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ATYPICAL PRESENTATION OF CEREBRAL CAVERNOUS HEMANGIOMA

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INTRODUCTION:

The first person to describe cavernous hemangioma was Hubert Luschka.[1] In the year 1854, he published the case of cavernous vascular brain tumor ("kavernöseBlutgeschwulst des Gehirns" – German) and provided a detailed description of its gross macroscopic aspect. The first histopathological description was given by the renowned pathologist Rudolf Virchow in the year 1863.[2]In histologic terms, cavernous angiomas (CA) are blood-filled cavities covered by a single layerof endothelium. The intervening tissue includes microglia but no neural elements.[3]

At present, the terms cavernoma, cavernous hemangioma/angiomaand/or cavernous malformation are being used interchangeably. We must also note that the terminologies cavernoma, cavernous angioma, and cavernous hemangioma are misnomers since they tend to point towards a neoplasticorigin.However, these represent true vascular malformations.[4]

CASE DESCRIPTION:

A 47-year-old male came with a history of seizure disorder for the past 21 years and is on regular medication. Last episode was one month prior to the visit and lasted for 2 minutes. He also complained of headache and mild left sided weakness for 3 months. History of deep venous thrombosis was present.

Vitals:Pulse rate – 72/minute, BP – 120/70 mm Hg, SPO2 -98% @ room air, GCS -15/15.

Routine blood investigations were within normal range.

MRI (done in an outside institution) was referred to us from the Department of Neurosurgery. Imaging revealed a space occupying lesion in the right fronto-temporal lobe with imaging features of a malignant neoplasm – possibly Glioma.

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MRI FINDINGS:

- A large well-defined intraparenchymal multiloculated cystic lesion with significant surrounding edema is noted involving the right frontal lobe. The locules appear hypointense on T1 and hyperintense on T2 and show variable intensities in FLAIR images.
- DWI-ADC sequence showed no evidence of restricted diffusion.
- Mass effect is noted the form of effacement of adjacent sulcal spaces, compression of right lateral ventricle and midline shift to the left.
- On post contrast images, this lesion shows peripheral rim like enhancement with enhancing septations.No evidence of any enhancing solid components seen.
- Few blooming foci and multiple serpiginous flow voids are seen on SWI images.
- With the imaging findings of large intraparenchymal lesion showing significant perilesional edema, mass effect and peripheral rim enhancement with enhancing septations, we gave a diagnosis of cystic neoplasm.

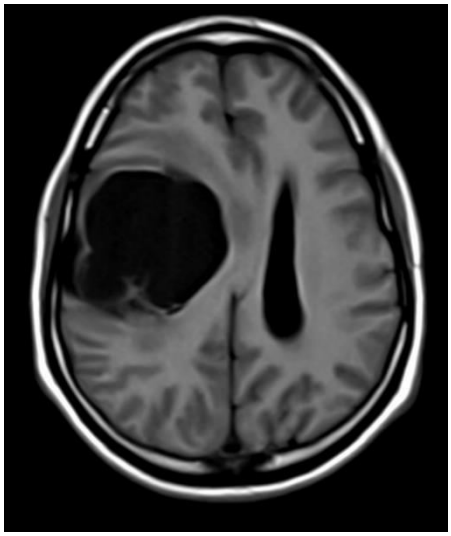

Figure 1: T1 axialMR sequence - Multiloculated cystic lesion with involving the right frontal lobe. The locules appear hypointense on T1.

