

Peripheral Congenital Arteriovenous Malformation - Report of Two Cases



Medical Science

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ABSTRACT

Congenital arteriovenous malformation is still a challenging clinical entity. It has varied clinical presentation and an unpredictable course. We present 2 cases of peripheral congenital AVM.

INTRODUCTION:

Arteriovenous malformation (AVM) encompass a variety of lesions. They are etiologically Congenital or Traumatic. A multidisciplinary approach is often required for their management.

CASE REPORT:

Case 1 :- A fifteen year old girl presented with history of swelling and pain during walking over the left knee of four years duration. No history of fever. There was no preceding trauma .An orthopedic surgeon had done an open synovial biopsy of the left knee a year ago and reported as Non –specific Synovitis. Clinical examination was unremarkable except for a longitudinal surgical scar (Fig.1) over the left knee joint .Computerized Tomography of the left lower limb (Fig.2) revealed numerous dilated tortuous veins in the **Vastus** lateralis without any identifiable feeding vessel. Surgery was done under Epidural anesthesia via a longitudinal incision over the surgical scar. The multiple vessels were ligated and resected . Post operative Doppler study (Fig. 3) did not reveal any residual lesion. The patient was followed up for 5 years and was lesion and symptom free.

Case 2 :- A three year old girl weighing twelve kilograms presented with history of swelling in the right calf of one year duration and local pain while walking since six months. Clinical examination showed a swelling in the upper half of the right calf with ill-defined borders. The approximate size was 6 cms by 4 cms. The swelling was non –tender and there was no change in the size of the swelling on walking. Magnetic Resonance Angiogram (Fig.4) displayed an ill – defined lesion with delayed enhancement of multiple , tortuous veins in the subcutaneous plane and lateral head of right Gastrocnemius , communicating with the Short Saphenous Vein. There was no immediate post-contrast enhancement and no skeletal abnormality noted. Surgery was done under general anesthesia via a longitudinal incision over the lateral aspect of right calf . The multiple tortuous veins were ligated and resected. Post- operative Doppler (Fig.5) did not demonstrate any residual lesion. The child has been followed up for 5 months and is doing well.

DISCUSSION :-

Congenital AVM is classified into Truncular and Extratruncular lesion according to Hamburg classification¹. The former is embryonal dysplasia of differentiated vascular trunk and involves central axial vessels. The two types are superficial and deep. The Extratruncular type is rare and derived from the remnant of primitive capillary network and involves peripheral vessels. The two types are infiltrating and limited lesion. The infiltrating lesion has diffuse involvement; the limited lesion has highly tortuous and ectatic dysplastic vessels.

Angiographic classification² of AVM into 3 groups is based on the flow characteristics. Group1 :- Predominantly arterial or arteriovenous lesions.

Group 2:- Predominantly capillary lesions.

Group 3 :- Predominantly venous lesions.

The primary effect on the surrounding is by the lesion itself with compression and erosion. The secondary hemodynamic effects could be due to a steal phenomenon (ischemia , gangrene) or venous hypertension (stasis dermatitis , ulcer , gangrene). They retain the evolutionary potential to grow and this is often represented clinically as recurrence³. Their behavior is therefore unpredictable and growth can be accelerated by stimulations such as injury, surgery and systemic hormone effect. Recurrence and unbridled growth are their trademarks³.

The symptoms could be pain, cosmetic disfigurement , bleeding , limb length discrepancy, high output cardiac failure or even psychiatric problems. Duplex ultrasonography is valuable as an initial diagnostic and follow- up tool. Magnetic Resonance Imaging is the gold standard for assessing the anatomic status. Ablative surgery with or without embolo/sclero therapy is the only option for potential cure⁴.

Hemangiomas differ from AVM in that they are :

1. Neoplasms
2. Encapsulated
3. Have a cellular stroma and show endothelial proliferation.
4. Grow in tissue culture.



Fig.1 Surgical scar of previous open synovial biopsy of left knee joint

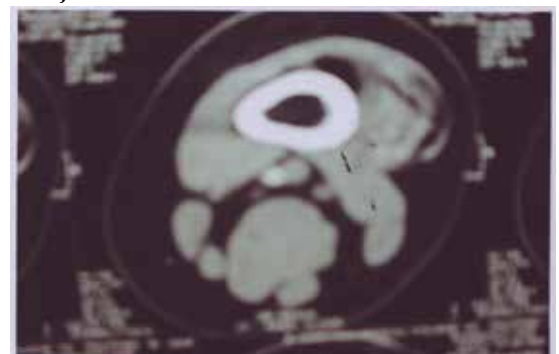


Fig.2 Computerized Tomography of the left limb showing dilated tortuous veins in the left Vastus Lateralis

