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### RESEARCH ARTICLE

#### SPORADIC CEREBRAL CAVERNOMATOSIS WITH HYPOTHALAMIC LOCATION REVEALED BY LEFT LATERAL HOMONYMOUS HEMIANOPSIA: A CASE REPORT

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#### Abstract

Cavernous malformations are low-flow vascular malformations that are histologically characterized by the lack of mural elements of mature vascular structures and intervening parenchymal neural tissue. They occur in sporadic and familial forms. Familial cases are associated with a high frequency of multiple lesions, which are less frequently associated with sporadic cases. They represent 10–20% of all central nervous system (CNS) vascular malformations. A supratentorial location of lesions has been reported to be more frequent than an infratentorial location. Hypothalamic location of cavernoma is extremely rare. We present a case of 40-year-old female who presented with progressive left lateral homonymous hemianopsia. Magnetic resonance imaging (MRI) is pivotal in the management of this condition and it is more sensitive than computed tomography (CT) for the detection of cavernomas. We describe the imaging characteristics of the lesion in such a rare location, highlighting the role of MRI which guarantees a higher anatomic contrast resolution. We also suggest some clues for differential diagnosis.

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#### Introduction:-

Cavernous angioma (CA) is a congenital vascular malformation consisting of thin walled sinusoidal spaces lined with epithelium without interposing glial or neural tissue (1).

The incidence of CA is 0.4% to 0.9% in the general population (2) representing 10%-20% of all vascular malformations of central nervous system (3). Cavernous malformations of the optic pathway and hypothalamus are extremely rare. They represent 1% or less of all cavernous malformations. Here, we describe the main characteristics of this very unusual lesion found. MRI is pivotal in the management of this condition particularly the susceptibility weighted imaging (SWI- SIEMENS) and T2\*- Gradient-echo sequences. In this report, we present a patient who had multiple cerebral cavernomatosis presenting with progressive loss of visual acuity and field, with history of seizures never explored.

Surgical removal is the recommended treatment to restore or preserve vision, to decompress the visual apparatus, and to eliminate the risk of future hemorrhages.

#### Case Report:-

A 40-year-old female patient, a known diabetic type 2 on treatment, presented with a history of progressive loss of visual acuity and field of 3 months duration which got aggravated since the last 25 days with appearance of

hemianopic visual field loss on the left side of both eyes evoking a left lateral homonymous hemianopsia. She also had headache for the past 5 years and history of seizures since childhood, unexplored. There was no history of focal neurological deficit. Clinical neurological examination was normal and vital functions were stable. No alteration of consciousness. Cerebellar signs were absent.

Magnetic resonance imaging (MRI) revealed a multiple heterogeneous lesions in the supratentorial compartment and ambient cistern, especially subcortical in the frontal ant temporal lobe with mixed signal intensities (isointense and hypointense) on T1 and T2 weighted images suggestive of blood of different ages (figure 1a, c and d). The lesions were often surrounded by a peripheral rim of hypointensity on T2\* Gradient-echo sequence demonstrated blooming consistent with old hemorrhage, and they are hypointense on SWI sequence (figure 2). Minimal peripheral enhancement occurred after the administration of gadolinium (figure 3b). There was no edema or mass effect as noted on FLAIR images (figure 1b).

A well-defined suprasellar mass on the right side of the hypothalamus was detected responsible for the ophthalmic clinical manifestations, measuring 20 mm (H) x 12 (AP) x 14 mm (T), with a mixed intensity core appearing like “popcorn”. The mass has the same characteristics as the lesions described above. It has a mass effect on the right optic tract which appear hyperintense on T2 weighted images extended to the optical chiasma (figure 3a), and raise the third ventricle. The pituitary gland and pituitary stalk were normal. No other vascular malformation was found especially developmental venous anomaly (DVA)

Presence of mixed intensity core with a hypointense rim noted on T2 Gradient-echo sequence were the basis on which a diagnosis of cavernomatosis was made. Hypothalamic location is responsible for the ophthalmic clinical manifestations in our patient.

