Cavernous malformation

- Cellular Heterogeneity and Tissue Specificity in Venous Malformations: Implications for Pathogenesis and Targeted Therapies
- Persistent trigeminal artery variant functioning as a duplicate superior cerebellar artery
- Surgical treatment to resect giant intraspinal epidural cavernous hemangioma of Cobb syndrome: illustrative case
- Comparative outcomes of surgical and conservative management of pediatric intracranial cavernous malformations: a systematic review and meta-analysis
- The parapharyngeal vein-an accessory communication between the middle cerebral veins and the internal jugular vein: a case report
- QSM predicts haemorrhage risk in brainstem cavernous malformations: a multicentre prospective study
- Cavernous Hemangioma of the Anal Canal Mimicking a Neoplasm: A Rare Cause of Lower Gastrointestinal Bleeding Successfully Treated With Transanal Excision
- Single-cell transcriptome profiling reveals dynamic cell populations and immune infiltration in cerebral cavernous malformation

Cavernous malformations are clusters of abnormally formed blood vessels with thin walls, resembling a small raspberry. They can cause bleeding in the brain, leading to neurological symptoms, seizures, or headaches.

In 1928, Cushing and Bailey classified cavernous malformations as a solid subtype of hemangioblastomas [1)]

General information

Key concepts

usually angiographically occult. May show up on MRI (open channels → flow void on T2 weighted image, previous hemorrhage →“popcorn” pattern especially on T2* gradient echo) or contrast CT

● low-flow. No intervening neural parenchyma, no arteries. Associated with venous anomaly (represents venous outflow and should be preserved)

● XRT is a risk factor for developing cavernous malformation

● presentation: usually seizures:

The 5-year risk of first-time seizure was 6% among CM patients presenting with symptoms vs. 4% for those with incidental CMs

● Hemorrhage: rare, the risk is difficult to predict (see Cavernous malformation hemorrhage)

● treatment:

a) surgery is the treatment of choice for symptomatic accessible lesions

b) radiosurgery should not be considered as a treatment option

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Cavernous angioma (CA) is also known as cavernoma, cavernous hemangioma, and cerebral cavernous malformation (CCM) (National Library of Medicine Medical Subject heading unique ID D006392).

It is classified as a group of vascular malformations, where a collection of dilated blood vessels form a tumor.

Hubert von Luschka was the first to describe a cerebral (Cavernöse Blutgeschwulst) (cavernous malformation) in 1854.

Synonyms

The terms cavernoma, cavernous hemangioma, cavernous angioma, and cavernous malformation are used interchangeably and appear equally frequent in the literature. The terms cavernoma, cavernous angioma, and cavernous hemangioma suggest that it is a neoplastic lesion, which is inaccurate as cavernous malformations are true vascular malformations.

The term cavernous hemangioma is also confusing as cavernous hemangiomas in the cavernous sinus or in the orbit; both are slow-growing neoplastic diseases unrelated to vascular malformation

see Cavernous sinus hemangioma

Classification

Cavernous Malformation Classification.

Etiology

see Cavernous malformation etiology.

Pathology

Cavernous malformation (CCM)s are benign, low-flow vascular lesions with thin elastic endothelial walls that lack adventitial smooth muscle and frequently present with little intervening brain parenchyma.

Microscopically, cavernous angiomas consist of sclerotic, variably calcified compact vessels in a honeycomb pattern.

Unlike telangiectasis and arteriovenous malformation these lesions do not contain interstitial parenchyma.

The cells that form the vessels do not form the necessary junctions with surrounding cells and the structural support from the smooth muscle is hindered causing leakage into the surrounding tissue.

Clinical features

It is the leakage of blood, known as a hemorrhage from these vessels that causes a variety of symptoms known to be associated with this disease.

Diagnosis

Cavernous malformation diagnosis.

Differential Diagnosis

Cavernous malformation differential diagnosis.

Treatment

see Cavernous malformation treatment.

Complications

Cavernous malformation hemorrhage

Outcome

When cavernous malformations can be completely removed, the risk of further growth or hemorrhage is essentially permanently eliminated [2] (however, recurrence of symptoms has been reported after partial and even seemingly-complete removal [3] [4]. For CMs treated surgically, patients need to be aware that post-op neurologic worsening is very common, especially with brainstem CMs [5] Worsening may be transient, [6] but may take months to resolve.

Herten et al. performed a cross-sectional observational study on patients with cavernous malformation of the CNS (CCM). admitted to the University Hospital Essen. from 01.11.2017-10.01.2020 using standardized interviews (short-form -36 questionnaire, hospital anxiety

and depression score (HADS-A/D), CCM-perception questionnaire). Including criteria was diagnosis of an untreated CCM and information about the diagnosis in a specialized CCM consultation. HRQOL data was analyzed and compared to German normal population. Uni- and multivariate analyses were carried out to identify variables with impact on outcome.

219 (93%) of 229 eligible patients were included. Mean age was 46.3 ±14.7 (18-86) years, 136 (62%) were female. 98 (45%) patients presented with symptomatic hemorrhage and 17 (8%) with repetitive SH. 92 (42%) patients were asymptomatic. 37 (17%) suffered from cavernoma-related epilepsy. 28 patients (13%) suffered from familial CCM. Patients showed significantly decreased component scores and subdomain scores compared to normal population with effects ranging from small to large. This accounted largely also for asymptomatic patients (except for physical component score and main physical subdomains). Multivariate regression analysis confirmed impact of functional impairment on physical component score. HADS-A was significantly increased. HADS-A/D strongly correlated with mental component score and individual perception of the CCM.

Patients with the diagnosis of a CCM showed decreased HRQOL compared to normal population even when not suffering functional impairment or neurological symptoms. This data may function as benchmark in evaluation of different (future) management strategies [7].

References

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