Caroticocavernous Fistula (CCF)

Forhad H. Chowdhury, Shamshul Alam, Nazmin Ahmed and Mohammod Raziul Haque

Abstract: A carotid cavernous fistula (CCF) is a relatively common intracranial arteriovenous fistula, usually traumatic and spontaneous in origin. A CCF may be of direct or indirect variety, and clinically presents with pulsatile proptosis, chemosis and red eye. Investigations include a CT of the head, an MRI of the head, and a CTA, MRA and DSA of the brain. Endovascular therapy comes first in line, but microsurgical treatment is needed where endovascular therapy is not possible or fails. Here, the etiopathogenesis, classification and management of a CCF are discussed concisely.

Abbreviations

AV	Arteriovenous	CCF	caroticocavernous fistula
CT	computed tomography	CTA	computed tomographic angiogram
CS	cavernous sinus	DAVF	dural arteriovenous fistula
DSA	digital subtraction angiography	ECA	external carotid artery
ICA	internal carotid artery	IPS	inferior petrosal sinus
MRA	magnetic resonance angiogram	MRI	magnetic resonance imaging
RTA	road traffic accident		

1. Introduction

A carotid cavernous fistula (CCF) is commonly traumatic in origin, following road traffic accidents (RTAs) due to the avulsion or tear of the cavernous carotid artery. Hence, blood goes to the cavernous sinus (CS) of the same side, followed by the opposite CS and the superior ophthalmic vein (Greenberg 2010).

2. Anatomy of the Cavernous Sinus

The CS is a complicated venous region that runs from the sphenoid bone to the periosteal, as well as the meningeal layers of the dura. On both sides of the sellae turcica, there is a twin venous space. The superior and inferior intercavernous sinuses provide open communication between the two regions. From the superior orbital fissure (SOF) to the petrous apex, the CS extends anteriorly. The diaphragm sellae are located cranially, and the larger wing of the sphenoid is located caudally. The dura forms a lateral boundary for the sinus (Figure 1). The ICA, with its periarterial sympathetic plexus, is located medially within the CS, and its involvement may produce Horner's syndrome. The sixth nerve is located to the lateral of the ICA. The ophthalmic and maxillary divisions of the trigeminal nerve, oculomotor nerve and trochlear nerve are located within the lateral dural boundary of the CS. Chemosis and proptosis can occur when venous drainage is compromised. The venous linkages in the CS are complicated and valveless. It communicates with practically every key venous component in the head and neck, either directly or indirectly. The superior and inferior ophthalmic veins, the sphenoparietal sinuses, and the middle meningeal vein all drain the CS. The superior and inferior ophthalmic veins connect the CS to the facial vein and pterygoid venous plexus. The CS drains into the superior and inferior petrosal sinuses, which then flow into the sigmoid sinus and internal jugular vein (Chowdhury et al. 2012; Tang et al. 2010; Barrow et al. 1985).

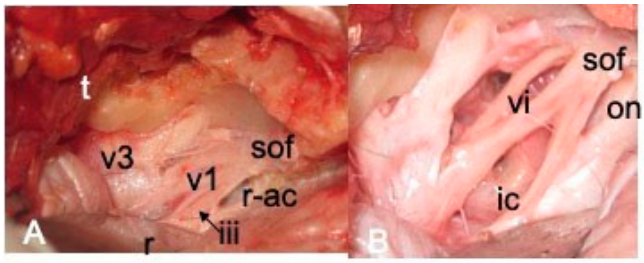

Figure 1. (**A**,**B**) Cadaveric dissection of the left CS after peeling of the dura from the CS with retraction of the temporal lobe extraduraly. (r—retractor, t—temporalis muscle, V3—mandibular nerve, V1—ophthalmic nerve, sof—superior orbital fissure, iii—oculomotor nerve, r-ac—root of anterior clinoid process (after drilling), vi—abducent nerve, ic—internal carotid artery and on—optic nerve). Source: Figure by authors.

3. Classification of the CCF

The CCF is generally classified depending on the arterial supply (Table 1, Figure 2). The clinical features and management approach are, however, mainly based on venous drainage.

Table 1. Barrow Classification.

Type	Pathogenesis	Arterial Supply	Hemodynamics
A	Head trauma/aneurysm rupture	ICA	High flow
B	Spontaneous	Dural branches of the ICA	Low flow
C	Spontaneous	Dural branches of the ECA	Low flow
D	Spontaneous	Dural branches of the ICA and ECA	Low flow

ICA: internal carotid artery, ECA: external carotid artery. Source: Authors' compilation based on data from Barrow et al. 1985; Cruz 1998.

