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CAVERNOUS MALFORMATION OF THE SEPTUM PELLUCIDUM: A CLINICAL RARITY

Radiology

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KEYWORDS

CASE REPORT

A 32-year-old female presented to the outpatient clinic with a complaint of persistent neck and back pain. There was no history of seizures, head trauma, or prior radiotherapy. Neurological examination was unremarkable, and no focal neurological deficits were identified.

An MRI of the spine was initially performed to evaluate the source of neck and back pain, which did not reveal any significant vertebral or disc abnormality. However, during the same study, two incidental lesions were identified. These lesions, located in the septum pellucidum and the left temporal lobe had heterogeneous hyperintense central cores with a peripheral hypointense rim on T2-weighted sequences (Figure 01).

Given these findings, a contrast-enhanced MRI (CEMRI) of the brain was recommended for further characterization. MRI brain confirmed both lesions, one each in the septum pellucidum (Figure 02) and in the left temporal lobe (Figure 03), measuring approximately 1.0 × 1.0 × 1.5 cm. On T2/FLAIR images, the lesions showed heterogeneous central hyperintensity with a classic "popcorn-like" internal appearance surrounded by a peripheral hypointense hemosiderin rim. On T1-weighted images, punctate hyperintense foci suggestive of subacute blood products were observed within both lesions.

On susceptibility-weighted imaging (SWI), the lesions demonstrated prominent "blooming," consistent with hemorrhagic components. Phase images confirmed blood signal intensity. Mild post-contrast enhancement was noted, but no restricted diffusion was seen on diffusion-weighted images (DWI) or apparent diffusion coefficient (ADC) sequences. Based on these imaging features, the diagnosis of septum pellucidum and left temporal lobe cavernomas was made, corresponding to Zabramski classification type II (1).

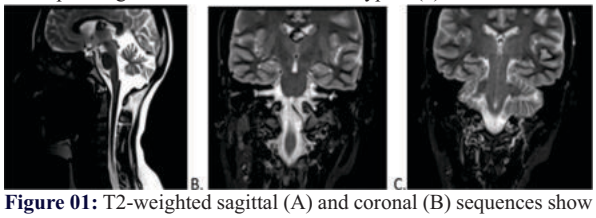
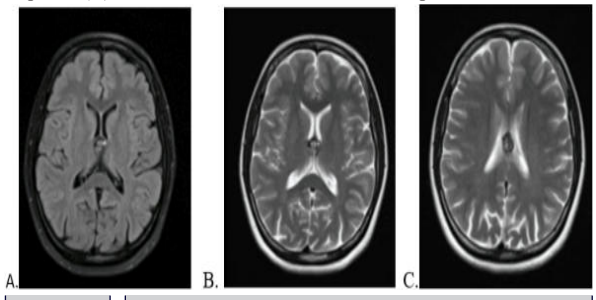


Figure 01: T2-weighted sagittal (A) and coronal (B) sequences show an incidentally detected, well-defined, heterogeneously hyperintense central lesion, 'popcorn' appearance with peripheral hypo-intensity located in the septum pellucidum. The T2-weighted coronal sequence(C) shows a similar lesion in the Left temporal lobe.



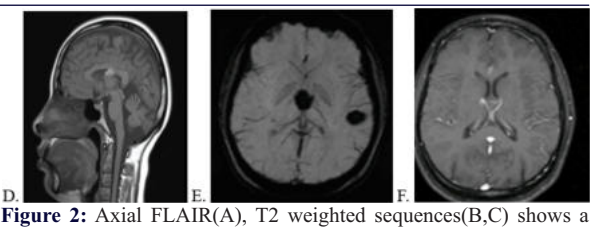


Figure 2: Axial FLAIR(A), T2 weighted sequences(B,C) shows a well-defined T2/FLAIR heterogeneously hyperintense lesion with 'popcorn' appearance located in the septum pellucidum. D : T1 weighted mid-sagittal image showing punctuate foci of hyperintensity within the lesion. E: T2* gradient echo image showing significant 'blooming' at the site of the lesion. F: Post-contrast axial T1-weighted axial image showing mild enhancement.

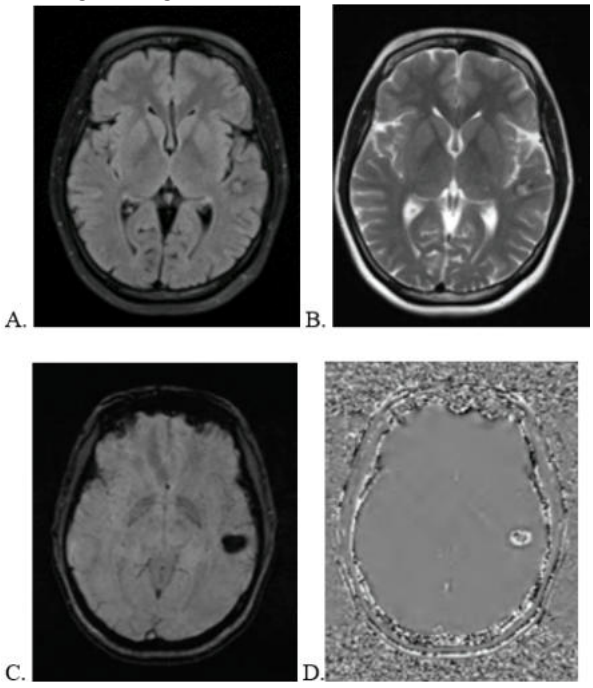


Figure 3: Axial FLAIR(A), T2 weighted sequences(B) show a well-defined T2/FLAIR heterogeneously hyperintense lesion in the Left temporal lobe. C: T2* gradient echo image showing significant blooming at the site of the lesion. D: Axial phase image showing blood signal intensity.

