RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 4 | April 2015 | ISSN - 2249-555X

Arteriovenous Malformation of Gluteal Region.

KEYWORDS: arteriovenous, extracranial, gluteal.

Sandeepa S	Hemalatha AN
Department of Pathology, Dr B R Ambedkar Medical college Kadugondanahalli,Bangalore,Karnataka.	Department of Pathology, Dr B R Ambedkar Medical college, Kadugondanahalli,Bangalore, Karnataka.

Mahesh A	Ananda BB	Shenoy V
Department of Surgery, Dr B R Ambedkar Medical college, Kadugondanahalli,Bangalore	Department of Surgery, Dr B R Ambedkar Medical college, Kadugondanahalli,Bangalore	Department of Surgery, Dr B R Ambedkar Medical college, Kadugondanahalli,Bangalore

ABSTRACT *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.[1,2,3] The majority of extracranial AVMs involve the head and neck, followed by the lower extremities and trunk.[2,3]Less than 2% of the total AVMs involve pelvis; there are less than 60 cases described in the world literature [4 ,5,6].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 [1,3,7]. This case is reported for its rare incidence and uncommon location.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.*

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.[1,2,3] The majority of extracranial AVMs involve the head and neck, followed by the lower extremities and trunk.[2,3] Less than 2% of the total AVMs involve pelvis; there are less than 60 cases described in the world literature [4 ,5,6]. 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[1,3,7]. 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 of13cm. On examination 13 cm swelling was non tender and pulsatile. By applying pressure, thetumors could be emptied almost completely, but refilledrapidly. 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(figure 1). Microscopy showed AVM withvenules showing arteriolisation consisting of hyalinization and fibrotic changes. Arteries had thickened irregular wall with narrow lumen.(figure 2)

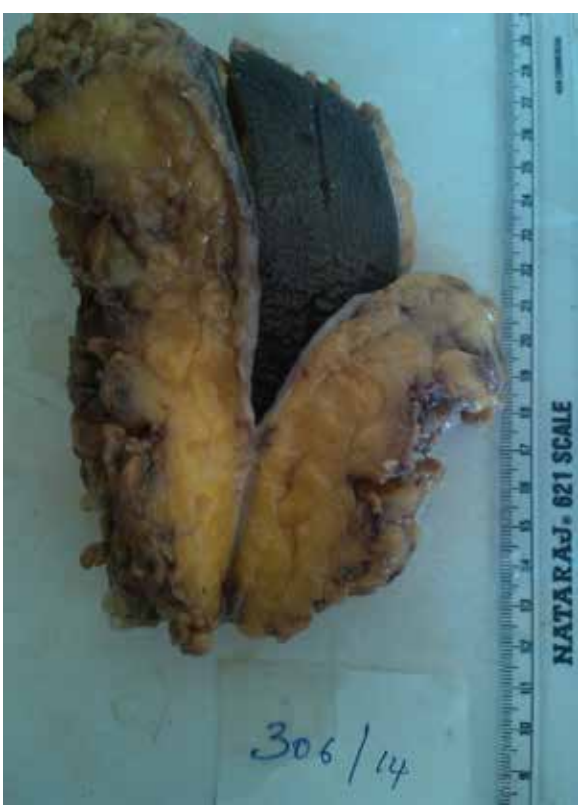

Figure 1. Photograph of cutsurface of AVM showing numerous blood vessels.

Volume : 5 | Issue : 4 | April 2015 | ISSN - 2249-555X

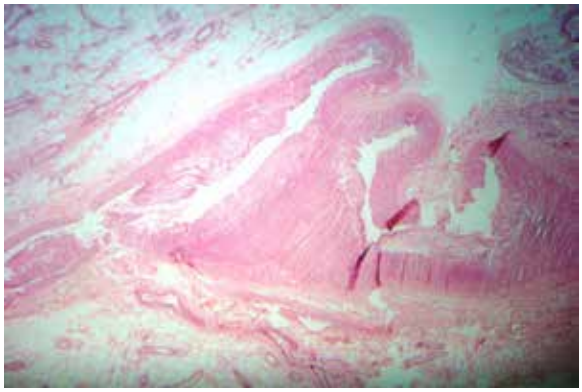

Figure 2.photomicrograph showing AVM consisting of numerous vessels with thickening, hyalinization and fibrotic changes.(hematoxylin and eosin stain, original magnification x100).