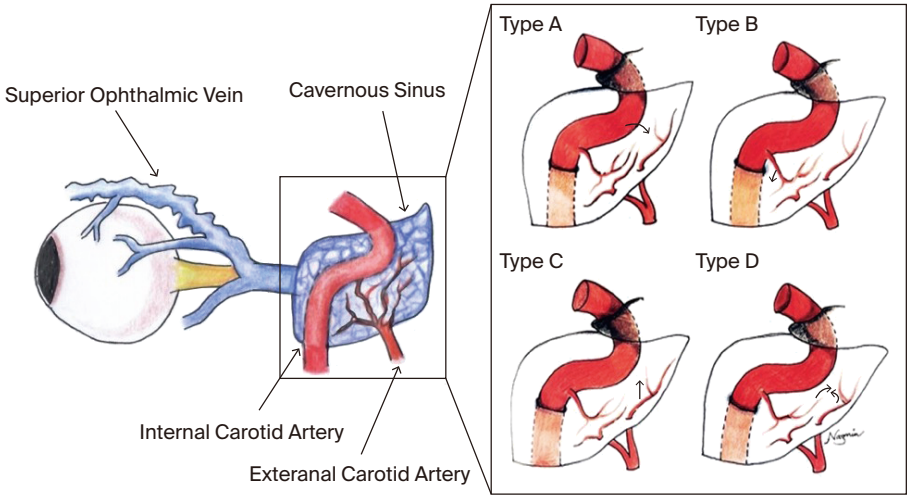

Figure 2. Types of carotid cavernous fistulas. Source: Figure by authors.

4. Pathophysiology

The underpinning pathophysiology (Greenberg 2010; Korkmazer et al. 2013; Ertl et al. 2019) for the clinical consequences of all caroticocavernous fistulas (CCFs) is characterized by elevated intracavernous venous sinus pressure. This is also likely to lead to retrograde venous drainage to the eye by the superior ophthalmic vein. Because of the interconnections between the two cavernous sinuses, contralateral eye involvement is common. Occasionally, due to occlusion of the superior ophthalmic vein by anterior intracavernous thrombosis, the clinical presentation may only occur contralateral to the fistula.

5. Natural History of CCFs

The natural history for the eye and vision is poor in extreme cases. Threats to vision can arise from a secondary glaucoma and extreme exophthalmos with consequent corneal damage. In the case of indirect CCFs, the clinical manifestations may be due to the combination of cavernous sinus thrombosis and a DAVF. In such cases, an extremely small DAVF may be responsible for extreme clinical manifestation. In the case of a direct CCF, retrograde flow in the ophthalmic artery may occur, contributing to retinal ischemia that, when combined with the high venous pressure, may lead to immediate permanent loss of vision.

Problems other than those with the eye can occur. Retrograde cortical venous drainage may be present (middle cerebral veins or pontine venous tributaries to the inferior petrosal sinus). When present, the considerations discussed relating to DAVFs need to be taken into account (Greenberg 2010; Macdonald 2008).

6. Etiology

Head or orbital injury;
Rupture of a cavernous ICA aneurysm;
ICA dissection (Greenberg 2010; Korkmazer et al. 2013; Ertl et al. 2019).

7. Clinical Presentation

7.1. Symptoms

Headache
Impaired vision
Double vision
Tinnitus (pulsatile)

7.2. Signs

Proptosis
Chemosis
Orbital bruit
Cranial nerve palsy
Corkscrew vessels of the conjunctiva
Increased intraocular pressure
Ophthalmoplegia
Ptosis
Venous pulsations
Heme in Schlemm's canal on gonioscopy (Greenberg 2010; Korkmazer et al. 2013; Ertl et al. 2019; Macdonald 2008; Kalangu et al. 2009; Bennett et al. n.d.).

8. Radiological Assessment

Besides a CT scan with a CTA (Figure 3) and MRI of the brain with an MRA (Figure 4), the definitive investigation method is a DSA of the brain to identify the feeding artery, draining vein and location of the AV fistula.

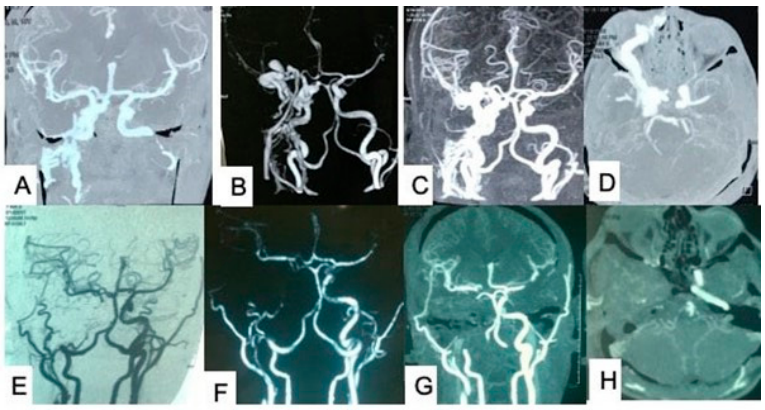

Figure 3. (**A**–**D**) Preoperative CTA showing a right-sided direct CCF. (**E**–**H**) CTA on the first POD after an STA–MCA bypass and ICA trapping. Source: Figure by authors.