After making diagnosis, the patient was referred to the neurosurgery department for a therapeutic decision: surgery treatment or radiosurgery.

### **Discussion:-**

We use the terms ‘cavernoma’ and ‘cavernous malformation’ (CM) to designate a vascular hamartoma with a prevalence of 0.4–0.9% (2) within the general population. The malformation grows as a result of recurrent internal haemorrhages (4), unlike hemangiomas which are ‘true’ benign vascular neoplasms. CMs represent 10–20% of all central nervous system (CNS) vascular malformations (3), being located everywhere inside the CNS (80% supratentorial), especially subcortical in the frontal and temporal lobes (4; 5). Hypothalamic location of cavernoma is even rarer accounting for only 1% or less of the cavernomas (6; 7). Both sporadic (75%) and multiple/familial forms (10–30%) are recognized.

In our patient, the family investigation did not find any genetic mutation. We also did not find a family history of multiple cavernomas, epilepsy and focal neurologic deficits, particularly in his parents and his children. It was concluded that our case was most probably sporadic.

Patients with CA are often asymptomatic and when symptoms are present, they depend on the location and size of the lesion. Most frequent presentations are focal neurological deficits, hemorrhage and epilepsy (8). Optic pathway and hypothalamic CA are usually brought to clinical attention by visual deterioration. Chiasmal apoplexy is the most common clinical presentation, characterized by sudden visual disturbance, headache, retroorbital pain, and nausea. Apoplectic symptoms are often preceded by transient blurred vision and headaches that occur weeks or months beforehand. Symptoms can also occur in a chronic or progressive manner with intermittent episodes of headache and visual loss. Transient or progressive symptoms are likely to result from recurrent episodes of hemorrhage and lesion growth.

Magnetic resonance imaging is reliable in the detection, follow up and diagnosis of cavernoma. It is the most sensitive and specific imaging modality for identifying this vascular malformation. These lesions usually appear as areas of mixed signal intensity with a hypointense rim. A typical “popcorn” appearance is characteristic of hemorrhagic components of different ages. Minimal or no enhancement occurs after the intravenous administration of Gadolinium. Lesions of the optic tract may appear as nerve thickening on coronal views, whereas lesions in the chiasm or hypothalamus often appear as focal and round masses. SWI sequence is preferred over other imaging techniques as it uses relaxation and magnetic susceptibility differences between oxygenated and deoxygenated blood

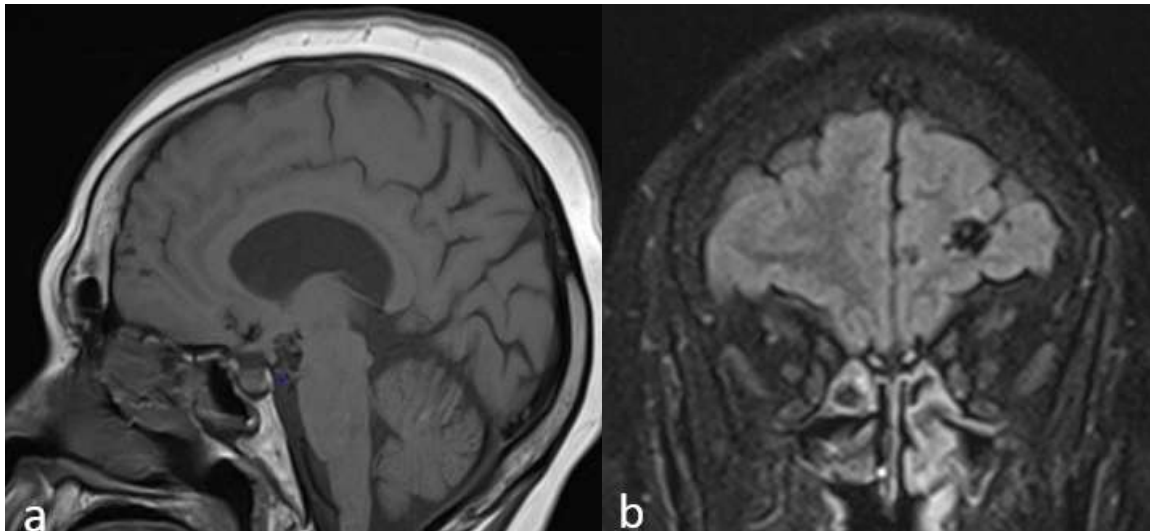
of arterial and venous circulation to detect lesions [9]. However, the lesional size in SWI appears larger than their real size [10]. Gradient-echo sequences are particularly sensitive to small hemorrhages and are ideal for detecting the presence of additional lesions in patients with multiple CMs. The hemosiderin rim appears particularly dark on T2 gradient-echo sequences. It is notable that peripheral hypointensity was not found in more than one-third of the cases providing detailed MR imaging information. The absence of the hemosiderin rim may have resulted from blood washout by CSF. Diffusion and perfusion weighted imaging are less useful for diagnosis (mask effect of hemosiderin). On CT scans, Cavernous malformations of the hypothalamus (OPH) appear as areas of hyperdensity with or without calcification and can sometimes mimic the appearance of a tumor or thrombosed aneurysm (11). Angiography does not typically show any pathological vessels. In some cases, however, an associated venous angioma (developmental venous anomaly), which are veins arranged in a radial pattern draining into a single larger vein, can be visualized.

