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Case Report

Extracranial arterio-venous malformation presenting as a scalp swelling

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ABSTRACT

It is a challenge to diagnose and treat extracranial arteriovenous malformations (AVMs) because of complex vascular malformations. AVMs are congenital vascular shunts with long standing expansion of vascular channels, collateralization of microcirculation and localized tissue infiltration. Even though the exact etiopathogenesis of AVMs remains undefined however newer genetic/molecular basis of the same are evolving. Any age can be affected by AVMs however it is seen to increase in dimension after an early period of inactivity. They may present at any age following an early quiescent period. Diagnosis is based on vascular staining, soft tissue expansions, progressive growth/ warmth and pulsations.

Keywords: Arterio-venous malformation; Extracranial; Head and neck

INTRODUCTION

Cervicofacial or subcutaneous haemangiomas or venous malformations are common compared to extra-cranial Arterio-venous malformation (AVM) in the scalp. It is an abnormal fistula communication between feeding arteries and draining veins without intervening capillary bed. It usually presents in 6 to 11 years of age, 10-19 years or 18-30 years of age. It can also cause massive haemorrhages due to dryness of the overlying skin and injuries[1]. In this report we describe the clinical, radiological and histopathological features of a patient with a scalp AVM.

CASE HISTORY

A 22-year-old female presented at tertiary care hospital with complain of a painless forehead swelling since birth, increasing in size for the past three years. Initially, it was a reddish small growth. Local pulsations felt and raised local temperature. Patient did not complain of tinnitus, giddiness and headache. The lesion bled once following trauma about a year prior to admission.

MRI of the swelling revealed a predominantly hyper-intense subcutaneous mass in the midline of the forehead on both T1 and T2 weighted images suggestive of vascular malformation. Biopsy of the lesion was sent for histopathological examination. Gross finding shows small globular structure measuring 3x2.9x2.5 cm in size, greyish-black in colour and having hairs on the surface. On cut section, the firm growth was homogenous whitish in colour (Figure 1).

Microscopic examination: Pathological examination and microscopic sections revealed skin and subcutaneous tissue with proliferation of variably sized thickened medium to large-sized blood vessels separated by fibro-collagenous tissue and adipocytes. Microscopy also shows admixture of malformed vessels such as capillaries, arteries and venules and abrupt changes in thickness of medial and elastic layers of vessels with abnormal vascular dilation (Figure 2 and Figure 3).

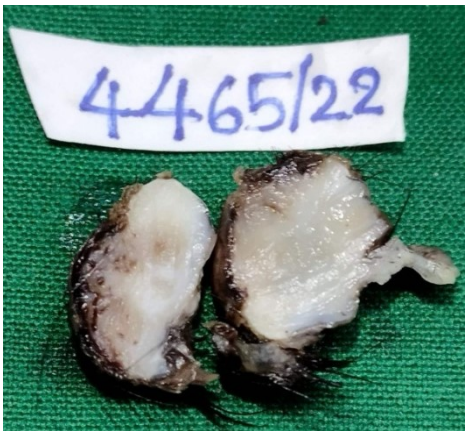

Figure 1: Gross photograph of excised scalp tissue showing external surface covered with hairs. Cut surface shows homogenous white, firm areas.

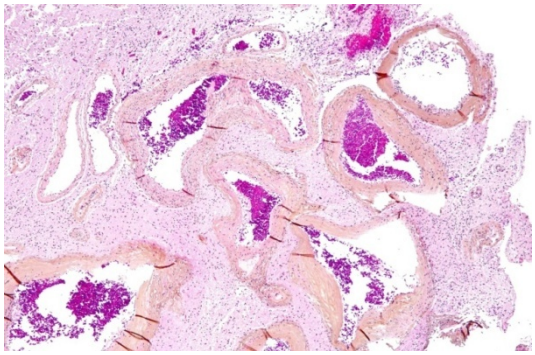

Figure 2: Microphotograph showing variably sized thickened, dilated blood vessels with fibrocollagenous tissue (H&E, x 400).

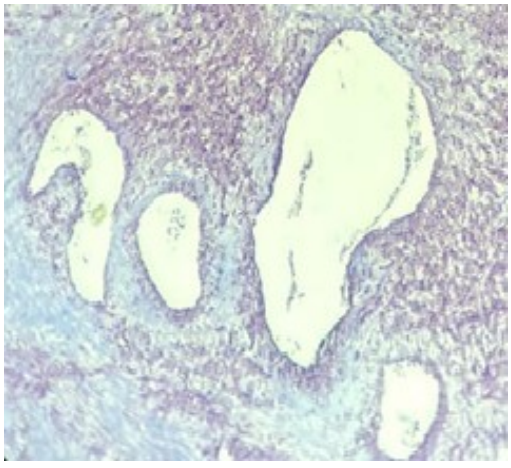

Figure 3: Microphotograph showing variably sized dilated blood vessels with surrounding expanded connective tissue stained pale blue in colour (Masson Trichrome stain, x 400).

DISCUSSION

Scalp AVMs are rare. Not many latest publications exist in this regard. Furthermore, these lesions are sometimes under recognized and managed as a simple case of presumed lipoma or sebaceous cyst, especially when the lump was not pulsatile[1,2].

The etiology of these lesions are classified into congenital or acquired. Acquired are usually after trauma, surgery or hair transplantation. The exact mechanism of congenital formation is not yet fully understood. Still, multiple theories like either formed from persistent embryonic arterio-venous communication, arise from vascular hamartoma or form at the site of arterio-venous crossing[3].

The median age of presentation of scalp AVM is 25 years, with noticeable male predominance; ratio is M: F, 2.5:1[3,4]. Our case was a 22-years-old female. The most common presenting symptom for such lesions is pulsatile mass followed by headache and tinnitus. Hemorrhage has been seen as the presenting symptom of these cases in up to 11 %.

Although these lesions are benign and exclusively extracranial, they can cause cerebral pathology which resulting in neurological deficits or seizures, not seen in this case. The suggested mechanism develops a stealing phenomenon that diverts the blood towards the external carotid artery leaving cerebral watershed areas prone to ischemia[3].

Imaging modalities that helps in the diagnosis include Doppler ultrasound that can detect the vascular etiology of a lump in the clinic. Magnetic resonance imaging (MRI) can demonstrate the dimensions and complexity of these lesions, especially with MRA sequences. MRI is an important preliminary investigation as different cervicofacial vascular lesions can be assessed to arrive at a single diagnosis of AVM. MRI can assess intracranial extension in such cervicofacial AVM lesions.

Angiography is undertaken to delineate the lesion and to exclude an intracranial component. Computed tomographic angiography (CTA) can also display the vascular pathology and depicts excellent correlation with the underlying bony structures. Digital subtraction angiography (DSA) is accepted as the 'gold standard' when evaluating AVMs. It is crucial to detect any intracranial extension and the number of feeding arteries which is essential for planning a management[2,4]. However since DSA is costlier and not available in all medical set-ups, clinico-histopathological correlation with local MRI of extra-cranial swelling.

The diagnosis of extra-cranial AVM can be effectively done clinically by experienced clinicians/surgeons. Like in our case, the final diagnosis is established on Histopathology. Pathology of AVM shows admixture of malformed vessels such as capillaries, arteries and venules and abrupt changes in thickness of medial and elastic layers of vessels, abnormal vascular dilation with proliferation of fibro-collagenous tissue and adipocytes formation.

CONCLUSION

Scalp arterio-venous malformation is a rare condition but it is more common in younger age compared to extremities of the ages. Its final diagnosis is established on multi-disciplinary approach by clinical evaluation with radio-pathological correlation.

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