Original Articles

Vascular anomalies - diagnosis and therapy

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ABSTRACT

Objective: Vascular anomalies were once thought to be impossible to properly diagnose and treat. Hence, we aimed to evaluate the different diagnostic and therapeutic modalities in the management of vascular anomalies.

Methods: We carried out a retrospective review of our experience to evaluate different diagnostic and therapeutic modalities in the management of 25 patients with vascular anomalies over a 2-year-period at King Khalid University Hospital, Riyadh, Kingdom of Saudi Arabia and follow-up period ranging from 2 months to 2 years.

Results: Vascular anomalies were more common in male patients (N=19). Age range was 7 to 46 years. Vascular anomalies were categorized as hemangioma (N=2) or malformation (N=23). The vascular malformation were further subdivided into slow flow (N=5) and fast flow (N=18). Duplex (N=12) and radiographic studies; angiography (N=21), venography (N=7), computerized tomography (N=10) and magnetic resonnance angiography (N=8) were used to confirm diagnosis. The treatment of hemangiomas were surgical resection (N=1) and conservative treatment (N=1). Embolization was the main modality of treatment in vascular malformation (N=16), with surgical resection in 4 patients, sclerotherapy in one and conservative in the other 2. All cases had successful outcome with no complications.

Conclusion: Control of large vascular malformations with acceptable results can be achieved nowadays. Intra-arterial embolization is the mainstay of treatment and long term follow-up with serial physical examination, duplex and arteriography is required.

Keywords: Vascular anomalies, arteriovenous malformation, hemangioma, selective embolization.

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Inspite of rapid advances in surgery, particularly in vascular surgery, vascular anomalies remain a difficult surgical problem. How should vascular anomalies be classified? How should they be evaluated? Are the surgical outcomes predictable? What are the practical guidelines for treatment? Unfortunately, no large series of vascular anomalies is available. Hemangioma was the most common terminology used for diagnosis of vascular anomalies.[1] Great progress in classifying and understanding vascular anomalies has been made on the basis of cellular, kinetic and clinical behavior.[2] They are classified into 2 major categories: hemangioma and vascular malformation.[2,3] Hemangiomas are benign tumors of infancy characterized histologically by endothelial hyperplasia and clinically by a rapid proliferating phase lasting 8 to 10 months followed by slow, spontaneous involution over a period of several years.[4] In contrast, vascular malformations are dysplastic vessels with normal endothelial behavior, they remain stable or slowly grow and typically require some form of therapy when cosmetic disfigurement, bleeding or functional impairment occur.[4,5] We reviewed patients admitted to our hospital with diagnosis of vascular anomalies over a

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period of 2 years with emphasis on clinical presentation, usefulness of a biologic classification systems and different diagnostic and therapeutic modalities.

Methods. The medical records of 25 patients with diagnosis of vascular anomalies in the period between January 1999 to February 2001 were reviewed. The collected data included: 1) patient's age first time the anomaly was noted; 2) presenting symptoms; 3) clinical course; 4) investigations performed; 5) management and; 6) outcome. Hemangiomas were distinguished by their biphasic growth and spontaneous involution while vascular malformation did not involute and were subdivided into capillary malformation (CM), venous malformation (VM), lymphatic malformation (LM), arteriovenous malformation (AVM) or combined. These malformations were categorized as slow flow (CM, VM, LM) or fast flow lesions (arterial). Slow flow lesions were painless, compressible masses while fast flow lesions were not easily compressible and were warm with palpable thrills and bruits.

Results. Vascular anomalies were more common in male patients (male:female-19:6). Age range was 7 to 46 years (mean age ± 26). Vascular anomalies were categorized as hemangiomas (N=2) and vascular malformation (N=23). The malformation were further sub-divided into AVM (N=18) which were the most common type, comprising more than two thirds of all vascular malformation, VM (N=3) and CM (N=2). Fast flow AVM (N=18) were 3-4 times as common as slow-flow lesions (N=5). Although abnormal vascular channels are presumably present at birth, not all malformations were congenital. Fifteen out of 23 (87%) were present at childhood while 8 manifested at birth. History of trauma which triggers appearance of malformations were noticed in 9 cases while history of previous biopsy or attempted surgical excision were obtained in malformation of extremities (N=3), face and neck (N=2) and abdomen (N=1). Anatomical sites of vascular anomalies were commonly seen in extremities (N=17) with no spared region, **Table 1**. Most common presenting symptoms were diffuse swelling (N=17), localized mass (N=6), pain with exercise and warmth sensation (N=4), and dilated vessels (N=5). All fast flow AVM evolved with prominent thrills and bruits and one case (AVM) presented with limb hypertrophy and lengthening. Diagnostic studies were occasionally used when the diagnosis was in doubt. Color duplex (N=12) was used to confirm the presence of fast flow anomalies and to separate hemangioma from malformation. Computerized tomography (CT) scan (N=10) and magnetic resonance imaging (MRI) (N=8) were used to localize the vascular malformations in relation to involved tissues. Diagnostic angiography with super selective catheter immobilization using evalon particles, sponge or coils was the main line of treatment in fast flow AVMs (N=16) **Figure 1.** One hemangioma and 2 VM had conservative management. Compression garment was used for extremity VMs (N=2). Sclerotherapy with ethenal 100% was applied to one lesion (VM) located in the right side of the neck. A satisfactory result has been achieved after 5 sessions of sclerotherapy with no complication. Surgical excision was the treatment for localized vascular anomalies: hemangioma (N=1), fast flow AVM (N=2) and slow flow vascular malformation (N=2).