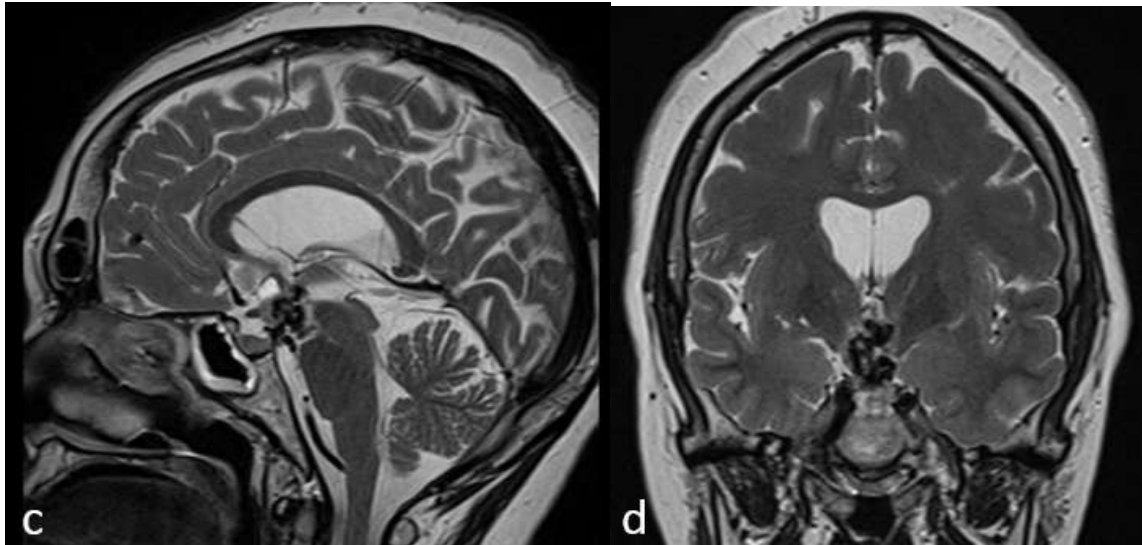
CA varies in size from a few millimeters to a few centimeters. The majority of them are small, but may reach a significant size. Those measuring more than 6 cm in diameter are termed as giant CA (12).

Actually, literature review showed that only seven cases of cavernomas have been reported in the hypothalamus [7; 13].