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.3,4,9Natalidescribed a woman who developed a pelvic AVM a year after trauma to her buttock. However, some authors believe that trauma simply calls to attention congenital lesions.Mekkes JR et al reported a case of arteriovenousanastomoses due to tissue repair and angiogenesis developed after ischemic tissue damage and subsequent necrosis in the deep subcutisinduced by prolonged pressure (decubitus).[4,9-11]

Most are diagnosed in the second and third decades of life.There is no reported gender difference. As described by Schobingernatural 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]

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, [3,14-17]Treatment depends onthe 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

1. Kohout MP, Hansen M, Pribaz JJ, Mulliken JB. Arteriovenous malformations of the head and neck: natural history and management. PlastReconstrSurg 1998;102:643-654. | 2. McCarron JA, Johnston DR, Hanna BG, Low DW, Meyer JS, Suchi M, Dormans JP. Evaluation and treatment of musculoskeletal vascular anomalies in children: an update and summary for orthopaedic surgeons. Univ Pennsylvania Orthopaedic J 2001;14:15- 24. | 3.Jui-Hung Chien ,Tzo-Peng Lee , Chih-Wei Wang , Shao-Liang Chen, Chih-Kung Lin. Intramuscular Arteriovenous Malformation of the Gluteus Maximus Muscle. J Med Sci 2010;30(5):225-229. | 4.Ledson MJ, Wahbi Z, Harris P, Walshaw MJ. A large pelvic arteriovenous malformation in an adult patient with cystic fibrosis. Postgrad Med J 1999;75:353-355. | 5.Szilagyi DE, Roger F, Smith MD et al. Congenital arteriovenous anomalies of the limbs. Arch Surg 1976;111:423–429. | 6. Rich, Spencer.Arteriovenous fistulae. in Vascular trauma. eds Rich NW, SpencerFC (WB Saunders and Co, Philadelphia), 1977. p 191. | 7.Wolf GT, Daniel F, Krause CJ, Kaufman RS.Intramuscular hemangioma of the head and neck. Laryn-229 Jui-Hung Chien, et al. goscope 1985;95:210-213. | 8.Vaisnyte B, Vajauskas D, Palionis D, Nevidomskyte D, Misonis N, Bilkis V. Complicated congenital gluteal arteriovenous malformation with hemorrhage in pregnancy. Ann VascSurg 2013;27(6):803. | 9.Mekkes JR, Pasch MC ,Meijs M, SillevisSmitt JH. Acquired arteriovenous malformation induced by pressure: a case report .Vascular Medicine 2003; 8: 201–202. | 10.Natali J, Jue-Denis P, Kieffer E, et al. Arteriovenous fistulae of the internal iliac vessels. J CardiovascSurg 1984;25:165–172. | 11.Mortensen JD, Ellsworth HS. Internal iliac arteriovenous fistula developing post partum. Am J Cardiol 1965;16:292–296. | 12..Decker DG, Fish CR, JuergensJC.Arteriovenous fistulas in the female pelvis.ObstetGynecol 1968; 31:799–805. | 13.Price AC, Coran AG, Mattern AL. Haemangioendothelioma of the pelvis: A cause of cardiac failure in the newborn. N Engl J Med 1972;286:647–649. | 14.Marler JJ, Mulliken JB. Current management of hemangiomas and vascular malformations.ClinPlastSurg 2005;32:99. | 15.Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. PlastReconstrSurg 1982;69:412-422. | 16. Meyer JS, Hoffer FA, Barnes PD, Mulliken JB. Biological classification of soft-tissue vascular anomalies: MR correlation. Am J Roentgenol 1991;157:559- 564. | 17.Lee BB, Do YS, Yakes W, Kim DI, Mattassi R, Hyon WS .Management of arteriovenous malformations: a multidisciplinary approach. Journal of Vascular Surgery 2004; 39(3):590–600. | 18.Katsimanis E, Jackson JE, Freeman RM. A major congenital vascular malformation of the pelvis in pregnancy. IJOG 2002 ;109:469–472. | 19.Beller U, Rosen J, Beckman M, Markoff G, Bernstein A. Congenital arteriovenous malformation of the female pelvis: a gynecologic perspective. Am J ObstetGynecol 1988;159:1153–1160. |