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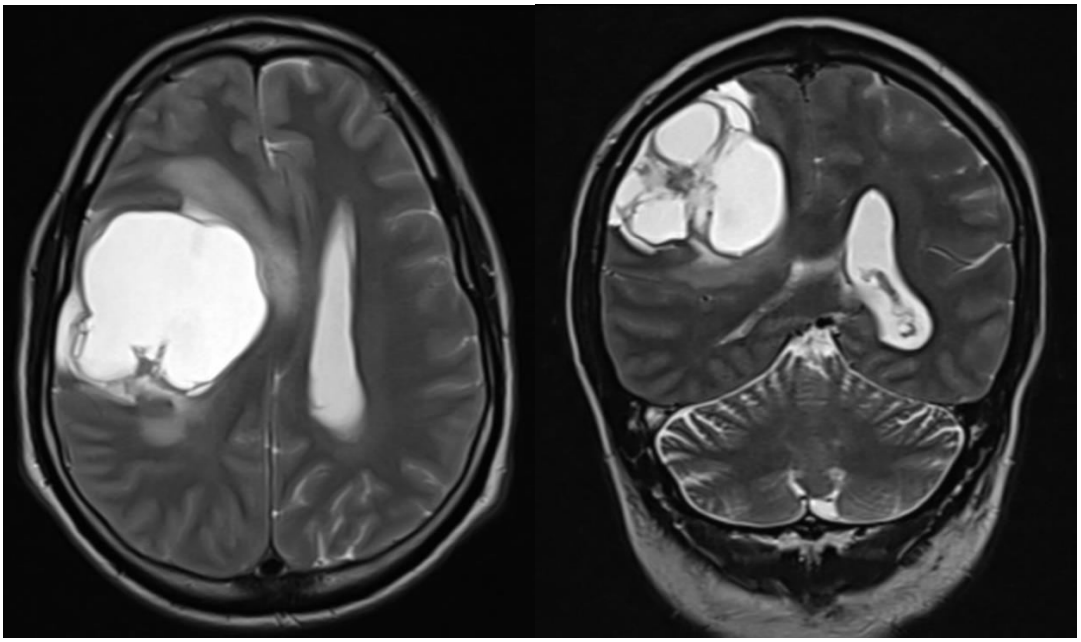


(A) (B)

Figure 2: T2 axial (A) and coronal (B) images show the multicystic lesion with hyperintense locules and surrounding hyperintense edema.

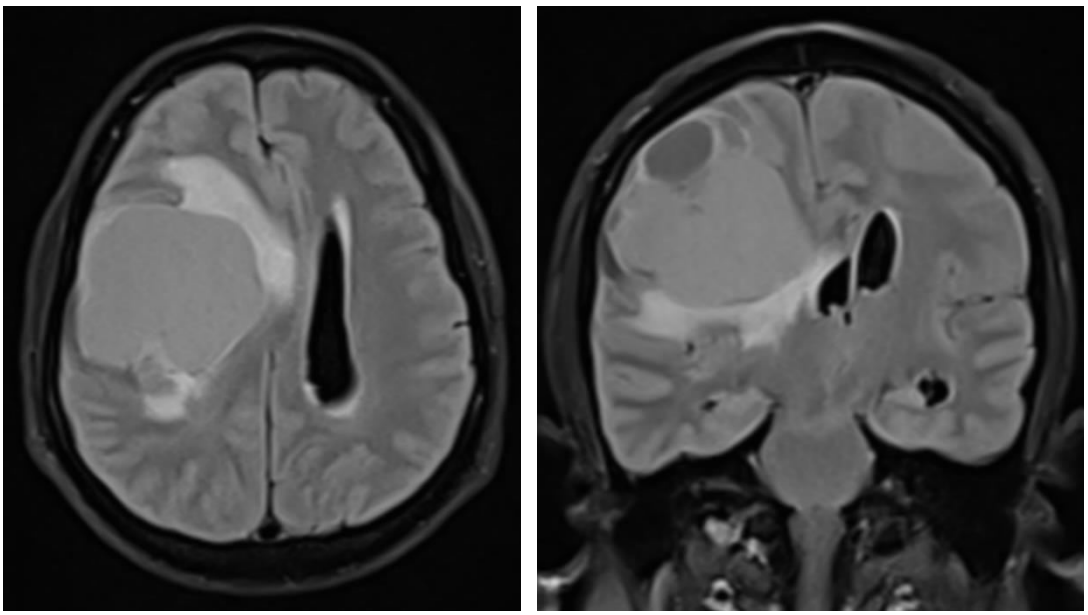


Figure 3: FLAIR axial (A) and coronal (B) images show the multicystic lesion with surrounding hyperintense edema and mass effect.