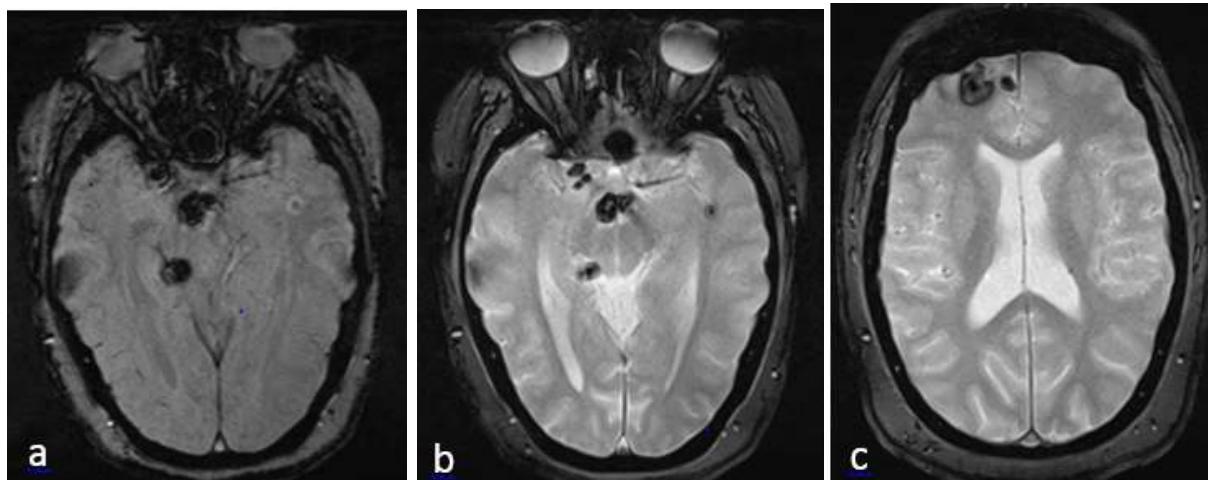
MRI findings are diagnostic of CA which usually do not require any further confirmation. By any means; however, in very rare cases of ambiguity, some of the differential diagnoses which may be thought of are hypothalamic glioma, germinoma, hamartoma, histiocytosis and granuloma, but presence of mixed intensity “popcorn” like core, representing blood in various stages of degeneration, surrounded by a hypointense halo due to hemosiderin on T2 weighted images helps in ruling out these conditions.

Surgical excision is mandatory for symptomatic hypothalamic and optochiasmatic cavernoma. Free margins of resection prevent re-haemorrhage, providing a good prognosis (80–90% clinical stabilisation and improvement/recovery of the visual function) [7; 14]. Radiosurgery should be excluded to avoid re-bleeding and damage to the optic pathways [7; 15].