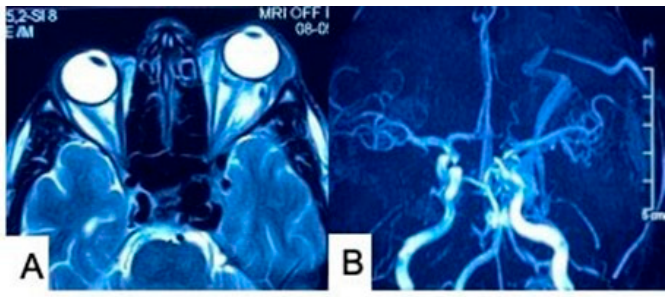

Figure 4. (**A**) MRI of the brain with an orbit showing the proptosis of the left eyeball, along with some irregularities in the cavernous sinus (left); (**B**) MRA showing the same constriction in the cavernous carotid artery, along with left-sided cavernous sinus dilatation and cortical venous drainage. Source: Figure by authors.

Proposed venous drainage-based classification system for CCFs;

Type	Venous drainage
I	Only posterior/inferior drainage
II	Posterior/inferior, as well as anterior drainage
III	Only anterior drainage
IV	Retrograde drainage into cortical veins ± other routes of venous drainage
V	High-flow direct shunt between a cavernous ICA and CS (Barrow type A) ± multiple routes of venous drainage (Kalangu et al. 2009).

9. Treatment of CCFs

Some traumatic CCFs may undergo spontaneous closure. There are various treatments for a CCF (Greenberg 2010; Cruz 1998; Korkmazer et al. 2013; Ertl et al. 2019; Macdonald 2008; Kalangu et al. 2009; Bennett et al. n.d.).

9.1. Carotid Compression

Carotid compression treatment may be successful in the closure of 17% of direct and 30% of dural CCFs.

9.2. Endovascular Management Is the Main Stay of Management

Approaches to carotid cavernous fistulas:

Transvenous routes via the following:

(a) Inferior petrosal sinus (IPS);
(b) Superior ophthalmic vein via the transfemoral route or direct surgical exposure;
(c) Facial vein via the transfemoral route;
(d) Transarterial route:

1. Direct CCFs are best managed with a detachable silicon balloon via an endo- arterial route;
2. Stent-assisted coil closure of a fistula may offer safe and effective management;
3. Onyx embolization by a transvenous route;
4. Microcoil embolization by a transvenous route;
5. Combination of Onyx and detachable coils through a transvenous route.

9.3. Surgical Trapping with or Without an STA–MCA Bypass

We commonly do surgical trapping of an ICA after thorough evaluation of the CCF by a DSA of cerebral vessels, cross-circulation study and balloon test occlusion (Figure 5).

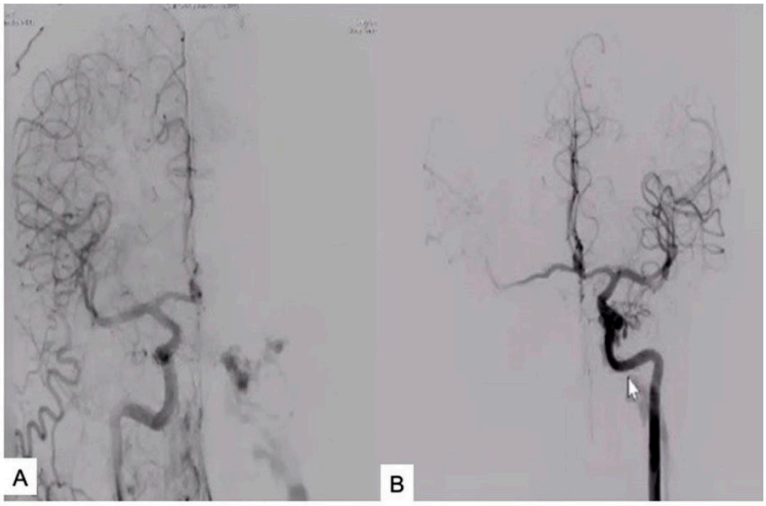

Figure 5. (**A**,**B**) Left and right cerebral ICA DSA, respectively, with a contralateral carotid occlusion in patients with a CCF. Source: Figure by authors.

We perform surgical trapping of the ICA by exposure and ligation at the high neck or Glasscock triangle in the middle fossa base, and ligation of a supraclinoidal ICA proximal to ophthalmic artery after anterior clinoidectomy. Some surgeons also use CS packing with trapping of the ICA.

[Balloon test occlusion: Before going to the occlusion or entrapment of an ICA, we need to evaluate the patency of the circle of Willis by performing a balloon test occlusion for half an hour.]

9.4. Gama-Knife Radiosurgery

In patients who fail or are unable to have an endovascular intervention, radiosurgical therapy of indirect CCFs has been advocated as a viable, non-invasive adjunct or primary treatment (Cruz 1998).

