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Septum Pellucidum Cavernous Malformation

A rare entity

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A 30-year-old man with no medical background presented to the emergency department in 2022 at a tertiary care hospital, Muscat, Oman, with a one-week history of persistent headache and a few episodes of vomiting. There was no history of seizure, visual symptoms, neurological deficit or loss of consciousness. On examination, he was fully conscious and alert. His neurological examination was unremarkable. The ophthalmology examination was normal with no evidence of papilledema. Computed tomography (CT) showed a well-defined hyperdense lesion in the septum pellucidum bulging bilaterally to the lateral ventricles and minimal dilatation of the lateral ventricles with no signs of acute hydrocephalus. Magnetic resonance imaging (MRI) of the brain was done and revealed a well-circumscribed lesion in the septum pellucidum with a heterogeneous signal intensity and hypointense rim of hemosiderin and an internal bubbly appearance, forming a "popcorn" appearance in T2-weighted images. Significant blooming in T2* gradient echo images was present at the site of the lesion, denoting blood products **(Figure 1)**. There was no enhancement in the lesion, and there was no surrounding edema, significant mass effect, or intraventricular hemorrhage. These imaging findings favored the diagnosis of the septum pellucidum cavernous malformation. As there was no objective clinical evidence of raised intracranial pressure (ICP) or radiological evidence of acute bleeding or ventricular obstruction, he was managed conservatively. He remained asymptomatic, and a follow-up MRI

after eighteen months showed an interval reduction in the size of the lesion with a siderotic rim and internal old blood products **(Figure 2)**. The patient will have an annual MRI of the brain for next two years, and he was instructed to report any new symptoms promptly.

We present a case of septum pellucidum cavernoma, its imaging features, differential diagnosis, and treatment options. Septum pellucidum cavernoma is an extremely rare entity, with only several similar cases reported in the literature.

Informed written consent for the publication was obtained from the patient.

Comment

Cerebral cavernous malformations, or cavernomas, are one of the central nervous system (CNS) vascular malformations with an overall prevalence of 0.02-13%. Among CNS vascular malformations, it is the third most common after developmental venous anomaly (DVA) and capillary telangiectasia, forming 5–13% of cases.[1,2] CNS cavernomas can be sporadic or familial. Positive family history, prior radiotherapy, or head trauma are considered the major risk factors for acquired cavernomas.[1,3] Cavernomas can be solitary or multiple, especially in hereditary types. It can also be associated with other vascular malformations, most commonly is DVA.[1] Around 50% of CNS cavernomas are incidental. It has a wide range of clinical presentations depending on location, size, and associated complications like mass effect or hemorrhage.[2,4] 74 to 90% of cerebral cavernomas are seen in supratentorial location.[1,2] Headache, seizure, and focal neurological deficits are commonly associated with supratentorial cavernomas compared to bleeding, which is commonly seen in infratentorial cavernomas.[2]

Intraventricular cavernomas are rare and seen in less than 10% of cases.[5] Septum pellucidum cavernoma is an extremely rare subtype of intraventricular cavernoma.[1,2] Septum pellucidum is a thin midline anatomical sheet seen between the anterior horn of lateral ventricles and extending from the corpus callosum to the fornix, which is an important structure connecting the hippocampus and other parts involved in memory function.[1] Furthermore, bleeding within the septum pellucidum cavernoma may damage the fornix and lead to long-term memory impairment and anterograde amnesia.[1,4] Septum pellucidum cavernomas can also extend to either

side of lateral ventricle and may lead to hydrocephalus secondary to compression on the foramen of Monro.[4] Haque et al. published a review article and descriptive analysis of septum pellucidum cavernoma that included ten cases found in English literature.[1] The median age at diagnosis was 42 years and majority of them were males. The most common presentation was headache seen in 70% of cases and 20% presented with impaired memory. One third of the included patients presented with complications like hemorrhage and hydrocephalus.[1]

Neuroimaging plays an important role in the diagnosis. Large cavernomas are seen as hyperdense lesions on CT. Small cavernomas can be difficult to visualize on CT.[4] MRI, on the other hand provides superior assessment in these cases. MRI shows the characteristic "popcorn" or "berry" appearance with a rim of hemosiderin as seen in our patient.[1] The signal intensity is variable depending on the age of the blood products. In lesions that bled recently, surrounding oedema may be seen. T2* gradient echo images, or susceptibility-weighted imaging (SWI), is the best sequence for detecting small cavernomas, particularly in the hereditary type. The enhancement is variable, ranging from none to a moderate degree of enhancement.[1,2] On digital subtraction angiography, cavernomas are occult lesions due to the lack of arteriovenous shunts.[1]

The differential diagnosis for septum pellucidum cavernomas includes hematoma, arteriovenous malformation, colloid cyst, neurocysticercosis, or other neoplastic lesions, especially when associated with hemorrhage or calcifications like central neurocytoma, ependymoma, and subependymal giant cell astrocytoma.[1,2,5]