As the patient was asymptomatic from these lesions, with no localizing neurological signs, and without mass effect or evidence of obstructive hydrocephalus on imaging, a conservative management strategy was adopted including serial imaging and regular neurological follow-up.

DISCUSSION

Cerebral cavernous malformations (CCMs) or cavernomas are vascular malformations of the brain, having dilated capillary-like

channels lined by a single layer of endothelium, without intervening brain parenchyma. They account for 10%–25% of all central nervous system vascular malformations, with an estimated incidence of 0.4%–0.8% (2). Cavernomas may occur sporadically or in familial forms. The familial type, caused by mutations in CCM1, CCM2, or CCM3 genes, is associated with multiple lesions and may coexist with developmental venous anomalies (DVAs). Other risk factors include prior cranial radiotherapy and head trauma.

Supratentorial lesions account for 74–90% of all reported cases, of which intraventricular cavernomas contribute only 2.5%–10% (3,4), majority being located in the lateral ventricles, followed by the third ventricle (3,5). Cavernomas in the septum pellucidum are exceedingly rare, with only 11 cases reported worldwide till date.

Cavernomas are often detected incidentally, as in our present case, or they may present with symptoms such as seizures, headaches, focal neurological deficits, or rarely with signs of raised intracranial pressure, depending primarily on their location. Cavernoma located in the septum pellucidum holds potential risk of obstructing cerebrospinal fluid (CSF) pathways, leading to hydrocephalus, while hemorrhage can cause acute neurological deterioration. The annual risk of bleeding is estimated between 0.7% and 1.1%, but this increases significantly, up to 4.5%, in patients with a prior history of hemorrhage (2). Due to its proximity to the fornix, a part of the brain involved in memory processing, hemorrhage within cavernoma of the septum pellucidum has added risk of precipitating long-term memory impairments.

MRI is the neuro-imaging modality of choice for diagnosing cavernomas. On T2/FLAIR, cavernomas typically demonstrate a “popcorn” or “berry” appearance with mixed signal intensities reflecting hemorrhage at different stages. A hypointense hemosiderin rim typically surrounds the lesion as sequel to past episodes of micro-hemorrhages . T1-weighted imaging may reveal intra-lesional hyperintense foci due to subacute hemorrhagic products. SWI and T2* GRE sequences are particularly sensitive in detecting small lesions in the form of microhemorrhages. Contrast enhancement is variable from none to mild, and cavernomas are also well-known to be angiographically occult as they lack arteriovenous shunts (2,3,4).

The differential diagnosis for septum pellucidum cavernomas includes a wide spectrum of conditions ranging from benign entities like hematomas, arteriovenous malformations and neurocysticercosis to neoplasms such as central neurocytoma, sub-ependymoma, ependymoma, meningioma, and subependymal giant cell astrocytoma (3,4).

Management depends on lesion size, location, symptoms, and complications, with asymptomatic lesions (like in our patient) without mass effect usually managed conservatively while being kept under clinical and imaging follow up. Symptomatic patients presenting with hemorrhage, seizures, neurological deficits, or hydrocephalus secondary to obstructed Foramen of Monroe would require surgical intervention. Successful surgical resection is curative, with low recurrence rates; however, functional outcomes depend on lesion location and even after complete resection, persistent memory deficits may occur in patients due to underlying fornix injury.

The most common surgical approach is the transcranial transcallosal anterior interhemispheric route, which provides good exposure but carries risks of disconnection syndromes. Neuroendoscopic excision is a minimally invasive alternate option with potentially lesser complications, with a caveat concerning adequacy of intraoperative hemostasis (3,4). The role of radiotherapy in cavernomas remains controversial as it could increase the risk of lesion growth, recurrence and bleeding (2), and hence may only be offered for lesions which are inaccessible with recurring symptoms.

CONCLUSION

Septum pellucidum cavernomas are exceptionally rare intracranial vascular malformations. They are usually discovered incidentally, as in our patient, but can present with hemorrhage, hydrocephalus, or memory impairment. MRI plays a pivotal role in diagnosis, with susceptibility-weighted imaging enhancing detection. Conservative follow-up is the recommended norm for asymptomatic cases and surgical resection reserved for symptomatic or complicated lesions. We are reporting this rare benign entity having potential for morbid complications, with an intent to augment awareness amongst practitioners in fields of Neurosciences.

Patient Consent

Written informed consent was obtained from the patient for publication of the duly anonymised clinical details and images.

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