Author Contributions: Conceptualization, methodology, validation, formal analysis, investigation, resources, data curation, F.H.C., S.A. and N.A.; writing—original draft preparation, F.H.C. and S.A.; writing—review and editing, F.H.C.; visualization, supervision, N.A. and M.R.H. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Conflicts of Interest: The authors declare no conflicts of interest.

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Cerebral Cavernous Malformation (CCM)

Shamshul Alam, Forhad H. Chowdhury, Nazmin Ahmed and Mainul Haque Sarker

Abstract: A cavernous malformation is relatively rare in the brain. The dilated capillaries conglomerate together to form a nidus and the cavernoma (CM) within the brain. Usually, a cavernoma is silent, but it can present with bleeding and a seizure. It can occur in any area of the brain, but it has a higher tendency in the brainstem, deep structure of the brain and temporal lobe. MRI GRE and SW images are diagnostic for a CM. A symptomatic or ruptured CM demands surgical removal. In this chapter, the pathology, distribution and management of cavernomas are mentioned.

Abbreviations

CCM	cerebral cavernous malformation	CM	cavernous malformation
CT	computed tomography	DT	diffusion tensor
DSA	digital subtraction angiogram	DVA	dural venous anomaly
fMRI	functional magnetic resonance imaging	GRE	gradient recall echo
MRI	magnetic resonance	MVM	mixed vascular malformation
SEZ	safe entry zone	SW	susceptibility-weighted

1. Introduction

A cerebral cavernous malformations is one variety of an arteriovenous malformation where the dilated capillaries conglomerate together to form a nidus and the cavernoma (CM) within the brain. Usually, a cavernoma is silent, but it can manifest when there is bleeding or when present with a seizure. It can occur in any area of the brain but has a higher tendency in the brainstem, deep structure of the brain and temporal lobe (Greenberg 2010).

2. Natural History of Cavernoma

The incidence of a CCM varies from 0.17 to 0.56 per 100,000 per population per year (Al-Shahi et al. 2003). A CCM frequently presents in the fourth and fifth decade, and there is small female preponderance (58%). Roughly 50% of CCM patients remain asymptomatic, whereas about 25% present with single or multiple attacks of seizure. Additionally, patients can present with either hemorrhage (Figure 1A) (~12%) or focal neurological deficits (~15%) (Salman et al. 2012). Overall, the annual risk of a first-time cavernoma-related hemorrhage is low (0.4–0.6% per year) and the risk of subsequent hemorrhage is much higher (3.8–23% per year). The risk gradually decreases over time. It can present as multiple cavernoma (cavernomatosis) (Figure 1B) and can be familial.

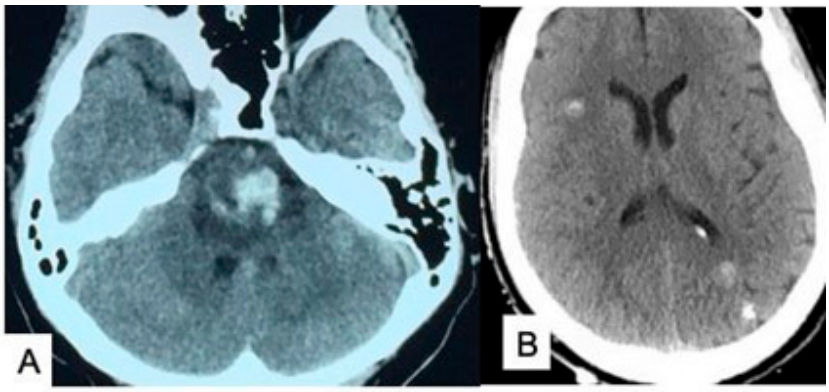

Figure 1. (**A**) CT scan showing bleeding in the pontine cavernoma. (**B**) CT scan showing multiple cavernoma. Source: Figure by authors.

3. Location

The distribution (locations) of CMs in a published series is shown in Table 1.

Table 1. Locations of cavernomas.

Location	Cavernoma	Percentage %
Cerebrum	84	69.4
Occipital	4	
Frontal	36	
Parietal	16	
Temporal	28	
Brainstem	17	14.0
Medulla	1	
Midbrain	2	
Pontomesencephalon	4	
Pons	8	
Pontomedullary	2	
Cerebellum	8	6.6
Cranial nerves	4	3.3
Spinal cord	8	6.6
Cervical	3	
Cervicomedullary	2	
Lumbar	1	
Thoracic	2	
Total CMs	121	99.9

Source: Authors' compilation based on data from Kirollos et al. (2019).

4. Developmental Venous Anomaly (DVA)

Developmental venous abnormalities (DVAs), also called venous malformations or venous angiomas, are frequently related to cavernomas. A DVA is a vascular abnormality that does not create any clinical signs on its own. In the proximity of a DVA, at least 40% of isolated cavernomas can occur.