Discussion. A classification system for any disorder is useful only if it has diagnostic applicability and helps in patient's management.[2,4] Vascular anomalies are classified on the basis of history and physical examination. Rapid growth phase followed by spontaneous involution differentiate hemangiomas from malformations in over 96% of all vascular anomalies.[5] The only surgery that is necessary for children with

Table 1 - Anatomical sites of vascular anomalies.

Site of lesion	Patients N
Lower limb	10
Upper limb	7
Face and neck	7
Abdomen	1
N - number	

Table 2 - Schobinger clinical staging system for arteriovenous malformations.

Stage	Description
I. Quiescence	Pink-bluish stain, warmth and arteriovascular shunting by continuous Doppler scanning or 20-MHZ color Doppler scanning.
II. Expansion	Same as stage 1 plus enlargement, pulsations, thrills and bruit and tortous/tense veins.
III. Destruction	Same as Stage II plus either dystrophic skin changes, ulceration, bleeding, persistant pain or tissue necrosis.
IV. Decompensation	Same as Stage III plus cardiac failure.

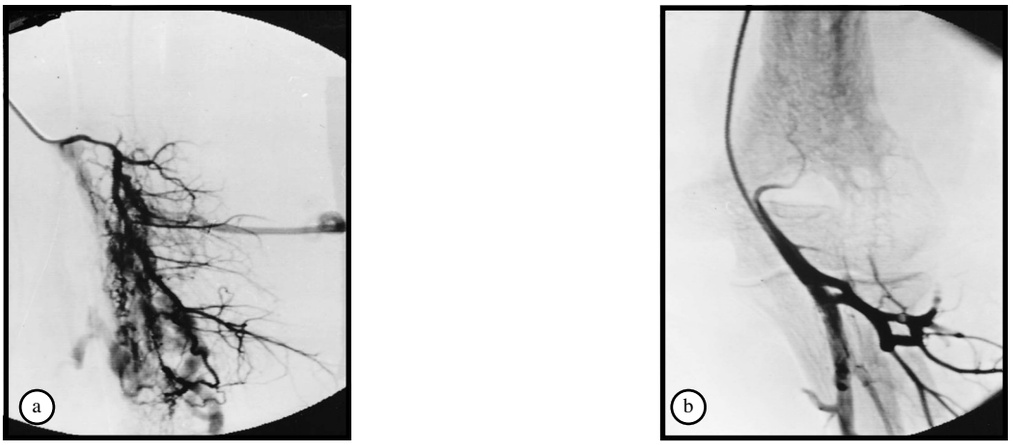

Figure 1 - Angiogram showing **a)** left upper limb arteriovenous malformation pre-embolization **b)** disappearance of arteriovenous malformation post-embolization.

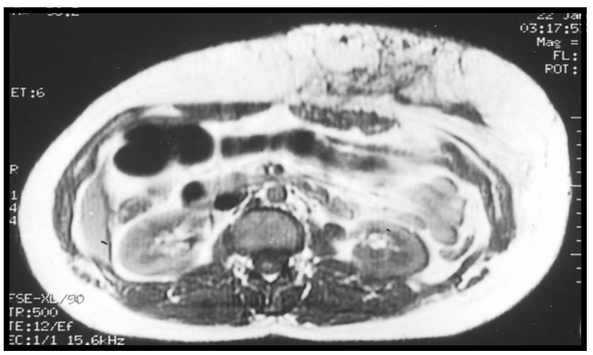

Figure 2 - Magnetic resonance imaging showing extensive arteriovenous malformation of left site abdominal wall.

