Neuroepithelial (colloid) cysts of the septum pellucidum

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The authors present two cases of neuroepithelial (colloid) cysts found above the diencephalic roof, occupying the space between the two fornices and the two leaves of the septum pellucidum, and describe the clinical history, neurological, and neuroradiological findings. Both lesions were removed through a right transventricular-transseptal approach. No other report of such a lesion in this location could be found in the review of literature. The pathogenesis of these cysts, both above and below the diencephalic roof, is discussed in light of the developmental anatomy of the area under consideration. In view of their origin from the neuroepithelium in the diencephalic roof, whether by a process of invagination into the third ventricle or by evagination into the velum interpositum, the so-called colloid cysts are more appropriately termed neuroepithelial cysts.

**KEY WORDS** □ neuroepithelial cyst □ colloid cyst □ septum pellucidum □ pathogenesis

**EVER** since the first report on a colloid cyst by Wallmann in 1858¹ and the first successful removal of this lesion by Dandy in 1922² the diagnosis and surgical treatment of colloid cysts have received wide attention in the literature.¹⁴,⁹⁻¹¹,²⁰ Perhaps even greater interest has been shown in the still somewhat uncertain pathogenesis of this lesion.²,⁵,⁷,⁸,¹²,¹³,¹⁵,¹⁶,²¹⁻²³ The purpose of this paper is to summarize two cases of colloid cysts that presented above the third ventricle and to explain the origin of these cysts in light of the developmental anatomy of the diencephalic roof and of the structures surrounding it.

**Summary of Cases**

The two patients were both women, aged 40 and 26 years. The older patient had a long history of petit mal seizures while the younger one exhibited manic-depressive episodes for a few years prior to admission. Both patients complained of intermittent and increasingly severe headaches for about 1 year prior to admission. The immediate reason for neurosurgical evaluation was the development of bilateral papilledema in both patients.

The neurological examination revealed no additional abnormalities in the older patient. When first seen, the younger patient was somewhat obtunded, irritable, had generalized hyperreflexia and bilateral Babinski signs. The skull x-ray films, electroencephalogram, and brain scan were of no localizing value in either patient, but bilateral cerebral arteriograms showed evidence of marked lateral ventricular dilatation. Positive contrast ventriculogram revealed a complete
obstruction of the right foramen of Monro in the older patient and a partial obstruction in the younger one (Fig. 1); in view of these findings both patients required ventriculoperitoneal shunts. After their neurological status improved, a pneumoencephalogram was obtained in both patients; in each instance it revealed an oval mass expanding the septum pellucidum (Fig. 2).

The lesions were exposed through a right transfrontal transventricular approach, with the aid of microsurgical techniques. Operative findings were similar in both cases with the posterior part of the septum pellucidum protruding markedly into the lumen of the right lateral ventricle. The foramen of Monro could therefore not be visualized. After the right leaf of the septum pellucidum was incised, a smooth, glistening cystic lesion was found occupying the space between the two fornices and separating the leaves of the septum pellucidum. It appeared firmly attached to the upper surface of the velum interpositum. The cyst content, a grayish-greenish gelatinous material, was evacuated. The cyst wall was then freed from the rostral portion of both internal cerebral veins and removed together with the rostral portion of the vela interpositi. The integrity of both fornices was preserved. Both patients had an uneventful postoperative course with no neurological deficit resulting from the operation.

The histological examination of the cyst wall in the older patient revealed a stratified, columnar, cilia-containing epithelium. On its outer surface the cyst wall was covered by a fibrovascular stroma from the vela interpositi and by portions of the choroid plexus (Fig. 3). The microscopic examination of the cyst wall in the younger patient was not obtained, since only the colloid material removed reached the laboratory.

Discussion

No other report of a neuroepithelial (colloid) cyst in this location could be found in the literature. We reviewed the embryology of the diencephalic roof in an attempt to explain the controversy surrounding the origin of these cysts. By the sixth week of gestation, five distinct cerebral vesicles can be recognized at the cephalic end of the neural tube. The most rostral ones are the diencephalic and the two hemispheric vesicles sprouting rapidly from the lateral walls of the diencephalon (Fig. 4, a and b). Their roof is covered by a layer of mesoderm which later transforms into the pia-arachnoid membranes. The initially slit-like communication between the diencephalic and the lateral ventricles enlarges anteriorly to form the foramen of Monro. While the lateral diencephalic wall and the opposing wall of the hemispheric vesicles undergo a rapid proliferation to form the thalamus and
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Fig. 2. Pneumoencephalograms showing expansion of the septum pellucidum, in the older (left) and younger (right) patient. In each instance the pneumoencephalogram was performed after a ventriculogram was obtained (Conray and Pantopaque, respectively) and ventriculoperitoneal shunts were inserted.

