Empty sella secondary to suprasellar colloid cyst of foregut (respiratory) origin

Case report

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An enlarged sella turcica was discovered in a 40-year-old man who had bitemporal headaches. A pneumoencephalograph revealed a third ventricular cyst, dilated lateral ventricles, and an empty sella. The colloid cyst was lined by foregut epithelium, probably originating in the respiratory tract, and dense connective tissue. This case is the first instance of an empty sella associated with a colloid cyst of the third ventricle. It is proposed that enlargement of the mass in the third ventricle caused increased pulsation pressure of the cerebrospinal fluid, and that in the presence of an incompetent diaphragma sellae the subarachnoid space expanded into the sella. The origin of third ventricle cysts is reconsidered. It is concluded that suprasellar colloid cysts may arise from endoderm, ectoderm, neuroepithelium, or a combination of these epithelia.

KEY WORDS • empty sella • colloid cyst • foregut cyst • respiratory cyst

The empty sella syndrome has been divided into primary and secondary syndromes. The primary syndrome usually is the result of expansion of the subarachnoid space into the sella turcica through a defect in the diaphragma sellae. The defect allows the pulsation of cerebrospinal fluid (CSF) to expand and erode the sella, and to compress the pituitary gland. The secondary syndrome occurs because of CSF filling the void in the sella after surgical or radiation therapy has obliterated part or all of the pituitary gland.

We report the first instance of a patient with an empty sella associated with a suprasellar, third ventricle cyst of foregut origin. It is postulated that the patient also had a defect in the diaphragma sellae, and that enlargement of the cyst increased CSF pulsation pressure and resulted in the subarachnoid space extending into the sella turcica, thereby creating the empty sella syndrome. We also have reconsidered the origin of third ventricle cysts, and conclude that they may arise from endoderm, ectoderm, neuroepithelium, or from a combination of these.

Case Report

This 40-year-old man presented with a 4-year history of mild intermittent headaches which became bitemporal and more severe 4 weeks before hospitalization. The headaches were occasionally related to changes in position. There was no history of seizures, loss of consciousness, fever, or endocrine symptoms.

Examination. The general and neurological examinations were normal, including visual fields. Admission laboratory data were unrevealing. The following endocrine studies were within normal limits: thyroxine, free thyroxine index, morning cortisol, prolactin, and basal fasting growth hormone. Skull films showed an enlarged sella turcica and eroded posterior clinoids. Computerized tomography (CT) of the head revealed dilated lateral ventricles and a large contrast-enhanced spherical structure above the sella turcica in the region of the third ventricle (Fig. 1). Pneumoencephalography (PEG) confirmed both the dilatation and the soft-tissue density in the third ventricle; air was easily introduced into the large sella turcica. A small amount of pituitary tissue was present posteriorly (Fig. 2). Bilateral carotid arteriograms were within normal limits.

Operation. A right transfrontal craniotomy revealed a cyst, filled with gelatinous material, in the third ventricle. The cyst did not shell out easily, but was entirely removed. Histologically, the cyst was
Colloid cyst and empty sella

**FIG. 1.** Enhanced computerized tomogram showing a spherical mass in the third ventricle, and symmetrically dilated lateral ventricles.

**FIG. 2.** Pneumoencephalogram reveals that the sella turcica is enlarged and the posterior clinoid processes are eroded. Air fills the sella except for a rim of pituitary tissue posteriorly and inferiorly.

**FIG. 3.** Photomicrograph showing foregut epithelium surrounding a core of dense connective tissue. *Inset* demonstrates multiple acini containing colloidal material. H & E, × 70.

composed of predominantly pseudostratified epithelium, occasional acini and colloidal material, and dense connective tissue (Fig. 3). An attempt to study the epithelial cells by electron microscopy failed because of insufficient material. Although cilia were not found, the respiratory rather than the gastrointestinal tract was considered the most likely source of the ectopic epithelium.

**Discussion**

An enlarged sella turcica usually is considered evidence of an expanding intrasellar lesion. A PEG, however, may reveal air in the sella rather than the presence of a pituitary tumor, that is, an empty sella. As early as 1898, Hrdlicka noted that the sella turcica was not a complete mold of the pituitary gland. Busch analyzed necropsy material from 788 patients with no known endocrine disorders, and categorized the contents of the sella into three types. Type IIIc became the "empty sella," and was seen in 40 of the 788 cases (5.5%).