The caput medusae indication of veins emptying into a solitary bigger collecting vein that then drains into either a dural venous sinus or a deep ependymal vein characterizes a DVA (Figure 2). The image has been compared to that of a palm tree. DVAs, on the other hand, can be found in any place, draining either superficially or deeply.

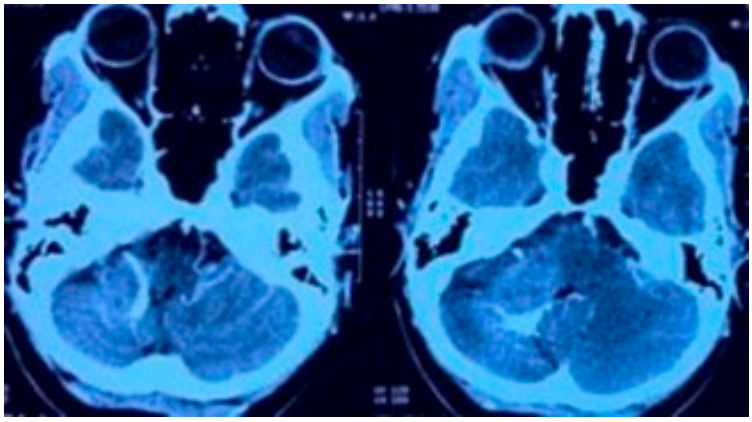

Figure 2. CT scan showing deep venous anomalies (DVAs), along with a cavernoma. Source: Figure by authors.

4.1. Associations

With the exception of the blue rubber bleb nevus syndrome, lesions are generally isolated (75%);

Mixed vascular malformations are found in 20% of cases (range: 8–33%) and are related to cavernous malformations (MVM)s;

Malformations of the venous system in the neck and head.

4.2. Classification Based on the Location

The most common locations (Figure 3) are as follows:

Fronto-parietal CM (36–64%), generally draining toward the lateral ventricle's frontal horn (Figure 3A,B);
Cerebellar hemispheric CM (14–27%), draining toward the 4th ventricle (Figure 3C);
Brainstem cavernoma (Figure 3D);
Spinal cavernoma (Figure 4).

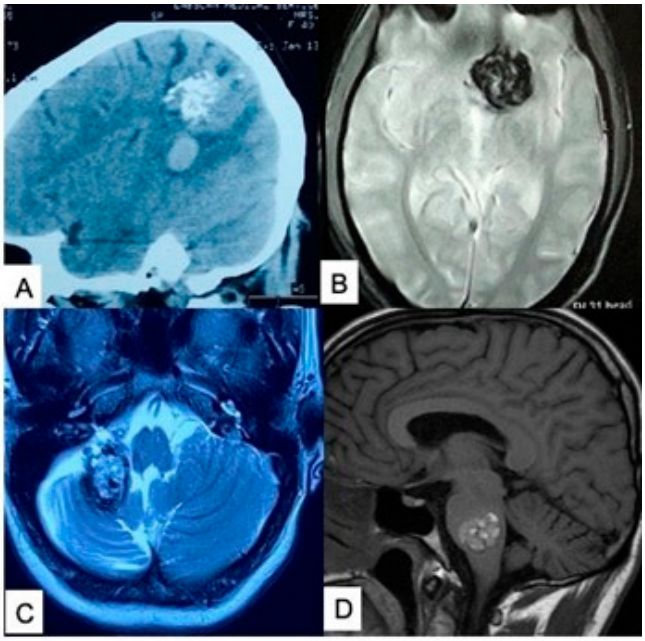

Figure 3. (**A**) CT scan showing a parietal cavernoma. (**B**) MRI showing a left basal frontal cavernoma. (**C**) MRI showing an rt cerebellar cavernoma. (**D**) MRI showing a pontine cavernoma. Source: Figure by authors.

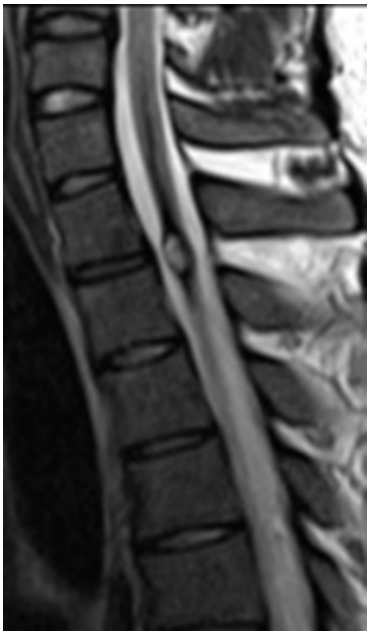

Figure 4. MRI showing a cervical spinal cord cavernoma. Source: Figure by authors.

5. Radiological Features of Cavernomas

State-of-the-art brain neuro-imaging techniques (called diffusion tensor tractography (DTI), gradient echo (GRE), as well as susceptibility-weighted (SW) sequences are used to permit for computational and noninvasive management planning (Table 2, Figure 5). Most cavernomas solely warrant observation with routine brain imaging to look for changes, recent bleeding ("hemorrhage") or new cavernoma/s.