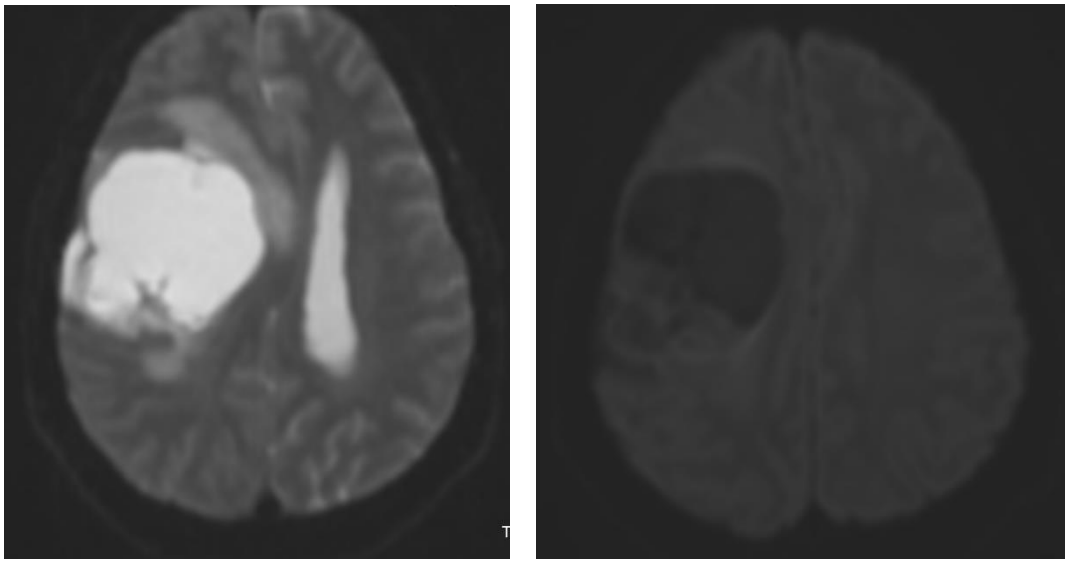
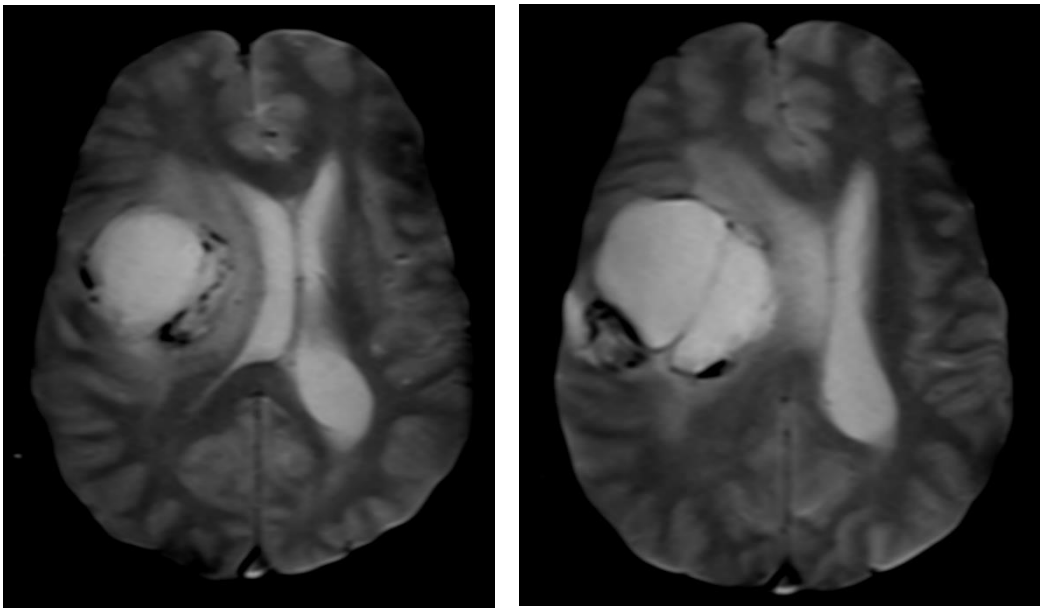


Figure 4: ADC (A) and DWI (B) images showing no evidence of restricted diffusion in the lesion.



(A) (B)

Figure 5: T2 gradient echo sequences showing flow voids within the lesion.