Fig. 3. Left: Microscopic section of the cyst wall includes portions of the surrounding choroid plexus. H&E, × 25. Right: Stratified and columnar epithelium of the cyst wall, containing cilia. H&E, × 400.

hypothalamus on one side and the corpus striatum on the other (Fig. 5, b) the roof of the diencephalon and the adjacent superior wall of the hemispheric vesicles fail to proliferate. Instead, they undergo a U-shaped infolding, resulting in a duplication of the overlying pia-arachnoid membrane which thus forms into the velum interpositum (Fig. 4, b and c). Further invagination and infolding of the fibrovascular stroma of the vela interpositi along the medial wall of the lateral ventricles leads to the formation of the choroidal fissure and of the choroid plexus respectively (Fig. 5).

A similar process of invagination of the fibrovascular stroma from the vela interpositi into the diencephalic vesicle leads to the formation of the choroid plexus in the roof of the third ventricle. Inside the ventricles the fibrovascular stroma of the choroid plexus remains covered by a layer of modified epithelium. At the base of the choroid plexus this modified neuroepithelium merges imperceptibly with the surrounding ependyma. The hippocampal fissure and formation form superior to the choroidal fissure; later, the corpus callosum develops between the two hemispheres at the level of the hippocampal fissure which thus becomes callosal fissure. Between the hippocampal formation of the rapidly growing temporal lobe and the hypothalamus the fornix develops (Fig. 5, b).
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**Fig. 4.** Drawings illustrating early development of neural tube. Note the U-shaped infolding of the diencephalic roof resulting in the formation of the velum interpositum (b and c).

**Fig. 5.** Drawings illustrating formation of the choroidal fissure and plexus (a) and of the corpus callosum, septum pellucidum and fornix (arrows) (b). The lateral walls of the diencephalon and the opposing wall of the hemispheres thicken to form the thalamus and corpus striatum respectively (b). (Modified from Truex and Carpenter, 1969, reference 18.)

The portion of the medial wall of the cerebral vesicles between the corpus callosum and the fornix fails to proliferate and persists as the septum pellucidum (Fig. 5, b). In its posterior part, the inferior edge of the septum pellucidum is attached to the rostral portion of the fornix. Even though the two fornices and the two leaves of the septum pellucidum usually adhere, the microsurgical dissection of this area in a number of autopsy specimens has shown that they may remain separated. With this in mind one can understand how a lesion with an origin in the diencephalic roof can present both inside the third ventricle as well as above it between the two fornices and the two leaves of the septum pellucidum.

Kappers and later Shuangshoti, et al., suggested that the so-called colloid cysts of the third ventricle originate from the neuroepithelium and the overlying stroma in the diencephalic roof by a process of invagination into the third ventricle and subsequent separation of the invaginated portion from the diencephalic roof (Fig. 6, d, e, and f, and cover illustration). It has been emphasized that a number of these cysts will actually remain attached to the choroid plexus in the roof of the third ventricle. A similar sequence of events can also explain the presence of a neuroepithelial (colloid) cyst

**Fig. 6.** Drawings illustrating development of neuroepithelial cysts from the diencephalic roof by the process of evagination into the space between the fornices (a, b, and c), or by the process of invagination into the third ventricle (d, e, and f). See also cover illustration.
Neuroepithelial cysts of the septum pellucidum above the diencephalic roof. This occurs when the neuroepithelium in the diencephalic roof evaginates into the space between the two fornices eventually resulting in a cyst formation (Fig. 6, a, b, and c). In our two patients the cysts remained attached to the rostral portion of the diencephalic roof. The histological examination of the lesion in our patient showed the epithelial lining to be on the inside and the fibrovascular stroma on the outside of the cyst wall. This appearance is consistent with the process of evagination of the neuroepithelium in the diencephalic roof into the overlying fibrovascular stroma of the vela interpositi described above.

According to this concept of their origin, the so-called colloid cysts of the diencephalic roof are more appropriately termed "neuroepithelial cysts" as first suggested by Fulton and Bailey. This term has been adopted since then by a number of other authors. The dispute concerning the role played by the paraphysis, the choroid plexus and the ependyma in the formation of neuroepithelial cysts is easily resolved by the developmental process just described, inasmuch as all of these structures derive from the primitive neuroepithelium and the surrounding fibrovascular mesenchyma of the diencephalic roof.

References


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