

Case Report

Septum Pellucidum Cavernoma: A Case Report and Anatomical Consideration of an Extremely Rare Lesion

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ABSTRACT

Cavernous angiomas (cavernomas) are vascular malformations of the brain characterized by abnormal capillaries. Ventricular cavernomas are considered rare; however, an extremely unusual topography is the septum pellucidum, with only five reported cases in the English literature. These malformations may rupture and cause very large hematomas, leading to neurological impairment. Cavernomas can be familial or sporadic; additionally, these may appear following brain radiotherapy in extremely rare cases.

Herein, we present an extremely rare and unique case of a septum pellucidum cavernoma that occurred in a young male who had previously undergone brain radiotherapy in childhood due to acute lymphoid leukemia. Following presentation, he was diagnosed with generalized seizures. The cavernoma was resected via an anterior interhemispheric transcalsal approach, following which the patient remained stable without neurological sequelae.

To conclude, ventricular cavernomas are rare lesions, especially when located at the septum pellucidum, thus constitute a challenging neurosurgical approach.

KEYWORDS: Cavernous hemangioma, Septum pellucidum, Radiotherapy, Cerebral ventricle neoplasm, Cerebral hemorrhage

INTRODUCTION

Cavernous angiomas (cavernomas) are rare lesions of the central nervous system (CNS) categorized as vascular malformations (5%–13%) (1). They may present as brain hemorrhage and seizures in young adults; in these patients, cavernomas generally occupy the brain parenchyma or ventricular system (2). Cavernomas may occur in familial or sporadic forms (6). Further, de novo growth has been reported, especially in pediatric patients with a history of CNS radiotherapy (6).

Involvement of the ventricular system is very rare (2.5%–10.8%) (6). However, a unique and rare topography is the septum pellucidum, which is surgically considered challenging. To date, only five cases have been reported in the English literature (1,4,5,7,8) (Table I). In this report, we describe the sixth

case of this rare entity in a patient without a familial history of cavernoma who had undergone whole-brain radiotherapy in his childhood.

CASE REPORT

A 19-year-old male presented to a neurosurgical outpatient clinic after experiencing brain trauma injury and seizures. Neurological examination revealed normal findings; however, brain computed tomography revealed a lesion with a ventricular topography. The patient's medical history included diagnosis of acute lymphoid leukemia (ALL) that was treated 14 years prior using chemotherapy and radiotherapy.

At that time, the patient was 5 years old and had undergone prophylactic whole-brain irradiation. Notably, CNS investigation did not reveal any involvement – cerebro spinal fluid, and

Table I: Reported Cases of Septum Pellucidum Cavernoma in English Language

Author/Year	Age	Sex	Clinical Feature	Family History	CNS Radiotherapy	Radiology	Surgical Approach
Kasliwal and Sharma, 2011 (4)	55	M	Headache	No	No	MRI brain showed heterogenous lesion on T1WI and T2WI, blooming on Gradient echo	Right frontal transcortical
Katoha M et al., 2013 (5)	44	M	Memory disturbance	No	No	MRI showed T2 WI hyperintense contrast enhancing lesion	Anterior interhemispheric transcallosal
Muzumdar D et al., 2015 (7)	35	F	Headache	No	No	MRI showing T2 mixed intensity T2-weighted arising from the inferior aspect of the septum pellucidum	Anterior interhemispheric transcallosal
Faropoulos K et al., 2015 (1)	56	M	Headache and lassitude	No	No	T1-weighted MRI showed a mixed signal mass; T2-weighted image a high signal image with no surrounded low signal rim	Right frontal craniotomy
Picolas C et al., 2016 (8)	58	M	Headache, personality and memory changes	No	No	MRI showed an oval tumor with mixed dispersed iso- and hyperintense nodular loci in T1 sequence and hypointense with dispersed nodular hyperintense loci in T2.	Inferior parietal transcortical transventricular
Paiva A et al. (present case)	19	M	Epileptic seizure	No	Yes	T1-weighted MRI showed a mixed signal mass at septum pellucidum without hydrocephalus	Anterior interhemispheric transcallosal

CNS: Central nervous system, **F:** Female, **M:** Male, **MRI:** Magnetic resonance imaging.

magnetic resonance imaging (MRI) findings were normal. In 2002, the total radiation dose was 18Gy administered over 12 days. Based on the chemotherapy strategy, he received cycles of methotrexate treatment in 2002, which was repeated in 2004.