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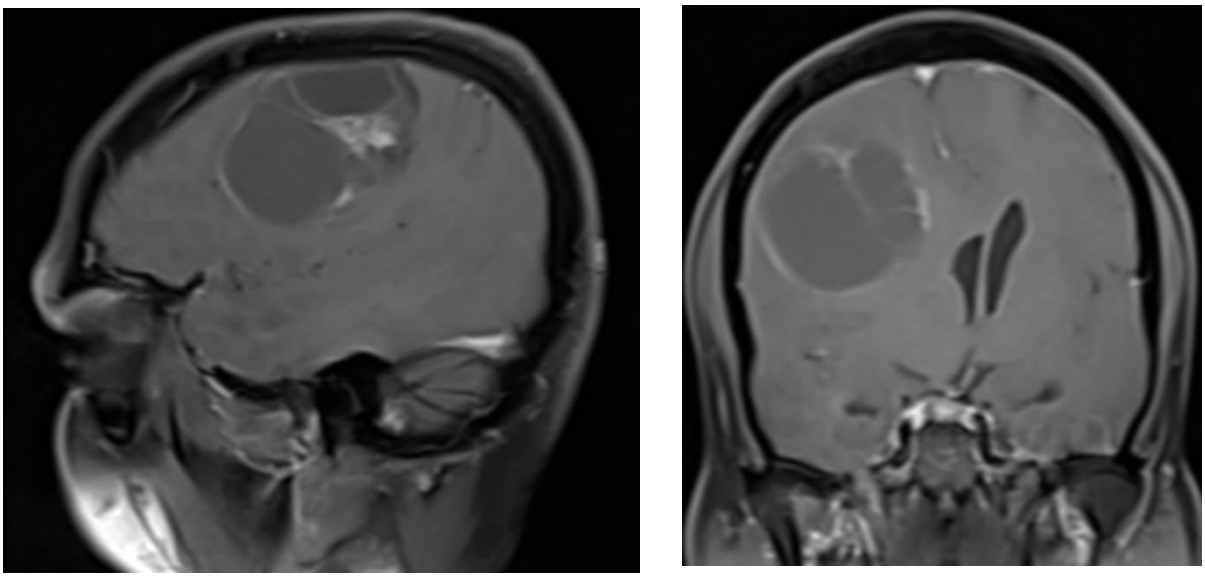

(A) (B)

Figure 6: Post contrast sagittal (A) and coronal (B) images showing peripheral rim like enhancement with enhancing septations within.

After preoperative evaluation, the patient underwent surgical excision of the lesion. Intraoperatively (Figure 7), a large cystic lesion was found and the same was excised with the help of intraoperative nerve monitoring (IONM). The lesion was removed in toto and sent for histopathological analysis. The post operative period was uneventful and the patient showed improved in leg power to normal.

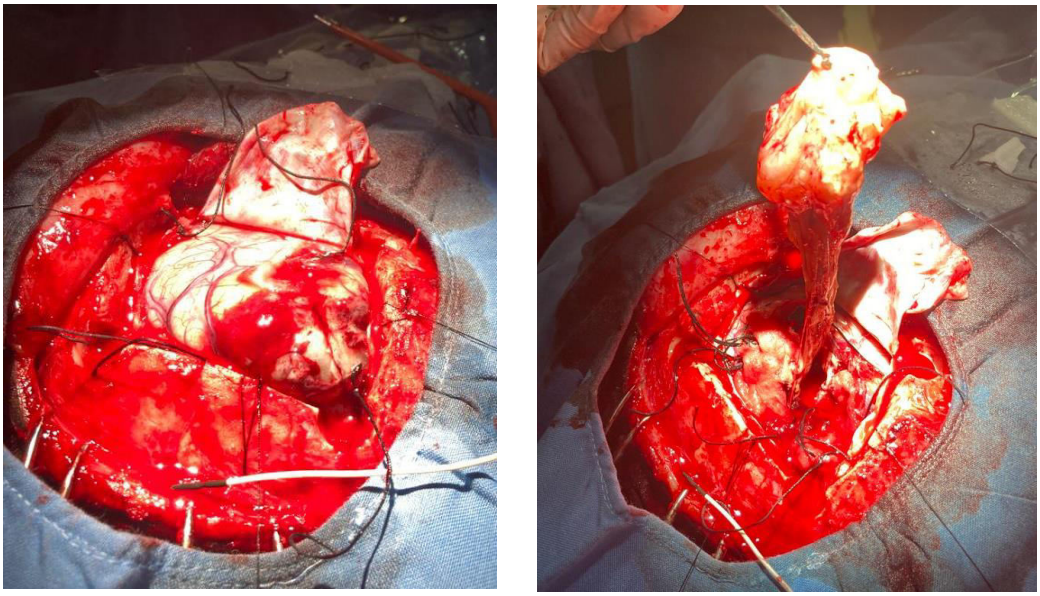

Figure 7:

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Histopathological analysis:

The gross specimen was a grey white cystic lesion that showed white exudates and areas of congestion on surface. The cut section revealed cystic spaces lined by thickened wall. Focal areas of hemorrhage and calcification were noted. Microscopy demonstrated thickened cyst wall lined by granulation tissue and hemosiderin laden macrophages. Focal ectactic and hyalinized vascular channels with scanty intervening neuroglial tissues were also noted. No evidence of malignancy was found in the given specimen. The final diagnosis was ruptured vascular malformation, probably cavernous angioma, leading to hemorrhage, calcification, gliosis and cyst wall formation.