The management of CNS cavernomas depends on the location and symptoms. Asymptomatic lesions can be managed conservatively. Symptomatic lesions due to mass effect, epilepsy, focal neurological deficit, and repeated hemorrhage necessitate surgical intervention. Cavernomas of the septum pellucidum with established or imminent ventricular obstruction warrant prompt surgical intervention. Different surgical approaches were reported, but the most common method is transcranial transcallosal anterior interhemispheric approach.[1,2,4] Although neuroendoscopy is a minimally invasive procedure with fewer complications, its role in the excision of septum pellucidum cavernoma is questionable and unclear due to the possible difficulty of controlling the bleeding.[1] The role of radiotherapy is debatable and is used for symptomatic control of some

intraparenchymal cavernomas.[5] Some studies believe that radiation increases the risk of cavernomas bleeding, interval growth, and recurrence.[1,3] Complete surgical resection of septum pellucidum cavernomas is curative, with clinical improvement and no recurrence.[1,3] Few cases reported persistent memory impairments that were present at the time of diagnosis which could be attributed to irreversible damage to the fornix.[1,3]

Authors' Contribution

AA collected the clinical data, reviewed literature and drafted the manuscript. RK reviewed the clinical aspects of the case description and reviewed the manuscript. EA supervised the work, selected the representative images and critically reviewed the manuscript. All authors approved the final version of the manuscript.

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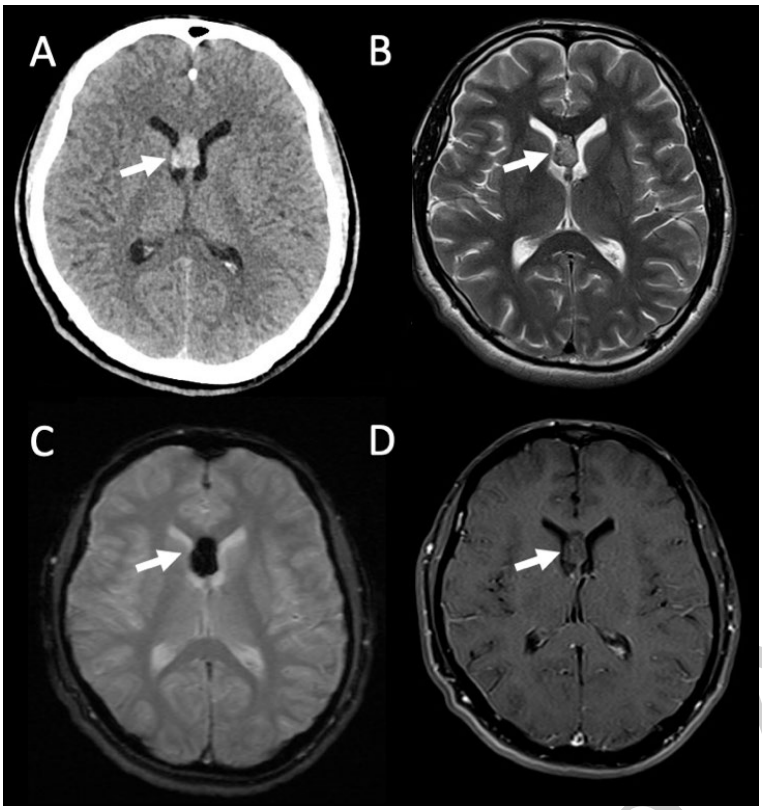

Figure 1: (A) Axial non-enhanced CT image shows a hyperdense lesion of the septum pellucidum at the level of the frontal horns bulging bilaterally to the lateral ventricles. **(B)** axial T2 weighted image shows the lesion to be T2 hyperintense with a bubbly appearance. **(C)** T2* gradient echo image shows significant blooming at the site of the lesion. **(D)** Post-contrast axial T1-weighted image shows no significant enhancement.

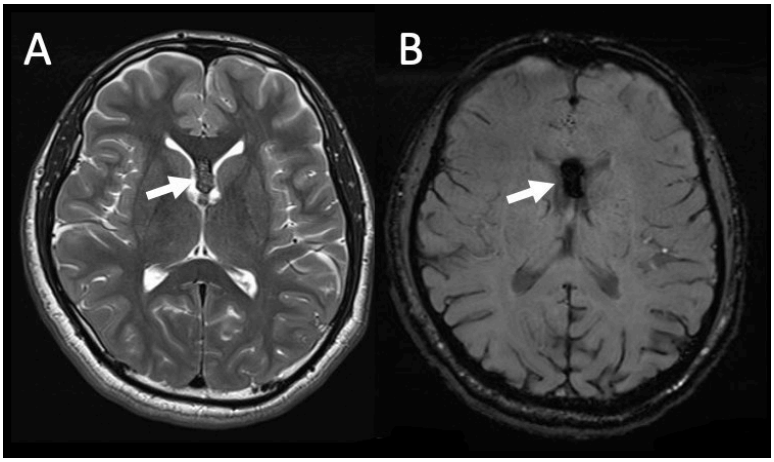

Figure 2: MRI follow-up after 18 months shows **(A)** interval reduction in the size of the lesion with a siderotic rim in axial T2, and **(B)** susceptibility effects due to old blood products in susceptibility-weighted imaging.