For a comprehensive investigation of this lesion, the patient underwent MRI, which revealed the presence of a nodular lesion with a heterogeneous sign on T1-weighted, T2-weighted, and FLAIR modalities; the lesion exhibited septum pellucidum topography (Figure 1A, B). The initial hypothesis was a cavernoma based on these MRI characteristics.

An anterior interhemispheric transcallosal approach was used for corrective surgery (Figure 2). Histopathological analysis confirmed the suspicion of a cavernoma. The patient continued to exhibit no neurologic deficits. MRI performed during the first postoperative 24 hours confirmed complete removal of the lesion (Figure 3). After 60 hours of observation, the patient was discharged.

Gradually, anticonvulsant administration was withdrawn, and the patient remarkably showed no signs of epilepsy. A subse-

quent neuropsychological evaluation revealed no changes in memory relative to that determined in the preoperative evaluation.

■ DISCUSSION

Cavernomas are CNS vascular malformations comprising clusters of abnormal capillaries surrounded by hemosiderin deposits and a gliotic margin (6). These lesions exhibit a high risk of bleeding and serve as a differential diagnosis for parenchymal hemorrhage, particularly in younger patients. Ventricular bleeding may be another catastrophic manifestation (4,6).

Occasionally, large lesions may be asymptomatic. Seizures, headache, and focal neurologic deficits comprise a subset of manifestations. Lesions in supratentorial compartments exhibit a prevalence of 70%–80% (6). However, bleeding is more common in infratentorial cavernomas, with a prevalence of 3.8% relative to the prevalence of supratentorial cavernomas (0.4%) (6). These malformations can be sporadic or occur in relation to an inherited pattern. Approximately 40%–60% of

