

## Extracranial Arterio-Venous Malformation in the Gluteal Region - A Rare Case Report.

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### ABSTRACT

Extracranial arteriovenous malformations (AVMs) are less common than intracranial AVMs. Extracranial AVMs mostly involve the skin, subcutaneous tissue and even the musculoskeletal system. The prevalence of vascular malformations is estimated to be 1.5% in the general population. The goal of treatment is to control the AVM, using tumor debulking or arterial embolization; complete tumor excision is usually not possible and the local recurrence rate is high. Intramuscular AVMs are rare and these lesions can be curable with complete surgical resection. This case is reported for its rare incidence and uncommon location in a 17 year old male who underwent surgical excision of the lesion and diagnosis of AVM was confirmed on histopathology.

**Keywords:** AVM- Arterio-venous malformation, Angiography and MRI- Magnetic Resonance Imaging.

### INTRODUCTION

Arteriovenous malformations (AVMs) are high-flow lesions providing a direct connection between an artery and a vein. AVMs are usually latent during infancy and childhood and may enter an active expanding phase in adolescence. Extracranial AVMs are very less common than intracranial AVMs.<sup>[1]</sup> The majority of extracranial AVMs involve the head and neck, followed by the lower extremities and trunk.<sup>[2,3]</sup> These lesions 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. Less than 1% of vasoformative tumors throughout the body occur in the skeletal muscle and AVMs here are extremely rare.<sup>[4]</sup> 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].

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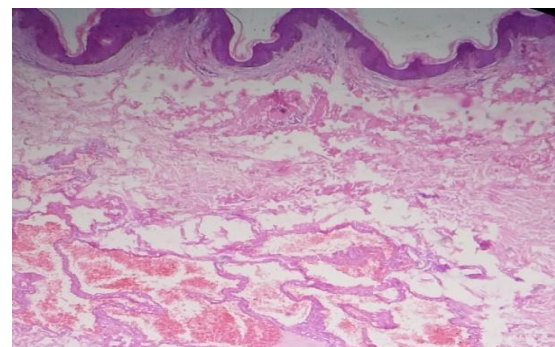
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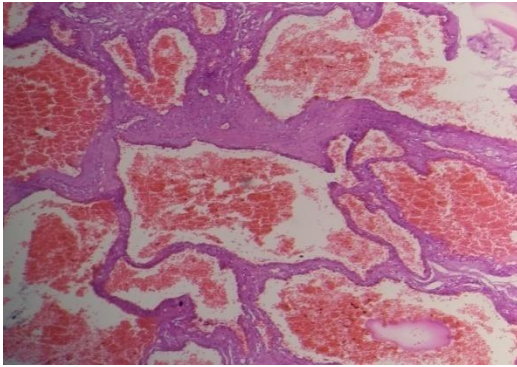
Angiography is the ideal modality to determine the anatomy of the feeding and draining vessels including the extent of arteriovenous shunting,

fistulae and vessel tortuosity. It can also be used for therapeutic embolization of the AVM.<sup>[5]</sup>

### CASE REPORT

A 17 year old male presented with c/o dilated vessels in the right gluteal region since 4 years which started increasing in size since 1 year. It was also associated with c/o pain since 6 months which occurred while sitting and walking. A clinical diagnosis of AVM was made. Surgical excision under spinal anesthesia was done and the resected tissue was skin covered and measured 9x3.5x3 cm in size. External surface of skin showed bluish worm-like dilatations. Microscopy showed malformed and dilated blood vessels with hypertrophied vessel wall and haemorrhages in the dermis with hyperkeratotic stratified squamous epithelium and mild chronic inflammatory infiltrate which were consistent with the clinical diagnosis of AV malformation. Follow up examination was unremarkable.





**Figure 1 & 2: Photomicrographs (H and E 100x and 400x) showing hyperkeratotic epidermis and dilated vessels in the dermis.**

### DISCUSSION

Hemangiomas and vascular malformations are benign lesions of blood vessels. Currently the classification scheme suggested by Mulliken and Glowacki in 1982, based on the histological and clinical features of the lesion is being used. 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>[6]</sup> Arteriovenous malformations can be either congenital or acquired. Formation of communicating channels between mature arteries and veins, associated with the appearance of supernumerary branches due to overgrowth of vascular elements can be caused by developmental arrest or misdirection. Acquired AVMs are rarer. Infection, trauma or hormonal changes in puberty and pregnancy are known factors that cause the arteriovenous anastomoses and rapid expansion of AVMs.<sup>[7-9]</sup> Patient history and clinical examinations are usually adequate for an accurate diagnosis of these lesions. Angiography, Magnetic resonance imaging, CT, Plain or Color Doppler Ultrasonography are effective in evaluating these malformations.<sup>[3,10]</sup>

### CONCLUSION

Extracranial AVMs are extremely rare than intracranial AVMs. Asymptomatic cases are treated conservatively. Diagnosis and management of AVM integrates surgical therapy with embolism and sclerotherapy. It improves the results with less morbidity and less recurrence rates.

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