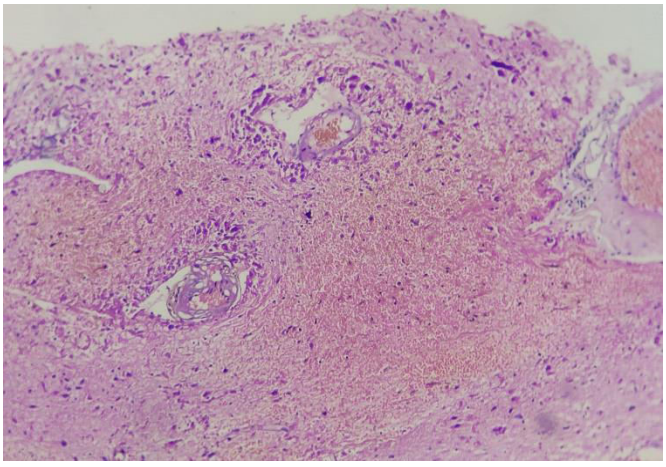

Figure 8:
Microscopy showing areas of hemorrhage.

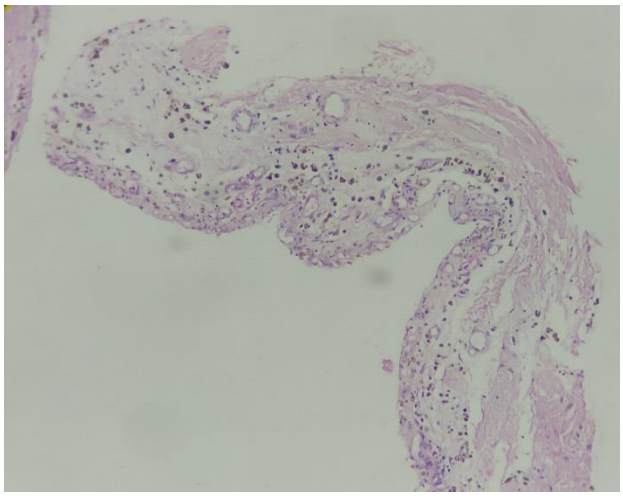

Figure 9:
Cyst wall lined by granulation tissue and hemosiderin laden macrophages.

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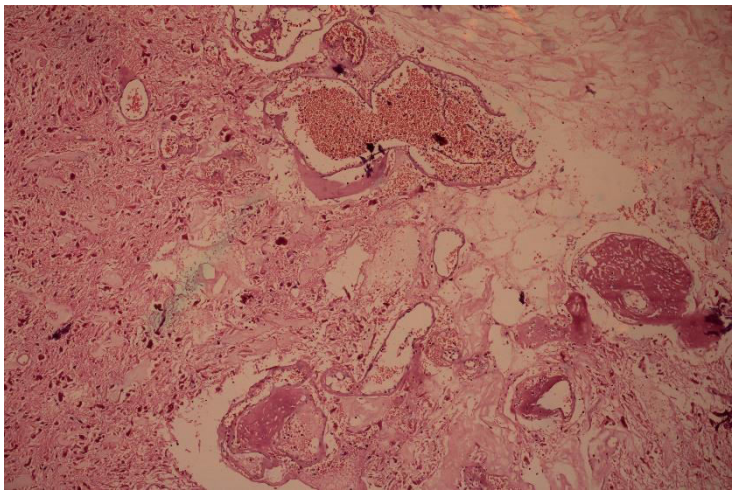

Figure 10:
Cyst wall with ectatic vascular channels.