Table 2. Imaging appearance of CMs.

CT	MRI	Angiography
Hyperdense	T2 bright areas with the susceptibility effect	Occult
	T1 can have bright areas, no appreciable enhancement, small adjacent DVA, if present strengthens the diagnosis	

Source: Table by authors.

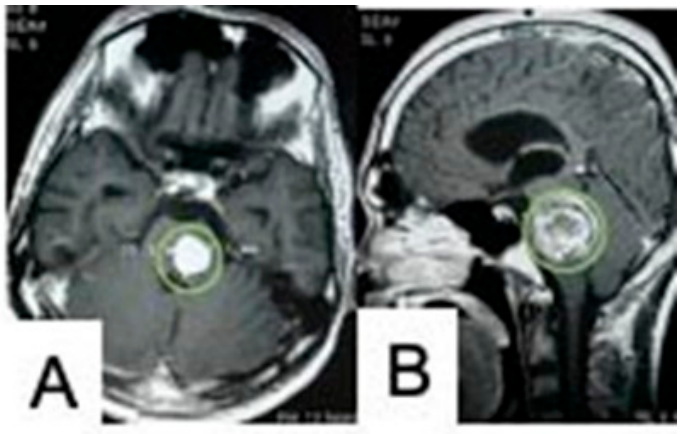

Figure 5. MRI of brain axial (**A**) and sagital (**B**) view showing a CM in the posterolateral pons with a halo sign. Source: Figure by authors.

A "popcorn" lesion is characterized by a center with a mixed signal in T1- and T2-weighted scans, that is bordered by a full hemosiderin ring with decreased signal intensity in T2W images. The severity of the hemorrhage affects the appearance of a CM. CMs have a tendency to expand with time. The common coexistence of a CM and DVA is thought to be the result of repetitive minor hemorrhages (D'Souza and Vadera 2022).

a. Zabramski classification of cerebral cavernous malformations (Zabramski et al. 1994):

Type I: Subacute hemorrhage. T1—hyperintense. T2—hyper- or hypointense.
Type II: The most common type—classic "popcorn" appearance. T1—heterogenous signal intensity at the center.
Type III: Chronic hemorrhage. T1—isointense to hypointense at the center.
Type IV: Numerous punctate micro-hemorrhages. T1—Hard to identify.

b. T 1 (Figure 6A): Variable signal based on the duration of the hemorrhage; Minimum fluid–fluid levels may be seen.

c. T 2 (Figure 6B):

- Rim is hypointense;
- Variable internal signal based on the duration of the blood products;
- In a recent hemorrhage, an adjacent edema may be observed.

d. Gradient Recalled Echo (GRE) MRI:
GRE T2/SWI (Figure 6C)

- Blooming is prominent;
- Helpful for finding tiny lesions that would otherwise go undetected by traditional spin echo sequences, particularly in individuals with familial or multiple cavernous malformations.

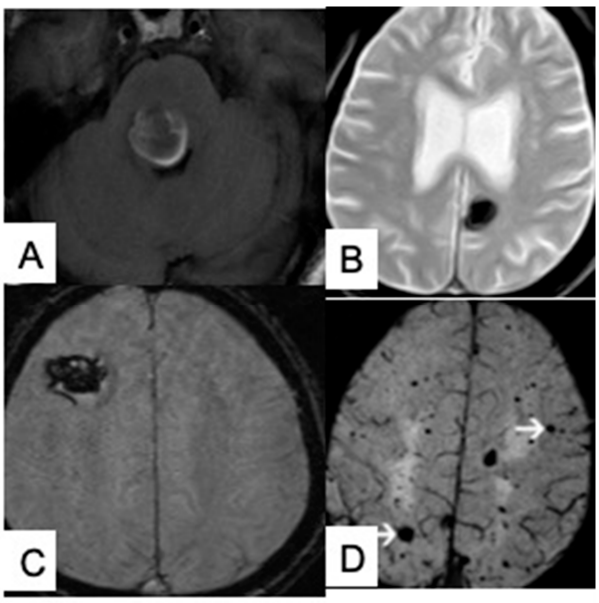

Figure 6. (**A**) MRI T1W image showing a high-signal hemosiderin ring. (**B**) MRI T2W image showing a hypointense lesion. (**C**) SW MRI showing an rt frontal cavernoma. (**D**) MRI showing multiple cavernomas. Source: Figure by authors.

