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Arteriovenous malformations of the head and neck: current concepts in management-Review

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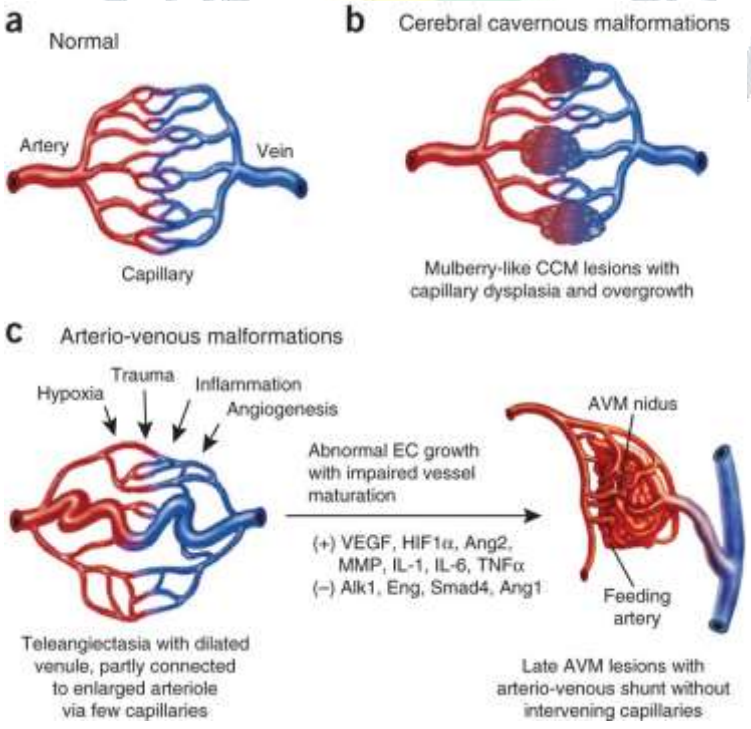
INTRODUCTION

AV malformations are the most dangerous type of vascular malformation and considered the most challenging to manage. Recurrence rates of over 80% after embolisation and surgical resection have been reported even after the recent advances and improved multidisciplinary approaches. The traditional capillary network between the arteries and veins is usually lacking or bypassed in arteriovascular malformations. Direct shunts cause a lack of normal down-regulation in pressure and have the potential to increase and recruit new vessels. The central section of the abnormal connection and the first section of the adjoining dilated vein termed the nidus.



CLASSIFICATION

Cho et al. modification of the Houdart et al. classification of arteriovenous malformations based on the number of arterial and venous communications and niduses.

Type	Description
I	No more than 3 arterioles shunting to initial part of venous component
II	Multiple arterioles shunt to initial part of venous component, with arterioles showing a plexiform appearance
IIIa	Multiple shunts between arterioles and multiple non-dilated venules
IIIb	Multiple shunts between arterioles and multiple dilated venules

Clinically they may present with pain,tissue expansion and destruction, ulceration, disfigurement, bleeding, warm, often pulsatile with the presence of a thrill.

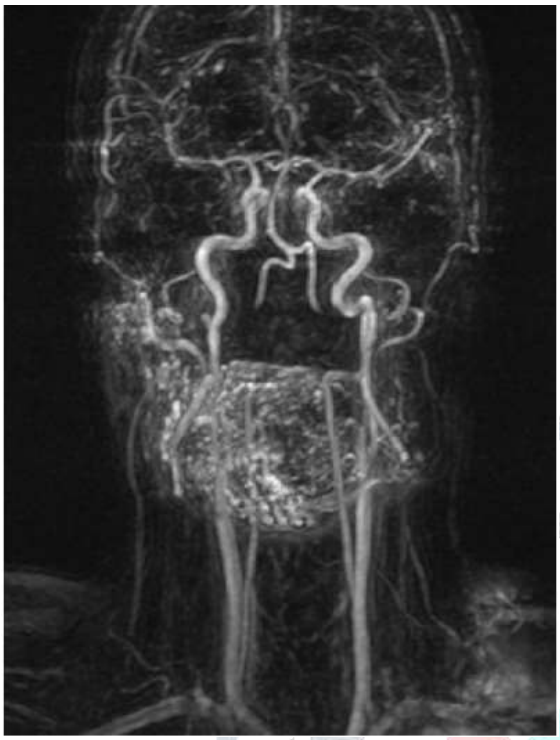
Schobinger modification of the Houdart classification of high-flow arteriovenous malformations.[7]

Stage	Description
1	Quiescent: stable
2	Increasing in size
3	Symptomatic: pain, bleeding, disfigurement, disruption of function
4	Decompensating: high-output cardiac failure

IMAGING AND DIAGNOSIS

- Ultrasound.
- Magnetic resonance imaging (MRI) – shows their size, shape, and association with neighbouring tissues.