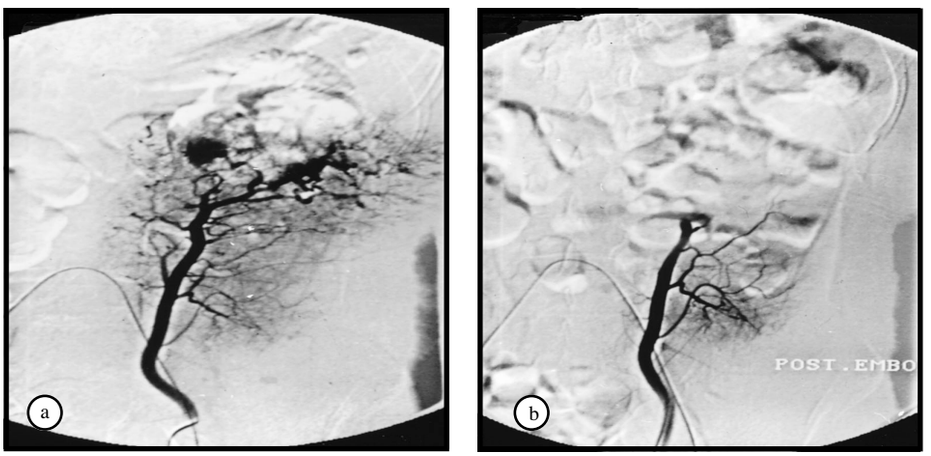

Figure 3 - Angiogram showing **a)** pre-embolization - feeding artery to arteriovenous malformation inferior epigastric artery. **b)** post-embolization with occlusion of inferior epigastric artery.

hemangiomas is excision of fibrofatty residue after involution is complete.[6] Malformations are categorized as CM, VM, lymphatic combined or arterial.[6,7] They are also divided into slow-flow (CM, VM, LM) and fast-flow (AVM) subgroups.[2,3] Arteriovenous malformations in the head and neck region are the most common followed in frequency by AVMs of the limbs, trunk and viscera.[8] In our series, vascular malformations were more common in limbs. Arteriovenous malformations are unusually noted at birth due to their innocent appearance. Sometimes puberty or trauma seem to trigger expansion.[6] An extensive AVM, either in a limb or the pelvis, can cause increased cardiac output. The natural history of AVMs can be documented by a clinical staging system introduced by Schobinger, **Table 2**.[8] Twenty five percent of our cases were misdiagnosed by physician based on physical examination. Virtually 100% of these malformations referred to us were correctly diagnosed by their natural history, clinical examination, duplex, and MRI. The extent of a vascular malformation is usually under estimated by physical examination. Magnetic resonance imaging and CT are used for an accurate representation of the size and involvement of contiguous structure.[7,9,10] Color duplex scanning is simple and documents the size of the anomaly and its blood flow characteristics.[4,11] Complex vascular malformations sometimes associated with the overgrowth of soft tissues and the skeleton.[1] Plain radiographs are useful in these conditions to assess skeletal lengthening as seen in one of our cases with AVM of the upper limb. The indications for treatment of vascular malformations are either progressive symptomatology or presence of complication, otherwise, conservative treatment is the choice. Surgical excision is applied for localized lesion of vascular malformation. Malformations are not composed of actively proliferating endothelial cells. Enlargement of new areas following surgery is a reflection of redirection of flow into previously uninvolved anomalous channels within malformation.[2,3] That was the explanation for one case presented to us with recurrent diffuse abdominal wall AVM after 2 attempts at excision **Figure 2**. Angiography is no longer used for diagnostic purposes alone. However, angiography combined with super selective immobilization is the mainstay of treatment in AVM as many of these lesions are not localized and surgical resection not feasible.[12] In our series, all fast-flow lesions have been treated by repeated sessions of super selective catheter embolization using particles or coils. Sclerotherapy has a role in the treatment of venous malformation.[13,14] These injections directed into the lesion must be repeated for control of large diffuse lesions. Absolute ethanol (100%) is commonly used in the Unites States of America (USA),[13] whereas ethibloc, a mixture of zein (a corn protein), alcohol, and contrast medium, is used outside the USA.[14] Sclerotherapy is potentially dangerous and requires the skills of an experienced radiologist. Local complications include blistering, full thickness cutaneous necrosis, or damage to local nerves.[13] A good result has been achieved in one of our cases with diffuse VM of neck after 5 sessions of local ethanol injections. Despite advances in the field of angiogenesis, we don't have a pharmacologic therapy for vascular malformation.[15] However, anti-angiogenic drugs alpha 2 interferon and steroids are used for hemangiomas.[16] Extensive vascular malformations can exhibit a chronic form of consumption coagulopathy characterized by depressed platelet counts, prolonged prothrombin time and low fibrinogen.[17]

In conclusion, intra-arterial embolization is the mainstay of treatment for vascular malformation. However, long term folow-up with repeated sessions of embolization is required. To achieve the proper treatment, the vascular anomalies management program should include; surgeon (vascular and plastic), radiologist and dermatologist.

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