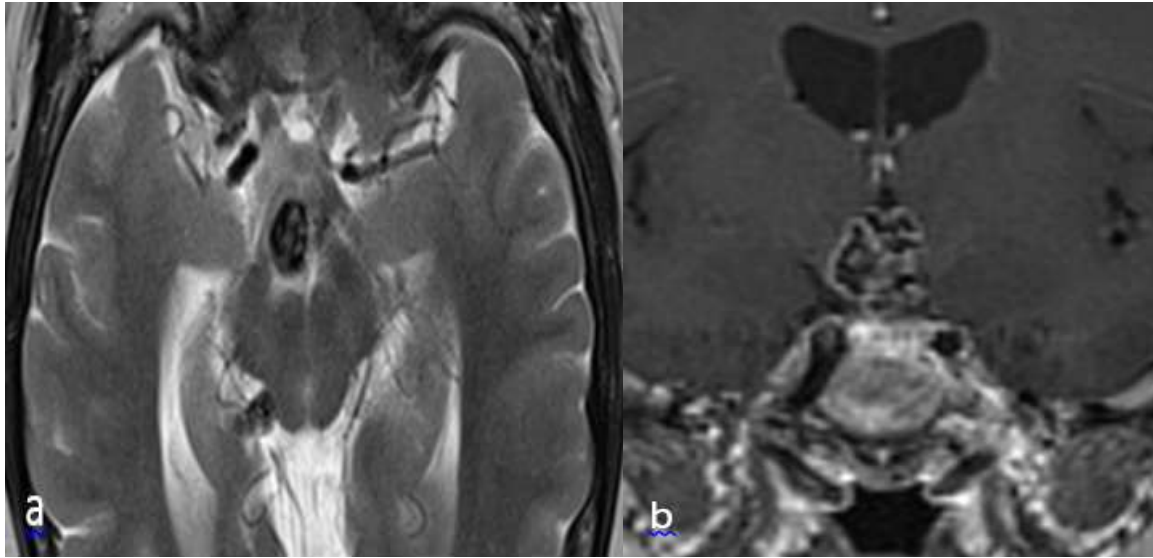




**Figure 1:-** A- sagittal T1 weighted; B- coronal FLAIR; C- sagittal T2 weighted; and D- Coronal T2 weighted images show a well-defined mass on the hypothalamus and on left frontal lobe with a mixed intensity core appearing like “popcorn” surrounded by a low signal intensity hemosiderin rim seen on T2 weighted images with no edema or mass effect as noted on FLAIR images.



**Figure 2:-** Magnetic resonance imaging showing lesions with peripheral rim of hypointensity on T2\* Gradient-echo sequence (b; c), and hypointense on the susceptibility imaging sequences (a).



**Figure 3:-** a- axial T2 weighted ; b- coronal post-Gadolinium T1-weighted MR images demonstrating lesion on the right side of the hypothalamus presenting mass effect on the right optic tract which appear hyperintense on T2 weighted images extended to the optical chiasma. Minimal peripheral enhancement occurred after the administration of gadolinium.

### Conclusion:-

Cavernous malformation of the hypothalamus are rare and challenging lesion. The most common presenting symptom was visual deficit. Magnetic resonance imaging is the most sensitive and specific imaging modality for identifying CMs with a mixed hyperintense reticulated central core, surrounded by a hypointense rim. Emergent surgical removal is indicated to prevent permanent visual damage.

### References:-

1. Akbostanci MC et al. Cavernous angioma presenting with hemidystonia. *Clin Neurol Neurosurg.* 1998;100(3):234-7.
2. Rotondo M et al. Cavernous malformations isolated from cranial nerves: unexpected diagnosis? *Clin Neurol Neurosurg* 2014; 126: 162–168.
3. Preetam Bhujagonda Patil et Al. Hypothalamic Cavernous Angioma Associated With Memory and Behavior Disturbance Attacks: Role of Imaging in Diagnosis. *Iran J Radiol.* 2012;9(1):42-44.
4. Arrue P et al. Cavernous hemangioma of the intracranial optic pathways: CT and MRI. *J Comp Assist Tomogr* 1999; 23: 357–361.
5. Alafaci C et al. Cavernous malformation of the optic chiasm: an uncommon location. *Surg Neurol Int* 2015; 6: 60.
6. Gross BA, Du R. Diagnosis and treatment of vascular malformations of the brain. *Curr Treat Options Neurol* 2014; 16:279. 2.
7. Liu JK et al. Cavernous malformations of the optic pathway and hypothalamus: Analysis of 65 cases in the literature. *Neurosurg Focus* 2010; 29:E17.
8. Attar A et al. Surgical treatment of intracranial cavernous angiomas. *J Clin Neurosci.* 2001; 8(3):235- 9.
9. Campbell PG et al. Emerging clinical imaging techniques for cerebral cavernous malformations: A systematic review. *Neurosurg Focus* 2010; 29:E6.
10. Tong KA et al. Susceptibility-weighted MR imaging: A review of clinical applications in children. *AJNR Am J Neuroradiol* 2008; 29:9-17.
11. Shibuya M et al. Cavernous malformations of the optic chiasma. *Acta Neurochir (Wien)* 136:29–36, 1995.
12. Son DW, Lee SW, Choi CH. Giant cavernous malformation: a case report and review of the literature. *J Korean Neurosurg Soc.* 2008; 43(4):198-200.
13. Toe BP, Ramli NM. Cavernoma of the hypothalamus. *Hong Kong J Radiol* 2011; 14:234-7.
14. Crocker M et al. Cavernous hemangioma of the optic chiasm: a surgical review. *Skull Base* 2008; 18: 201–212.
15. Panczykowski D et al. Optochiasmatic cavernous hemangioma. *Br J Neurosurg* 2010; 24: 301–302.