Fifth ventricle with bitemporal hemianopsia

Case report

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Symptomatic cysts of the septum pellucidum (fifth ventricle) are rare, and they are not usually considered among the causes of chiasmal compression. This report describes a case with a noncommunicating cyst of the septum pellucidum that presented with signs of chiasmal compression.

KEY WORDS □ septum pellucidum □ cyst □ chiasmal compression □ bitemporal hemianopsia

C AVITIES within the septum pellucidum are not uncommonly found at autopsy, and they do not usually have a clinical significance. The incidence of cavum septi pellucidi in adults is about 15%. Symptomatic cysts of the septum pellucidum occur only rarely. Dandy was the first to call attention to the pathological cavum in his report of two cases. He called it “congenital cerebral cyst of the cavum septi pellucidi” or fifth ventricle when it was anterior, and “cavum vergae” or sixth ventricle when it was posterior to the fornix. Fifteen cases of symptomatic septum pellucidum cysts were cited from the literature in 1969 by Shaw and Alvord, in addition to their two new cases. Since then, sporadic cases of cysts of the cavum septi pellucidi have been reported.

The purpose of this paper is to present an unusual case with a cyst of the septum pellucidum. To our knowledge, this is the first case reported in the English literature that presented with symptoms of chiasmal compression.

Case Report

This 8-year-old boy complained for 4 months of headaches and slowly deteriorating vision in both eyes. One year previously, he had started to watch television closer than usual.

Examination. There were no signs or symptoms of hypopituitarism. Neuro-ophthalmological examination revealed visual acuity of 6/15 in the right eye, and 6/12 in the left eye. He could not identify any of the numbers on Ishihara color plates. There was a bitemporal hemianopsic visual field defect. Both discs were pale, more on the right than the left. The remainder of the neurological examination was normal. Complete blood count, urinalysis, kidney and liver functions, and chest and skull x-ray films were normal. Computerized tomography (CT) scan revealed a faint demarcation of a central cystic mass in the area of the septum pellucidum. Absorption values were similar to the density of cerebrospinal fluid (CSF). The lateral ventricles were normal in size (Fig. 1 left). There was no enhancement with the injection of urographin 76%. Conray ventriculography was performed to delineate the area of this cystic mass. The opening pressure was 90 mm H2O. The CSF protein was 120 mg%, and glucose was 50 mg%. On the x-ray films the contrast material was seen filling the cyst; there was no communication with the ventricular system (Fig. 2). While the contrast material was in the cyst, CT sections were obtained, and revealed a midline cystic mass between the lateral ventricles just above the third ventricle (Fig. 1 right).

Operation. The chiasmal area was exposed through a right frontal craniotomy and extramedullary exploration. The chiasm was compressed by the third ventricle, which seemed in turn to be compressed by pressure from above. A right frontal transcortical incision disclosed the foramen of Monro and the lateral ventricle, with a large cystic mass of the septum
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**Discussion**

The septum pellucidum is a double membrane that separates the anterior horns of the lateral ventricles. The term “cavum septi pellucidi” is used for the non-dilated cavity with or without an opening to the ventricular system. A frankly dilated septum is referred to as a “cyst of the septum pellucidum.” Cysts of the septum pellucidum are of two varieties: 1) incidental, asymptomatic, communicating cysts, and 2) pathological, symptomatic, noncommunicating types with increased pressure in the cyst.

According to Shaw and Alvord, cavum septi pellucidi rapidly closes 2 months after birth, usually leaving only a small cavity just behind the corpus callosum. It is not explained why it persists in some persons. Liss and Mervis reported that these cavities in children are lined by unspecialized astrocytes, but in adults they may become lined by cells identical to the ependymal cells that line the ventricles. They suggested that there is an age-related evolution or transition from glial cells to ependymal cells. All of these observations suggest dynamic changing characteristics of the normal septum pellucidum, which may explain the formation of the cyst. A closed septum pellucidum does not admit air from the ventricular system, and appears as a widened septum. If fluid accumulates within a closed cavum septi pellucidi, it may behave as an intraventricular mass and its manifestations result from compression or distortion of neighboring structures. It occurs in association with symptoms such as mental retardation, headache, seizures, unilateral sensory motor dis-
orders, hydrocephalus, and other anomalies of neural tube development. Signs and symptoms produced by septal cysts may resemble those associated with tumors of the septum pellucidum.

Pressure from above the chiasm is seen characteristically in association with suprasellar lesions such as meningiomas, stalk tumors, gliomas, and aneurysms. An enlarged third ventricle may also act as a suprasellar mass and produce a chiasmal syndrome (a bitemporal hemianopsia and bilateral optic atrophy). The presence of the fifth ventricle is an unusual cause of chiasmal compression, and it is usually not considered in the differential diagnosis. In the present case, a large cyst of the septum pellucidum (fifth ventricle) without enlargement of the third ventricle was found compressing the chiasm, causing bilateral optic atrophy and bitemporal hemianopsia.

References

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