With its capacity to show hemosiderin-filled cerebral parenchyma with a highly identifiable low intensity, a GRE MRI scan is a significant tool for diagnosing CMs. Traditional MRIs reveal an average of five lesions per patient in studies on familial CMs, whereas a T2W GRE MRI discovers a mean of sixteen pathologies per person. A GRE MRI is capable of not only identifying all existing lesions, but also delineating them more accurately. While a GRE MRI provides various advantages, it is vital to keep in mind that it increases the relative size of the CM. GRE MR imaging may also reveal multifocal CMs in older persons with hypertension, as well as a history of stroke, but these should not be confused with familial CMs; hypertensive angiopathy causes them.

e. Susceptibility-Weighted MR Imaging (Figure 6D): Since it reliably distinguishes deoxyhemoglobin and hemosiderin, susceptibility-weighted (SW) scanning is highly useful for identifying CMs. SW imaging is also the only approach for differentiating CMs and telangiectasias that do not bleed. It has been demonstrated to outline CMs more precisely, as well as discover additional CMs that are not seen with traditional imaging modalities.
f. Diffusion Tensor (DT) Imaging (Figure 7): Even though CMs are deeply placed in certain areas, DTI and fMRI are utilized preoperatively for better visualization of the lesions and nearby parenchyma in terms of improving the surgical success. The surgeon can see the white matter tracts that regularly pass over the hemosiderin rim of the CM using DT tractography. When removing CMs in the brain, an fMRI captures activity-dependent variations in cerebral blood flow, which is highly beneficial.

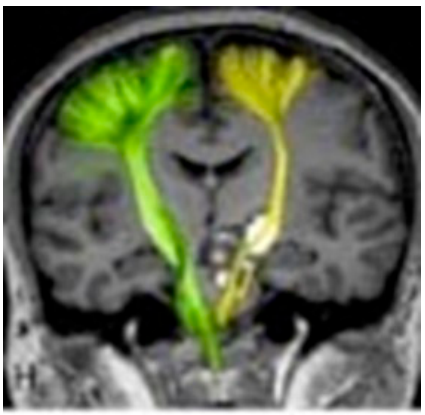

Figure 7. MR showing tractography in a cavernoma patient. Source: Figure by authors.

g. Angiography (DSA): CMs are angiographically occult and they do not have arteriovenous shunting.
h. CT scan of brain (Figure 8)

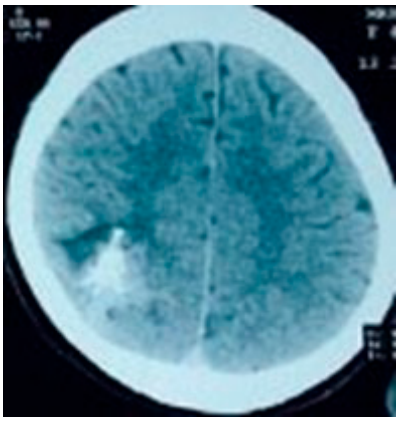

Figure 8. CT scan showing a calcified cavernoma. Source: Figure by authors.

6. Clinical Presentation

Seizures, headaches, neurologic deficits and asymptomatic presence are the four major kinds of clinical presentation. The most common presenting symptom is seizure, which affects 35–55% of patients. Several symptoms are found in many people. A bleed into the neighboring brain parenchyma occurs in some patients in each of the clinical groups. The hemorrhages are normally tiny, but they might be significant on rare occasions, causing the patient to rapidly deteriorate (Greenberg 2010).

7. Treatment

7.1. Microsurgery

Indications of surgery include a ruptured CM, CM with a mass affect, cranial nerve palsy and epilepsy. For a ruptured and symptomatic CM, it is the standard treatment (Greenberg 2010; D'Souza and Vadera 2022; Spetzler et al. 2020; Spetzler et al. 2017; Macdonald 2008; Mouchtouris et al. 2015).

7.1.1. Microsurgical Treatment of a Brainstem CM: Surgical Approaches

a. Brainstem anatomy: The diencephalon, midbrain, pons and medulla oblongata are the four components of the brainstem, which have an ectodermal origin. The brainstem links the cerebral hemispheres well with the spinal cord as well as the cerebellum. It is responsible for mandatory vital functions like respiration, cardiac pulsation, blood pressure, consciousness control and sleep. White and gray matter coexist in the brainstem.

Although complex, the internal anatomy of the brainstem is structured in three laminae (tectum, tegmentum and basis) that run the length of the brainstem (Spetzler et al. 2020).

b. Approaches to CMs in the brainstem

Microsurgical approaches to brain stem CMs are shown in Table 3.

c. Approaches to the midbrain
Supra cerebellar infratentorial approaches:

1. Midline;
2. Lateral;
3. Far lateral:

- Supracerebellar transtentorial approach;
- Occipital transtentorial approach.

The dorsal midbrain, pineal region and upper pons can also be approached by the occipital transtentorial approach:

d. Ventral midbrain approaches (shown Table 4);

e. For lateral midbrain and upper pons (shown Table 4).

Table 3. Surgical approaches to CMs in the brainstem.

Lesion location	Anterior	Lateral	Posterior
Midbrain	Pterional, orbitozygomatic (OZ) subtemporal	Lateral infratentorial supracerebellar (LIS)	LIS
Pons	Pterional, OZ subtemporal	Far lateral suboccipital retrosigmoid	Median suboccipital/4th ventricular
Medulla	Subtemporal, far lateral suboccipital transcondylar	Lateral suboccipital retrosigmoid	Median suboccipital/4th ventricular

Source: Authors' compilation based on data from Spetzler et al. (2017).

