Some Observations on the Septum Pellucidum

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Key Words

Septum pellucidum · Cavum vergae · Cysts · Septo-optic dysplasias

Abstract

The thin, vertically placed partition consisting of two laminae separated by a narrow chink constitutes the cavity of the septum pellucidum, known from the time of Sylvius. Traumatic lesions in boxers, rare congenital expanding cysts causing hydrocephalus, and a number of septo-optic dysplasias give clinical significance to the septum and its related cavum.

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The septum pellucidum is not just an anatomical structure of academic curiosity. Traumatic lesions in boxers, rare congenital expanding cysts causing hydrocephalus, and a number of septo-optic dysplasias give clinical significance to the septum and its related cavum.

Anatomy

The septum pellucidum is a thin, vertically placed partition consisting of two laminae, separated by a narrow chink, the cavity of the septum pellucidum. The original description is attributed to Franciscus de Le Bo Sylvius

[1, 2] in 1671. The Italian anatomist Andrea Verga (1811–1895) in 1851 recorded a fluid-filled space [3], the posterior extension of the cavum septi pellucidi. In a distinguished anatomical paper, Elliot Smith in 1895, observed:

‘The stretching of the thickened mass of the lamina terminalis by the extending arc of the dorsal commissure produces the septum pellucidum; and as a physical expression of the antagonism between the extension of the commissure and the retrogression of its “matrix” the cavum septi pellucidi or fifth ventricle appears’ [4; p. 193].

The lateral ventricles communicate through the inter-ventricular foramen with the third ventricle, but are separated by the septum pellucidum and cavum septum pellucidum, which do not communicate with the ventricles. Walter Dandy described the nomenclature as follows [5]:

‘The nomenclature, however, is not uniform. For example, the cavum septi pellucidi is perhaps better known as the fifth ventricle, and the cavum vergae is called Verga’s ventricle, the sixth ventricle, the ventricle of Strambio, ventriculus fornicis, ventriculus triangularis, and the canal aqueduct...’

The cavum septi pellucidi is bounded (fig. 1) by: anteriorly, the genu of the corpus callosum; superiorly, the body of the corpus callosum; posteriorly, the anterior limb and pillars of the fornix; inferiorly, the rostrum of the corpus callosum and the anterior commissure. Viewed laterally, the cavum septi pellucidi is roughly triangular with the base at the corpus callosum.

Fig. 1. Diagram of relations of septum pellucidum.

Development

The leaves of the septum form as a result of cavitation of the medial inferior commissural plate derived from the primitive lamina terminalis at the rostral end of the neural tube, during the formation of corpus callosum [6]. The leaves enclose the cavum septum pellucidum and appear during the 3rd month of intrauterine growth by secondary cleavage. By 6 months, starting posteriorly and progressing rostrally, the leaves begin to fuse. However, the cavum septi pellucidum and cavum vergae (the posterior extension of the cavum septum pellucidum) are frequently seen in premature and term infants. They seldom enlarge to become symptomatic. Because the cavum vergae does not contain cerebrospinal fluid, it is incorrect to label it as the sixth ventricle [7].

Variations and Incidence

Liss and Mervis [8] reported three variations. The first (50%) is the single midline membrane with an ependymal lining on each *ventricular* surface. The second (25%) consists of two separate but closely apposed leaves that delineate a potential space. The third (25%) is the cavum septum pellucidum in which the leaves of the septum are separated by a variable space. Congenital absence of the septum pellucidum occurs in 2–3 per 100,000 people in the general population. Congenital perforations of the septum pellucidum may be present. The incidence of the cavum vergae depends on the technique (ultrasound, CT, MRI, autopsy, etc.) used to demonstrate it. Pathologically, it was found in 20.3% of 1,032 brains [9].

Table 1. Lesions involving the septum pellucidum

Agnesis
Cysts
Trauma (boxers; other head injuries)
Septo-optic dysplasia
Apert syndrome
Cavernous angioma
Central neurocytoma
Astrocytoma
Association with schizophrenia, alcoholism

The asymptomatic cavum septum pellucidum is not a malformation or a true cyst; it has been recognized since Sylvius as a common incidental finding; it may be associated with a cavum vergae or cavum velum interpositum. Using ultrasonography, Mott et al. [10], demonstrated a cavum septum pellucidum in all normal infants less than 36 weeks gestational age and in 36% of full-term infants. Schwidde [11] found a cavum septum pellucidum in 20.3% of 1,032 formalin-fixed brains; MRI studies show a similar incidence.

Clinical Significance

Resulting from physical acceleration-deceleration forces, there is a higher incidence in professional boxers [12, 13]. Corsellis et al. [14] found it in 12 of 13 professional boxers' brains. The cavum in boxers showed a characteristic fenestration with detachment of the fornix from the undersurface of the corpus callosum, with the two flattened forniceal bodies splaying out horizontally.

Symptomatic cysts and giant cysts (communicating with the ventricles or noncommunicating) of the cavum septum pellucidum (table 1) and cavum vergae have been reported. An expanding septum pellucidum cyst does not communicate with the ventricular or cisternal space and can secrete fluid internally [1]. Thin-walled cysts may rupture. Dandy [5] described the clinical and radiographic evaluation, surgical exploration, and subsequent clinical improvement of a patient with an expanding septum pellucidum cyst (case 2). Silbert et al. [15] described 5 patients with persistent or intermittent obstructive hydrocephalus as a result of obstruction of the interventricular foramina. The presenting symptoms were intermittent postural headache and postural loss of consciousness. Behavioural, autonomic, neuroophthalmological, and sensorimotor symptoms may occur when an expand-

ing cyst impinges on the structures of the hypothalamo-septal triangle or impairs the deep cerebral venous drainage.

Amin [16] similarly described a patient with symptomatic hydrocephalus from a cyst of the septum pellucidum with intermittent obstruction of the foramina of Monro. Endoscopic pellucidotomy [17] or fenestration re-establish ventricular drainage with symptomatic relief. Shunt procedures are occasionally needed.

Familial examples have been described with macrocephaly, mental retardation and epilepsy [18]. The term 'septo-optic dysplasia' was coined in 1956 by de Morsier, who showed an association of optic nerve hypoplasia and absence of the septum pellucidum (gene map locus

3p21.2–p21.1, $\pm$ *Hesx1* mutations¹) [19]. Others have reported an association with pituitary dwarfism.

Statistical associations suggest that a schizophrenic illness [20, 21] or attention deficit syndrome may relate to such cystic enlargements or callosal malformations. However, a well-controlled study showed cavum septum pellucidum while prevalent, was not more frequent in a sample of patients with schizophrenia, and had few associations with symptom severity or neuropsychological deficits [22].

¹ *Hesx1* is expressed at the anterior extreme of the rostral neural folds, finally resolving to the ventral diencephalon. It is expressed in the oral ectoderm which gives rise to Rathke's pouch.

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