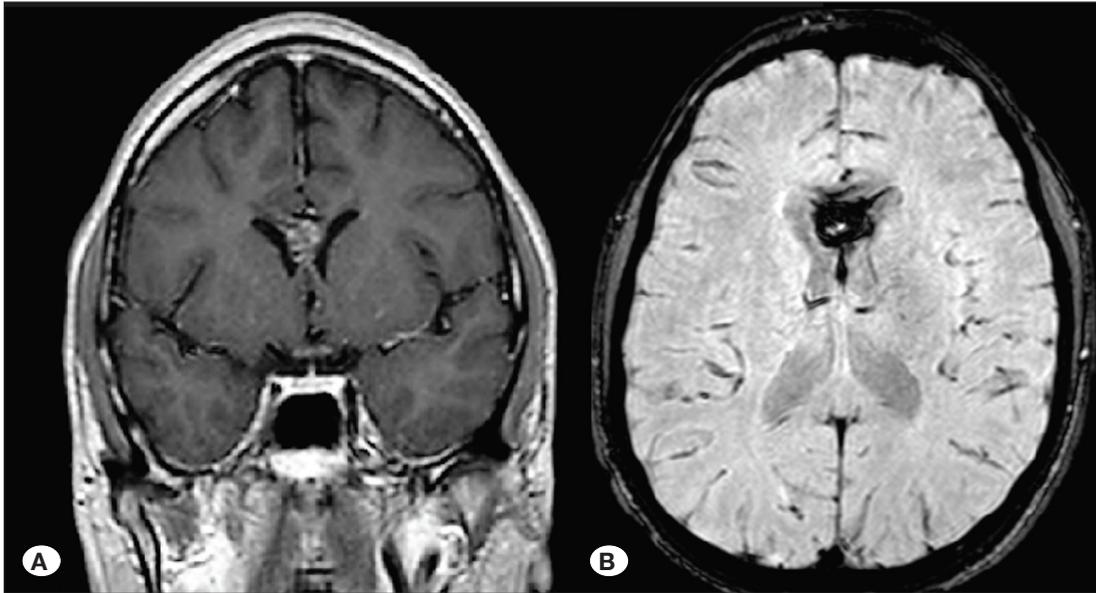


Figure 1: **A)** T1W contrast enhanced coronal MRI showing a heterogeneous lesion at septum pellucidum topography; **B)** axial SWI MRI showing signs of previous bleeding – heterogeneous lesion with hemosiderin inside.

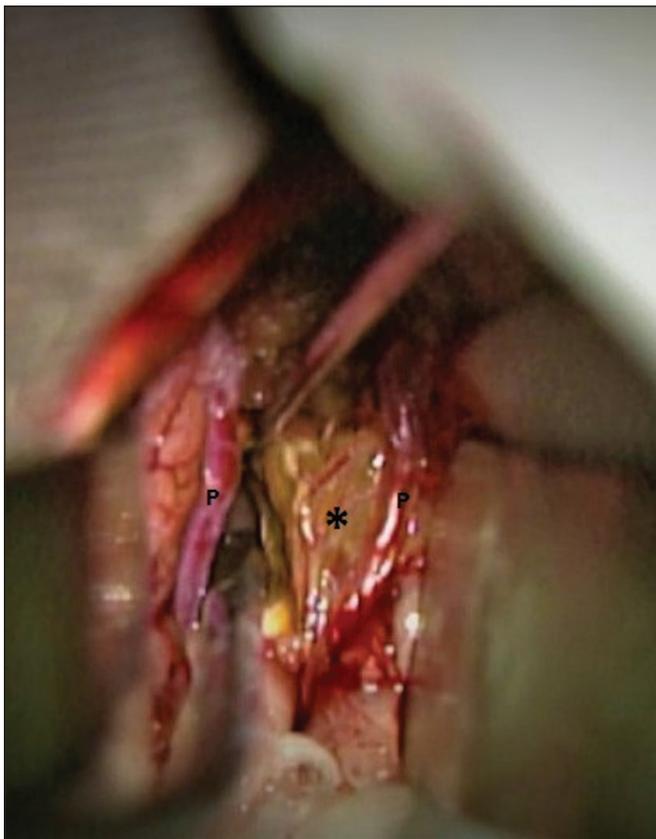


Figure 2: Microscopic intraoperative feature revealing a yellowish lesion; *: cavernoma; P: posterior choroidal arteries.

cavernomas occur in a familial form; notably, these types often involve multiple cavernomas (6).

Regarding sporadic cavernomas, there may be a relationship between the application of radiotherapy and onset of cavernoma, primarily in children who undergo cranial radiation in

