Arteriovenous Malformation of Gluteal Region.

KEYWORDS arteriovenous, extracranial, gluteal.

Introduction
The prevalence of vascular malformations is estimated to be 1.5% in the general population. Arteriovenous malformations (AVMs) are high-flow lesions providing a direct connection between an artery and a vein. Extracranial AVMs are far less common than intracranial AVMs. The majority of extracranial AVMs involve the head and neck, followed by the lower extremities and trunk. Less than 2% of the total AVMs involve pelvis; there are less than 60 cases described in the world literature. They present as an enlarging soft tissue mass in the subcutaneous tissue, or may be located below the deep fascia and involve the musculoskeletal system. AVMs are usually identified during adolescence because in this age group they are in active expanding phase and they are latent during infancy and childhood. This case is reported for its rare incidence and uncommon location.

Case report
A 23-year-old woman presented with a large swelling with blue discoloration over the left buttock since 4 yrs. Initially the swelling was small and gradually progressed to the present size of 13 cm. On examination 13 cm swelling was non tender and pulsatile. By applying pressure, the tumors could be emptied almost completely, but refilled rapidly. The diagnosis of congenital arteriovenous malformation was made which might have been present since birth but became symptomatic during adolescence. Surgically resected specimen was 13.5X11.5X4.2 cm in size. External surface of skin showed bluish worm-like dilatations. Cut surface showed numerous blood filled spaces. Microscopy showed AVM with venules showing arteriolisation consisting of hyalinization and fibrotic changes. Arteries had thickened irregular wall with narrow lumen.

Figure 1. Photograph of cut surface of AVM showing numerous blood vessels.
Most are diagnosed in the second and third decades of life. There is no reported gender difference. As described by Schobinger, natural history of these lesions follows four stages: quiescence, expansion, destruction, and decompensation. Symptoms depend on the size and site of the lesion: pelvic lesions may grow to a large size before they are detected. Paradoxically, systemic haemodynamic effects rarely occur because communications are multiple, tortuous, and narrow, maintaining peripheral vascular resistance. High output cardiac failure has been observed only in very bulky lesions and in pelvic lesions during pregnancy.[1,3,4,5,12,13] According to Schobinger, clinical staging system the progression of AVMs can be divided into the following stages: 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).[1,3]

Discussion
Extracranial congenital arteriovenous malformations (AVMs) are rare clinical entities that can be progressive in nature. According to Mulliken and Glowacki in 1982, vascular anomalies can be classified into intohemangiomas, which are neoplastic lesions with endothelial hyperplasia, and vascular malformations, which are congenital lesions with normal endothelial turnover.[3,8]

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. Engorgement and ageing of the component elements causes further increase in size. Acquired AVMs are rarer. The aetiopathogenesis of acquired AVMs is poorly understood. Infection, trauma or hormonal changes in puberty and pregnancy are known factors that cause the arteriovenous anastomoses and rapid expansion of AVMs. Acquired AVMs are neoplastic lesions with endothelial hyperplasia, and anomalies can be classified into intohemangiomas, which are neoplastic lesions with endothelial hyperplasia, and vascular malformations, which are congenital lesions with normal endothelial turnover.[3,8]

Treatment
The diagnosis and management of AVM is a multidisciplinary approach. Angiography, Magnetic resonance imaging, CT, and color Doppler ultrasonography are effective in evaluating these malformations,[3,14-17] Treatment depends on the timing of presentation, extent of malformation, any visceral involvement and the effect of selective embolisation.[5] Asymptomatic static lesions can be monitored or treated conservatively. The goal of treatment is to control the AVM using tumor debulking or arterial embolization. Major blood loss during surgery leading to incomplete removal of the lesion and its recurrence makes it problematic and dangerous which can be prevented by preoperative embolization. Oophorectomy in women may lead to regression of an AVM which shows association between AVMs and high oestrogen levels. However, oestrogen blocking agents have not been used to treat these lesions.[2,4,15,16,19]

Conclusion
Extracranial AVMs are far less common than intracranial AVMs. Asymptomatic cases are treated conservatively. Diagnosis and management of AVM is a multidisciplinary approach that integrates surgical therapy with embolism and sclerotherapy. It improves the results with less morbidity and recurrence during early follow-up.

REFERENCE