

CASE REPORT

CAVUM SEPTI PELLUCIDI AND CAVUM VERGAE CYST IN MAN

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ABSTRACT

Introduction: The cavum septi pellucidi (CSP) and cavum vergae (CV) are cystic anomalies of septum pellucidum.

Case: A 30-year-old man, which revealed cavities at the anatomical location of the CSP and CV on brain magnetic resonance imaging (MRI) because of recurrent headache.

Conclusions: The CSP and CV occur during the developmental process of the brain which regresses between the seventh month of intrauterine life and the second year of postnatal life. Persistence of these structures does not cause any symptoms, but sometimes related to malformations and psychiatric disturbances, mainly dependent on size. In this case, developmental disturbance is the underlying cause.

Key words: Septum pellucidum, cavum septi pellucidi, cavum vergae, midline cavities.

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INTRODUCTION

Septum pellucidum refers to a thin translucent plate of two laminae that parallels the interhemispheric fissure. It forms the medial wall of both lateral ventricles and extends from the lamina terminalis to the splenium of the corpus callosum. A cavity between two leaflets of septum pellucidum is called cavum septum pellucidum (CSP). The cavity is not connected to leptomeningeal or ventricular spaces. The presence of a cavum septum pellucidum represents a normal anatomical variant which is usually asymptomatic. The possible cause of the existence of this cavity is a developmental disturbance or an acquired abnormality.

CASE REPORT

A 30-year-old man underwent a brain MRI examination because of recurrent headache without trauma history. Neurological examination was normal. There were no psychiatric disturbances in this patient. The brain MRI which included T1-T2-weighted and flair sequences in all three planes (sagittal, axial, and coronal) revealed cavities in front of the fornix and extended to the caudal segment of septum pellucidum (figures 1 and 2).

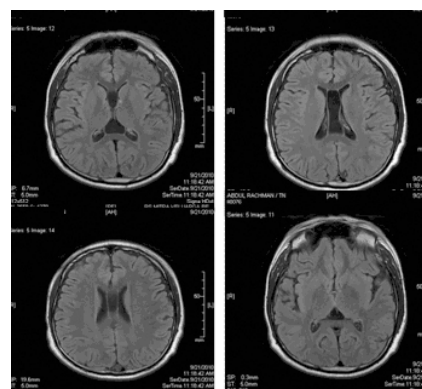


Figure 1. The Brain MRI : Axial T1. Arrow : CSP and CV.

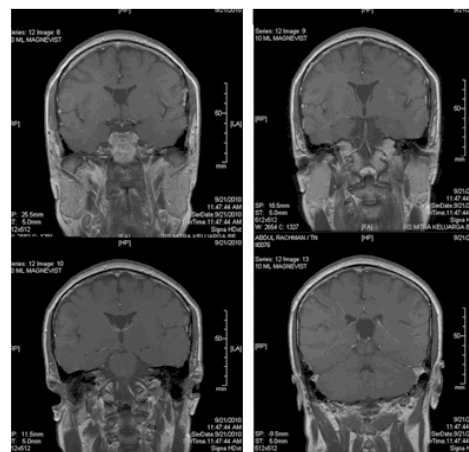


Figure 2. The Brain MRI: Coronal T1. Arrow : CSP and CV.

DISCUSSION

In most children and adults, septum pellucidum is a single, thin, vertically orientated structure that forms medial border of the frontal horns and bodies of the lateral ventricles and extends from the corpus callosum to the columns of the fornix. The septum pellucidum is usually 1 to 3 mm thick and consists of two fused leaves that were separated during fetal life.¹

Septum pellucidum is a part of the limbic system, with prominent connections in both medial (or Papez) limbic circuit and basolateral limbic circuit. This provides direct and indirect afferent and efferent connections to the hippocampus, cingulate gyrus, medial frontal cortex, anterior nucleus of the thalamus, mamillary bodies, hypothalamus, and amygdaloid nuclei. Septum pellucidum is also play as a relay station connecting the hypothalamic autonomic system to hippocampus, amygdala, habenula and brain stem reticular formation. However, the exact function is not completely understood.^{2,3}

During intrauterine life there are three potential midline cavities among the ventricles in the cerebrum. These midline cavities occur during the developmental process of the brain which regress between the seventh month of intrauterine life and the second year of post-natal life. They are normally present during fetal life and in certain proportion of the adult population, thus they are called "persistent primitive structures". As these three cavities in the cerebrum are not lined by ependyma or choroid plexus cells, they do not produce cerebrospinal fluid and they are not considered as part of the ventricular system.⁴ Persistence of these midline cavities from anterior to posterior is called Cavum Septum Pellucidum (CSP), Cavum Vergae (CV) and Cavum Velli Interpositi (CVI) (*figure 3*).

Cavum Septum Pellucidum (CSP) is also called the fifth ventricle.⁵ CSP is the potential space which develops when the two leaflets of a septum pellucidum fail to fuse. Embryologic development of CSP is intimately associated with corpus callosum.⁶ CSP is triangular and demarcated anteriorly by the genu of corpus callosum, posteriorly by the corpus and columns of fornix, superiorly by the body of corpus callosum, and inferiorly by the rostrum of corpus callosum.⁷

Cavum Vergae (CV) is also called the sixth ventricle. It is bounded superiorly by the body of corpus callosum, inferiorly by the hippocampal comisure, laterally by the crus of fornix, and posteriorly by the splenium of corpus callosum.