The usual anatomic feature needed to develop an empty sella is an incompetent diaphragma sellae. Other conditions associated with the primary empty sella...
sella include: 1) rupture of cysts of the intrasellar arachnoid or of Rathke's pouch;11,19,20,23,30,31 2) spontaneous infarction of a pituitary tumor, especially eosinophilic adenomas;22,24 3) pituitary hypertrophy during pregnancy and subsequent post-partum involution;1,28 and 4) benign intracranial hypertension.12

Kaufman, et al.,10,26 have detailed the x-ray findings of the empty sella, all of which occur below the level of the diaphragm. The sella turcica occasionally may be indistinguishable from a normal sella; it may be uniformly remodeled in a globular, ballooned shape with normal curvature of the diaphragm; or a thin diaphragm may be associated with a concave sellar floor, at times with a double contour. The degree of remodeling is a function of the incompetence of the diaphragma sellae. The compressed pituitary gland is usually located inferoposteriorly. The empty sella syndrome cannot be differentiated from an intrasellar tumor by examining plain skull films or by finding progressive sellar enlargement,14 hence further evaluation is necessary. Pneumoencephalography has been necessary for diagnosis, because air in the sella demonstrates the "emptiness."19 Increasing accuracy and specificity of the CT scan may obviate the need for a PEG, although Topliss, et al.,29 have reiterated the difficulty in using a CT scan to differentiate an empty sella from an arachnoid cyst within the sella.

The typical patient with an empty sella is a middle-aged woman, obese, and hypertensive, who presents with nonspecific symptoms: headaches, sinusitis, or syncope.5,52 In most instances, the discovery of an enlarged sella is fortuitous. The patients rarely have visual field defects.9 Most have normal endocrine studies, as did our patient.9,19,20,30 Although the lack of endocrine abnormalities usually correlates well with an empty sella, it does not exclude either an intrasellar tumor or the coexistence of pituitary tumor with an empty sella.9,30,31 Colloid cysts in the third ventricle have not been reported to be associated with an empty sella, but the latter has occurred in a case of a cyst within the sella.7

As early as 1924, Camp5 recognized that the sella may enlarge because of intracranial lesions outside the sella. In a more recent report, du Boulay and El Gamal16 found abnormalities of the sella turcica by skull series in 92 of 388 patients with intracranial tumors; none of these cases, however, was associated with an empty sella.

Another type of third ventricle cyst combined with an empty sella was described by Forsbach, et al.13 A 31-year-old woman at 22 weeks' gestation presented with convulsions, blurred vision, persistent severe headaches, and vomiting. She had bitemporal hemianopsia and papilledema. Skull films revealed an enlarged and ballooned sella turcica. Clinical endocrine deficiencies were absent. Four weeks later, a right frontal craniotomy disclosed an extracerebral, irregularly lobulated cyst in the third ventricle and secondary hydrocephalus. Histological examination revealed cysticercosis. The sella turcica was filled with CSF except for a flattened bit of putuitary gland.

Analysis of the patient described by Forsbach, et al.,13 and the case presented in our report, suggests that a previously undescribed entity is associated with an empty sella: cysts and probably other masses in the third ventricle. Our patient lacked overt signs of raised intracranial pressure (ICP) except for severe headaches. The PEG and CT scan revealed grossly dilated lateral ventricles that suggested raised ICP. In most instances of the empty sella syndrome, the patients do not have clear-cut signs of elevated ICP. We postulate that a mild degree of increased CSF pulsation pressure together with an incompetent diaphragm slowly allows the subarachnoid space into the sella turcica and creates an empty sella.

A review of the relevant embryological data is helpful in considering the origin of colloid cysts of the third ventricle. The stomodeum or primitive oval cavity is separated from the pharynx by the buccopharyngeal membrane. This membrane ruptures during the fourth week of gestation, and then completely disappears, obliterating the line of junction between endoderm and ectoderm. Both endodermal and ectodermal epithelia thus have to be considered in evaluating the origin of tissue arising from this region. The respiratory system begins as an endodermal protrusion from the ventral wall of the foregut. Subsequently, the esophageal septum separates the respiratory from the gastrointestinal tract except at the entrance to the larynx. The finding of cilia is useful in distinguishing respiratory from enteric epithelium. The ectoderm of Rathke's pouch initially is only slightly rostral to the stomodeum, and cysts arising from this epithelium cannot be distinguished from neuroepithelium.16

Histologically, colloid cysts of the third ventricle have been considered to be neuroepithelial27 or respiratory14 in origin. Enterogenous cysts thus far have occurred only in the spinal cord. The evidence in our patient is that the epithelial cells arose in the respiratory tract. Multiple sources of origin of colloid cysts were suggested by Challa and Markesbery.6 They described a colloid cyst outside the third ventricle and away from the midline, and found intermingled epithelium of the respiratory tract and of ependyma.

We conclude that a colloid cyst may arise from endoderm, ectoderm, neuroepithelium, or from a combination of them. It may be difficult to decide its origin with present information.2 The term "colloid cyst" has some disadvantages, but until more certain data regarding cells of origin are established, it remains the best term.

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References

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