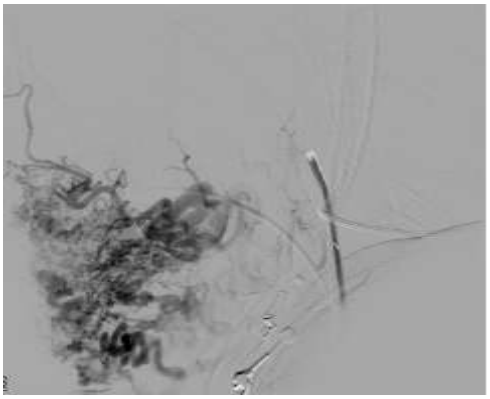
- Angiography - typically shows dilated and tortuous feeding arteries, arteriovenous shunts, and greatly dilated draining veins and to identify the detailed vascular anatomy.



Magnetic resonance angiogram of an arteriovenous malformation of the lower lip and chin showing extensive vascular supply.

EMBOLISATION - TANTALUM(ONYX®)

- Onyx® is a liquid embolic agent that is dissolved in dimethyl sulphoxide (DMSO) and opacified by tantalum powder.
- It became popular as a neurovascular embolic agent, and has recently been used in peripheral lesions.
- On contact with blood, the DMSO diffuses out of the mixture and leaves the ethyl vinyl alcohol to polymerise.
- Polymerisation first occurs peripherally within the mixture, and over a few minutes gradually hardens towards the centre of the plug. This extends the working time and allows more time for the injection.
- Once hardened, Onyx® leaves a rigid cast that plugs the nidus permanently, and gives a characteristic appearance on future imaging .



Lateral view of embolisation showing Onyx®in some of the niduses of the arteriovenous malformation.

EMBOLISATION – PHILTM

PHILTM(precipitating hydrophobic injectable liquid) is a new embolic agent that is being used in the endovascular management of cerebral neurovascular diseases.

Three concentrations are available, with various viscosities and different flow characteristics that lead to deeper penetration of the nidus. As the material looks white on clinical inspection, the risk of staining is minimised.
The benefits of PHILTM -in the management of extracranial arteriovenous malformations, with successful endovascular embolisation and subsequent resection. Like Onyx®, PHILTM also uses the solvent DMSO, and therefore has the same risk of perivascular inflammation and tissue damage, but it also has the less dangerous side-effect of a garlic-like odour from the breath and skin of patients. Patients may also report a garlic-like taste.

Surgical intervention

Early operation has been recommended to limit the destruction caused by the continuous growth of lesions.
Complete resection should be the aim, and partial excision avoided where possible. Well-localised lesions without previous embolisation are therefore ideal, as they are likely to have smaller feeder vessels and well-defined borders. They also have a better chance of cure, and there is less likelihood of intraoperative haemorrhage.
Partial resection can acutely worsen the disease due to re growth.
Subtotal resection can, however, be justified in certain cases, and indications include life-threatening haemorrhage, serious functional and cosmetic deformity, and continued growth despite previous, alternative interventions.resection after embolisation has become the most accepted treatment in case of high-flow lesions .
Preoperative embolisation can help delineate the extent of a lesion and dramatically reduce intraoperative bleeding, and multiple embolisations before operation have been found to lessen the risk of recurrence.

RE-EXPANSIONAND RECURRENCE

Embolic and surgical treatments can stimulate recurrence through the creation of a proangiogenic environment. Embolisation leads to local hypoxia and increased levels of VEGF, MMP-2, and MMP-9. Operation and healing also induce local hypoxia and inflammation, which are known stimulants of angiogenesis through VEGF, basic fibroblast growth factor, and MMP. To reduce hypoxia and recurrence after resection, several authors have proposed reconstruction with free-tissue transfer.

NEW TREATMENTS

Antiangiogenic drugs- Marimastat is an antiangiogenic MMP inhibitor, initially developed for the treatment of metastatic breast cancer and also used successfully in themanagement of a progressive lesion in the limb of a child.

Bevacizumab**,** a humanised anti-VEGF anti-body, has shown promising results in early clinical trials tocontrol nosebleeds and arteriovenous malformations of the liver. Side effects include hypertension, severe proteinuria, decreased wound healing, and gastrointestinal bleeding, and its effect on cutaneous and cerebral lesions is as yet unknown.

CONCLUSION

Multidisciplinary treatment is required to achieve an optimal outcome in the management of arteriovenous malforma-tions. Preoperative intravascular embolisation with excision is recognised as the treatment of choice, and earlier intervention is now deemed desirable**.**

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