The cavum is a potential cavity and therefore significant enlargement is best termed a cavum septum pellucidum cyst or cavum vergae cyst. The cavum septi pellucidi and the cavum vergae usually communicate with each other and obliterate from posterior to anterior, the posterior cavum vergae obliterating first and then usually the anterior cavum septi pellucidi.

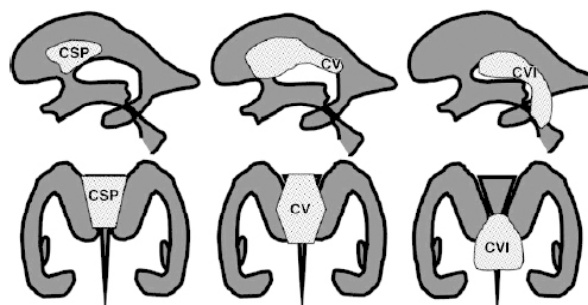


Figure 3. Midline cavities and their positions in the sagittal plane (top) and coronal plan (bottom). The shaded areas represent the fluid-filled spaces, which include the cavum septi pellucidi (CSP) (A), cavum vergae (CV) (B), and cavum veli interpositi (CVI) (C).²

CSP is noted in from 0.14 to 18.9% of cases and CV is less frequently from 0 to 1.3%.^{8,9} Wang et al reviewed the computerized database of 54,000 patients and found 0.04% involving a dilated cyst of the CSP.¹⁰

Cysts of the cavum septum pellucidum have been classified into two major groups. Incidental (asymptomatic) and pathological (symptomatic). The pathological cava are comparatively rare, are always non-communicating, and may be associated with other structural or inflammatory brain disease. In boxers the incidence of the CSP was 77%, which is significantly higher than in the general population.¹¹

The CSP is well known to the fetomaternal experts and represents a trusted landmark of normality in development of the midline telencephalic structures and it has been associated with a variety of neuropsychiatric disorder.^{12,13} Disease of the cavum septum pellucidum could therefore be expected to cause symptoms either by mass effect or by disturbance of the emotional and behavioural functions of the limbic system.

Persistence of these structures does not cause any symptoms but is statistically related to malformations and psychiatric disturbances, mainly dependent on size (*table 1*).

Expanding cysts of the septum pellucidum, although rare, may be a cause of significant neurological dysfunction. Most become symptomatic as a result of obstruction of the interventricular foramina and produce

	Measurement by Ultrasonography or MRI in controls	Size	Reference
36-40 weeks	Width	<9.5 mm	Mott et al 1992
Fetal age	Length	<8.0 mm	
Newborn 39-40 weeks age	Avarage width	6.3 mm ± 0.83 (1 SD)	Jou et al 1998
Children and adults	Maximum length	<6 mm	Nopoulos et al 1998
		<6 mm	Born et al 2004
		<5.6 mm	Takahashi 2008

Table 1. Midline cavities in normal persons.¹⁴

headaches, papilledema, emesis, and loss of consciousness. Behavioral, autonomic, and sensorimotor symptoms occur when an expanding cyst impinges on the structures of the hypothalamoseptal triangle or impairs the deep cerebral venous drainage. Neuroophthalmological symptoms may develop as a consequence of hydrocephalus or direct compression of visual structures. Wang et al found that 7.43% patients had acute episodic headache, 31.3% had chronic daily headache (CDH) with acute onset, and 4.25% had CDH with insidious onset.¹⁰ Silbert et al reported five patients presented with symptoms related directly to pressure effects from their cavum septum pellucidum with persistent or intermittent obstructive hydrocephalus. The most characteristic presenting symptoms were intermittent postural headache and postural loss of consciousness.¹⁵

CSP size was not significantly associated with the comorbid diagnoses of obsessive-compulsive disorder (OCD) or attention-deficit/hyperactivity disorder (ADHD). The presence of a cavum vergae was not significantly associated with a diagnosis of Tourette syndrome (TS).¹² Patients with schizophrenia have been reported to have a higher frequency of enlarged cavum septi pellucidi (CSP) in comparison with normal subjects.¹⁶ According to Rajarethinam R et al, they were doubt on the significance of CSP as markers of neurodevelopmental pathology in schizophrenia.¹⁷ Filipović et reported that CSP is more frequent, longer and wider in persons who are suffering from schizophrenia, alcohol addicts and those who had sustained one or several head blows in the past.¹⁸

Presence of CSP is statistically higher in epilepsy patients than in control group. This indicates that the presence of CSP may not be a simple normal variation, and it can be considered a developmental anomaly that

may contribute to epileptogenesis.³ A study by Varsik P et al was focused on frequent simultaneous occurrences of epileptic seizures and the defect or abnormality of the ventricular system - cavum septi pellucidi (CSP). They believed that there is an important role for septal and diencephalic midline structures in cerebral electrogenesis, and possibly in the origin of epileptic seizures.¹⁹

CONCLUSIONS

In this case, developmental disturbance is the underlying cause. The leaves of the septum form cavity in the medial inferior commissural plate derived from the primitive lamina terminalis at the rostral end of the neural tube and followed by posterior extension of the cavum septum pellucidum during the formation of corpus callosum. It is impossible to correlate the recent headache suffered by patient with the old standing cyst that could have been responsible for a compression of the neighboring neural structures.

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