ARTERIOVENOUS MALFORMATIONS

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Cutaneous arteriovenous malformations (AVMs) are fast-flow malformations as opposed to low-flow venous, capillary and lymphatic malformations. They consist of a connection between arteries and veins without any interpositional capillary bed, thus creating a „nidus” or shunt. They can be congenital or acquired. If acquired, they often occur after a trauma. If congenital, they are usually latent for many years and may be exacerbated at puberty or after a trauma or therapeutic procedure, even if minimal.

From a clinical viewpoint, the lesions may be small, such as located on a finger (1) or the nose. They involve a superficial telangiectatic vascular network and sometimes minimal swelling. The

Summary

Cutaneous arteriovenous malformations (AVMs) are fast-flow malformations. They consist of a connection between arteries and veins. The can be congenital or acquired.

The lesions may be small to large and aggressive. There are many particular clinical forms
- Stewart-Fluefarb
- Bonnet-Blanc Deschaumes (or Wyrburn – Mason)
- Capillary malformations –AVM syndrome with RASA-1 mutation
- Parkes-Weber syndrome
- Congenital lipomatous overgrowth vascular malformations and epidermal nevi (CLOVE) syndrome
- Cobb syndrome

AVM can be complicated by pain, ulceration, haemorrhage and necrosis. Evolution has 4 stages (Schrobringer classification).

Diagnosis is based particularly on two exams, doppler ultrasonography and MRI.

Treatment for voluminous forms used embolization, surgery, and new medical treatments with some anti VEGF drugs.

Key words: Angioma, malformations, arteriovenous malformations.
lesion may be located on a limb (fig 1) or on the face (fig 2-3) with various spreading. Sometimes the only features are superficial capillary malformations. The local temperature may be increased. Palpation can reveal a pulse. In contrast, large defects can affect any part of the face or a limb, with large swelling, superficial venous network of large diameter, and increased local heat. This lesion may be stable or may be the starting point of a large aggressive malformation. The evolution from this growth phase can be particularly aggressive, with haemorrhage, ulceration, and necrosis.

**Clinical forms of AVMs**

- **Stewart-Bluefarbsyndrome** is a purplish swelling tumor mimicking Kaposi’s sarcoma. The lesions may become necrotic and infected.
- **Bonnet-Blanc-Dechaume or Wyburn-Mason syndrome** stain consists of a forehead capillary malformation associated with an underlying AVM of the brain.

**Evolution**

AVM lesions can increase in volume. They can be complicated by pain, ulceration, haemorrhage, and necrosis. Heart failure is possible. AVM may rapidly progress during puberty or pregnancy. Trauma and infections are also factors of progression. Evolution shas 4 stages (Schobinger...
(classification): stage 1, quiescence with macular lesions; stage 2, expansion, enlarging lesions, palpable pulse; stage 3, infiltration; stage 4, destruction, necrosis, heart failure (fig 4).

Pathophysiology

AVMs are due to failure of complete involution of the fetal capillary bed. This situation leads to connections between arteries and veins. Most AVMs do not have a hereditary family character of transmission. The discovery of the mutation in RASA-1 has introduced an element of complexity with the realization of familial forms, but clinical manifestations are varied in the same family (9) (6) (5) (4).

In contrast, somatic mutations are sometimes identified on biopsy, in particular mutation in the PI3K gene (14) in the same pathway as the Proteus syndrome gene (AKT) (14). Somatic mutations play an important role in many AVMs. DNA libraries of tissues and circulating blood samples must be collected for future investigations of these malformations.

Diagnosis

The diagnosis is made on clinical examination: clinical features, location, and stage. Some areas such as the ear are more at risk. Local heat, which increases the hypertrophy of the auricle (15), palpation of pulse or thrill require exploration. Doppler ultrasonography allows for visualizing increased arteriovenous flow. MRI can visualize the enlarged superficial tissues, important for visualizing depth. Gadolinium injection allows for better visualization of vessels. MRI angiography can reveal the nidus squads that are vascular arteries and veins corresponding to extension zone angiomas.

Treatment

For quiescent, minor forms of AVMs, monitoring only is the rule; any action may lead to exacerbation, so the treatment should be conservative. Embolization can be performed in case of progression or complications.

For voluminous forms, the definitive treatment, when possible, is complete surgical wide excision of the nidus. This surgery may be difficult or impossible because of the location or volume of the AVM. It sometimes requires rapid intervention for heavy bleeding or rapid extension (16). Amputation may be necessary. Embolization is useful immediately preoperatively to improve intraoperative hemostasis. Embolization or ligation of vessels without subsequent surgery are of little use for large AVMs, and often a reversal in condition is fast and exacerbation may even succeed embolization.

Medical treatment

The aim is to decrease the action of growth factors (vascular endothelial growth factor [VEGF]). The immunosuppressant sirolimus can be tried as can some anti-VEGF drugs with or without embolization or surgery. Clinical trials are underway.

Bibliography


Conflicts of interest


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NONE DECLARED