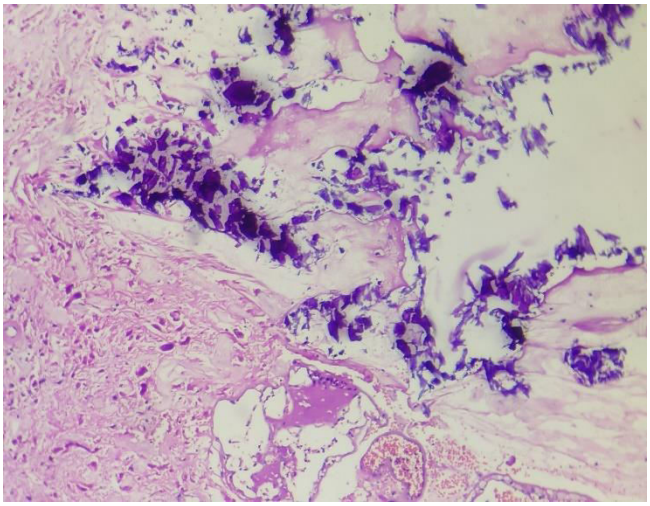

Figure 11:
Areas of calcification.

DISCUSSION:

Detecting cavernous angiomas by neuroimaging is based on certain morphologic and structural features. Non contrast CT scan may be negative in ~30-50% cases. In NCCT, it is often identified as well-delineated ovoid hyperdense lesion that is less than 3 cm with calcification seen in upto 60% cases.[5]These lesions have no specific localizations andmay be intra-or extra-axial.[6]In the pre-MRI era, CAs were difficult to diagnose. Angiogram may not detect these lesions and hence the term AOVM (angiographicallyoccult arteriovenous malformation) was sometimesused.[4] At present, even though no specific signs may be recognized and the most sensitiveimaging technique remains to be the MRI.[7]Since CAs are pathologically comprised of endothelial lined caverns with blood, thrombosis, calcification andhyalinization in varying stages of organization, these changes are reflected accordingly in the MRI with characteristic appearance.[8] Therefore, MRI shows a variable appearance depending onhemorrhage/stage.[5]Surrounding parenchymal tissue alsodemonstrates evidence of previous micro-hemorrhage andhemosiderin-filled macrophages reflected by the T2 hypointensitysurrounding the lesion. [8]

Zabramski classification is used depending on stage/ grade of CAs.[5]

- Type 1 = subacute hemorrhage (hyperintense on T1; hyper- or hypointense on T2).
- Type 2 = mixed signal intensity on T1, T2 with degrading hemorrhage of various ages(classic "popcorn ball" lesion).
- Type 3 = chronic hemorrhage (hypo- to isointense on T1, T2).
- Type 4 = punctate microhemorrhages ("black dots"), seen on GRE sequences.

The usual symptoms of a cavernoma are seizure, progressive neurologic deficit, hemorrhage.[9] Our case also presented with seizure.

To categorize as giant cavernoma (GCM), no threshold dimension has been universally accepted.[9]However, Lawton *et al*. described the lesions with a diameter greater than 60 mm as GCM.[11]Imaging appearance of GCM is variable. It can range from being a completely cystic lesion or a heterogenous lesion resembling neoplasms with mass effect and striking contrast enhancement.[9]Misdiagnosing a large cavernomas as pilocytic astrocytoma due to the isodense appearancewith poor contrast enhancement, as oligodendrogliomadue to calcifications, as ependymoma or metastatic melanoma with intratumoral hemorrhage has also been reported.[12]
Furthermore, cavernomas have also been reported in association with various intracranial tumors, including ependymomas and astrocytomas.These pathologies which are co-localizedwith features of both a cavernoma and a glioma are collectively termed 'angiogliomas'.[13]
Yun et al. found perilesional, hyperintense T1 signal to be a predictorof true CM in hemorrhagic lesions.[14]

Retrospective analysis:

Our inclination to give a diagnosis of cystic neoplasm was mainly based on the size, edema and mass effect caused by the lesion. There is also absence of typical T2 hypointense rim or popcorn appearance that is expected to be seen in cavernous angiomas.
But our case did not have restricted diffusion that is expected in malignant neoplasm. Presence of flow voids also favors consideration of vascular malformation. Moreover, the patient presented with a history seizure for almost 21 years. This long duration is unlikely to represent a neoplastic etiology.

Conclusion:

Since the imaging appearances of CAs are variable, the possibility of CA should be considered in the case of large intracranial tumor.

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Ethical consideration:

This article followed all ethical standards for carrying out research.

Data availability statement:

The authors confirm that the data supporting the findings of this study are available within the article.

Disclaimer:

The views and opinions expressed in this article are those of the authors and do not necessarily reflect the official policy or position of any affiliated agency of the authors.

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ABBREVIATIONS:

Cavernous Angioma – CA
Giant cavernoma – GCM
Magnetic Resonance Imaging – MRI