or without feature of overlying skin ulceration and hemorrhage. After reviewing literature, Mohanty reported a scalp AVM which was associated with epilepsy and mental

retardation<sup>2</sup>. Ohno et al. reported a case presented with features of cerebral ischaemia due to steal phenomenon<sup>8</sup>. In our reported case, there was a palpable pulsatile mass with audible bruit (Figure 1). Overlying skin was intact and free from the mass.

History, clinical examination, MRI, CT angiography and/ or digital subtraction angiogram are adequate for an accurate diagnosis of these lesions. MRI is helpful to differentiate AVM from other vascular lesions, and also defines its intracranial extension and additional intracranial vascular lesion. It can also differentiate high flow lesion from low flow lesion. CT angiography helps to understand the angioarchitecture of the lesion, involvement of adjacent bone and excludes intracranial extension. It is also helpful to determine the acquisition of intracranial feeders. Plain or color Doppler ultrasonography is effective in evaluating these malformations by analyzing its flow velocity. DSA is helpful for those patients requiring surgical intervention or preoperative

embolization. In our case, we did CT scan of brain with contrast with 3D CT angiography which revealed there was a serpiginously enhancing lesion overlying right fronto-temporal region, lying superficial to the temporalis muscle. There was a single compact nidus fed by both superficial and deep temporal artery. There was a dilated tortuous single draining vein at the posterior superior aspect which ultimately drained into right subclavian vein. There was no perinidal aneurysm and no intracranial feeder (Figure 2).

There are several treatment options of the management of scalp AVM that include surgical excision, ligation of feeding vessels, interlocking suture along the line of incision and the use of tourniquet and intestinal clamp over the base of the flap. Among them, total surgical excision is considered curative. Incomplete resection is responsible for hemorrhage, future recurrence and expansion of the lesion. Recurrence of the lesion was seen as late as 18 years after complete surgical excision. It should be emphasized that embolization or ligation of the feeding arteries without surgical resection usually results in rapid recruitment and dilation of previously microscopic collateral blood vessels<sup>9</sup>.

Now-a-days, endovascular management has been increasingly reported as the mainstay therapy or

surgical adjunct in the treatment of scalp AVMs especially for large one. Transarterial, transvenous, and percutaneous approaches with different embolic agents such as polyvinyl alcohol (PVA) particles, n-butyl cyanoacrylates (n-BCA), and EVOH copolymers are all described in the literature<sup>10, 12-18</sup>. Recently, percutaneous direct puncture embolization using SQUID is an additional treatment option for patients with scalp AVMs.

It can be considered as the primary treatment option for patients unwilling for surgery<sup>19</sup>.

Scalp AVM is an external lesion so attention should be paid on the cosmetic outcome too. Skin grafting may be necessary if AVM is large for aesthetic reason<sup>20,21</sup>. Our patient underwent complete surgical excision without significant peroperative hemorrhage and cosmetic deformity because AVM was small, overlying skin was free and there was no intracranial extension.

### Conclusion:

Because it is reported that scalp AVM will expand and captures additional feeders, even from branches of internal carotid artery with the course of time which ultimately leads to incomplete resection, cosmetic deformity and recurrence of the lesion. So, it is better to resect this lesion at the earlier stage. To achieve a permanent cure, a combination of surgery and preoperative embolization for selected cases to reduce intraoperative bleeding can be the best choice in the treatment of scalp AVM.

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