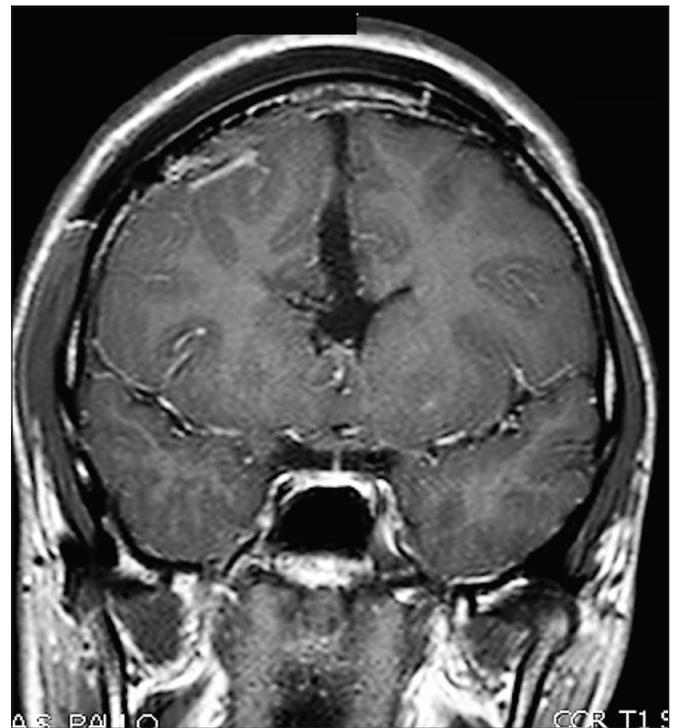


Figure 3: Immediate post-operative T1- weighted contrast enhanced coronal MRI showing complete resection.

childhood; in fact, most instances of radiotherapy-associated cavernomas are related to the treatment of hematologic malignancies (2). There are some prior reports in the literature, and a relationship between radiotherapy dose and the risk of developing a radiation-induced cavernous hemangioma (RICH) has been proposed (3,6). The primary hypothesis regarding the mechanism underlying RICH physiopathology is that RICH develops from a de novo response to radiation, thereby inducing histological changes in brain parenchyma.

Currently, there is a lack of available information regarding the behavior of such lesions because of the small number of case reports on this peculiar entity.

The patient described in this report had previously undergone prophylactic whole-brain radiotherapy in his childhood as a treatment for ALL, during which neurological involvement was not observed. Therefore, the septum pellucidum cavernoma might be a result of radiation exposure and be appropriately classified as RICH.

MRI is the gold standard neuro imaging modality for cavernoma diagnosis, in so far as these lesions are angiographically occult malformations. Nevertheless, developmental venous anomaly typically associated with and is visible during angiography and MRI (6). In the parenchymal form, a hypointense ring is observed on T1- and T2-weighted modalities due to hemosiderin deposits from recurring microhemorrhages (6).

Because typical images have not been established, ventricular cavernomas exhibit several differential diagnoses, including arteriovenous malformations, astrocytomas, choroid plexus papillomas, ependymomas, and colloid cysts (1). In some reports, biopsy was initially performed because MRI results supported a hypothesis of brain tumor (5). They are less frequent (2.5%–10.8%) (5), and the first description was done in 1905 by Finkelburg (2,6). Regarding ventricular topography, lateral ventricles are most commonly affected (5). In this patient, the initial MRI was highly suggestive of a cavernous angioma. An anterior transcallosal interhemispheric approach was found to be an appropriate surgical approach for the lesions in this topography because it provides good visualization of the anterior ventricular structures (Figure 2).

Furthermore, septum pellucidum cavernomas are extremely rare ventricular lesions; as noted above, only five cases have been reported in the English literature (1,4,5,7,8) (Table I). Cavernomas of the septum pellucidum do not exhibit any specific clinical presentation; however, patients might demonstrate memory disturbances and the signs and symptoms of hydrocephalus owing to the obstruction of the foramen of Monro (6).

While using neurosurgical approaches, damage to neighboring structures should be minimized, particularly the structures related to memory, such as the fornices, emphasizing the criticality of surgical route selection. The anterior transcallosal approach is adequate and is often superior to the ventricular transcortical approach for midline lesions without hydrocephalus as well as in cases where in the frontal lobe thickness is greater than 30 mm, as observed in the present case (9). Therefore, the anterior transcallosal approach (Figure 2) was used, following which the patient did not exhibit neurologic deficits.

■ CONCLUSION

In young patients, cavernomas constitute an important type of vascular malformation associated with brain hemorrhage. Although rare, their ventricular forms should be considered in differential diagnosis during MRI evaluation. The septum pellucidum is an extremely rare topography; considering its proximity with noble structures, the corrective surgical approach must be judiciously planned. In patients who have previously undergone brain irradiation, the diagnosis of cavernoma must be strongly considered, assessing its importance as noted in the current report and similar reports.

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