7.1.2. Safe Entry Zone (SEZ) to the Brainstem

A. Pons

Safe entry zones (SEZ) to the Brainstem are shown in Figure 9, and in pons are shown in Table 4.

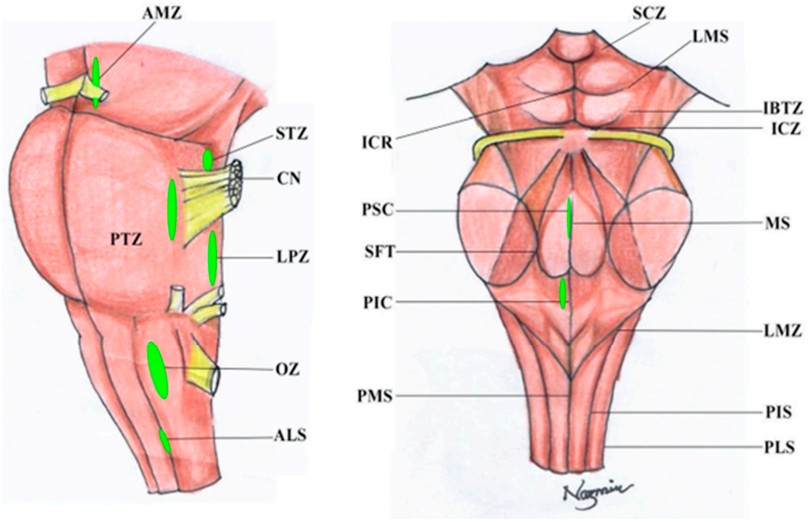

Figure 9. Principles of SEZs to the brainstem. The colored ellipses represent areas where petit neurotomies can be carried out to avoid minuscule perforators, prime nerve tracts and nuclei. (**Left**) Antero-lateral surface of the brainstem demonstrating some anterior and anterolateral SEZs. (**Right**) View of the dorsal surface of the brainstem demonstrating SEZs on the surface of the quadrigeminal plate, floor of the fourth ventricle. (ALS—anterolateral sulcus; AMZ—anterior mesencephalic zone; CN—cranial nerve; IBTZ—inferior brachium triangular zone; ICR—intercollicular region; ICZ—infracollicular zone; LMS—lateral mesencephalic sulcus; LMZ—lateral medullary zone; LPZ—lateral pontine zone; MS—median sulcus of fourth ventricle; OZ—olivary zone; PIC—paramedian infracollicular; PIS—posterior intermediatesulcus; PLS—posterior lateral sulcus; PMS—posterior median sulcus; PSC—paramedian supracollicular; PTZ—peritrieminal zone; SCZ—supracollicular zone; SFT—superior fovea triangle; STZ—supratigeminal zone). Source: Figure by authors.

Table 4. Pontine SEZ.

Approach	SEZ
Subtemporal transtentorial	Supratrigeminal
Anterior petrosectomy	Supratrigeminal, peritrigeminal
Suboccipital telovelar	Median sulcus of the 4th ventricle, paramedian infracollicular, superior fovea triangular
Retrosigmoid	Supratrigeminal, peritrigeminal lateral pontine
Retrolabrynthine	Supratrigeminal, peritrigeminal lateral pontine

Source: Authors' compilation based on data from Spetzler et al. (2017).

In the posterior pons/floor of the 4th ventricle, safe areas are the suprafacial triangle and infrafacial triangle (Spetzler et al. 2017). These areas are approached by a suboccipital teloveloar approach.

In the lateral pons, the safe areas are as follows:

- Supratrigeminal area;
- Peritrigeminal area;
- Infratrigeminal area (between 5th and 7th nerve);

The lateral pontine zone can be reached by a retrosigmoid approach.

To reach betel, more wide exposure is required and then we need to utilize a presigmoid approach.

B. Medulla Oblongata

Medullary safe entry zone: In the anterior medulla, the safe zone is the olivary area. The olive is a small elevation formed by the location of the inferior olivary nucleus.

Olivary zone: The olivary zone can be reached by the far lateral approach.

Dorsal medullary safe zone: The dorsal lateral medullary zone is a posterior midline sulcus and laterally medullary zone, respectively. This area is approached by midline suboccipital craniotomy.

7.2. Conservative Treatment

Conservative treatment is utilized for incidental CMs.

7.3. Stereotactic Radiosurgery

Stereotactic radiosurgery is usually not recommended.

Author Contributions: Conceptualization, methodology, validation, formal analysis, investigation, resources, data curation, S.A., F.H.C. and N.A.; writing—original draft preparation, S.A. and F.H.C.; writing—review and editing, F.H.C., visualization, supervision, M.H.S. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Conflicts of Interest: The authors declare no conflicts of interest.

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