Intrasellar epidermoid cyst

Case report

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The authors report the uncomplicated removal of an intrasellar epidermoid cyst that on presentation mimicked a pituitary adenoma. Current controversies regarding the differentiation of this cyst from other cystic lesions of the sellar region are reviewed.

KEY WORDS • epidermoid cyst • intrasellar cyst • craniopharyngioma • cyst • transsphenoidal surgery

Clinically significant intrasellar cysts unrelated to pituitary adenomas are rare. We report the case of a typical epidermoid cyst, located within the sella, which on presentation mimicked a pituitary adenoma. Because there is still debate regarding the origins of epidermoid cysts in the sellar region and about their ontogenetic relationship to craniopharyngiomas and intrasellar epithelial cysts, we review briefly the relevant theories.

Case Report

This 41-year-old man was admitted to the Neurosurgery Service at the University of California, San Francisco, with the diagnosis of an endocrine-inactive pituitary adenoma. Six years before admission, he began to experience the gradual onset of fatigue, intolerance to cold, and decreased libido. Two years before admission, an endocrinological evaluation revealed that he had panhypopituitarism. X-ray films of the skull and sellar tomography performed at that time were reported as showing no abnormalities. Hormone replacement therapy was started, with oral thyroid supplement and adrenal corticosteroid and monthly testosterone injections.

Examination. At a follow-up evaluation in February, 1981, the patient was asymptomatic and his physical examination showed no abnormalities. Hormonal studies confirmed that replacement therapy had been adequate, but the levels of the unreplaced pituitary hormones in serum remained below normal. Repeat hypocycloidal polytomograms of the sella revealed gross uniform enlargement and thinning of the sella turcica typical of an intrasellar tumor. High-resolution computerized tomography (CT) disclosed an enlarged sella turcica with an intrasellar region of decreased attenuation and slight suprasellar extension (Fig. 1).

Operation. On March 20, 1981, transsphenoidal exploration of the sella exposed a cystic lesion, 1.5 cm in diameter. Incision of the yellow-gray paper-thin anterior cyst wall released a white caseous amorphous material. There was no liquid component. The cyst wall was excised subtotally, and a fragment was left attached to the attenuated anterior pituitary lobe. Superiorly, the cyst wall was loosely adherent to a small area of thickened arachnoid and diaphragma sellae. When the cyst was removed, no leakage of cerebrospinal fluid (CSF) was observed. The cavity was filled with a free fat graft, and the sellar floor was reconstructed with a piece of nasal septal cartilage.

Postoperative Course. The patient's postoperative recovery was uneventful. One year after surgery, the patient is doing well. Laboratory evaluations show that his pituitary function is improved, but he still receives exogenous hormone replacement therapy. Follow-up CT scans showed no signs of tumor recurrence.

Pathological Examination. Histological examination of the surgical specimen revealed a cyst wall of
mature stratified squamous epithelium with keratinization. There was a well defined layer of prickle cells, but no clearly defined granular layer (Fig. 2). The cyst contained the typical laminated horny material common to epidermoid cysts (Fig. 3 left). The adjacent pituitary gland showed evidence of compression and atrophy (Fig. 3 right). Serial sections failed to disclose an area of tumor histologically different from the typical epidermoid cysts found elsewhere in the central nervous system (CNS).

Discussion

Intracranial epidermoid cysts are rare, representing 0.2% to 1.0% of primary neoplasms of the CNS.\textsuperscript{9,25,26,33} Examples of such cysts in the suprasellar and parasellar regions, although rarer than in other intracranial areas, have been amply reported.\textsuperscript{11,21,25,26} MacCarty, \textit{et al.},\textsuperscript{19} reported four intrasellar epidermoid tumors with presentations and radiological appearance that suggested that significant suprasellar or parasellar extension was also present. In our patient the tumor was entirely intrasellar.

Intracranial epidermoid cysts are generally thought to result from the inclusion of ectodermal elements at the time of closure of the neural groove, or later, during the formation of the cerebral vesicles.\textsuperscript{16,31,32} These cysts may result from iatrogenic or traumatic implantation of epidermis into the subarachnoid space.\textsuperscript{2,31} Epidermoid cysts in the region of the pituitary stalk are often attributed to the proliferation of foci of squamous cell rests in the anterior hypophysis.\textsuperscript{7,8} Some of these rests display cornification and keratohyalin typical of epidermoid tumors.\textsuperscript{5,12} It has also been proposed that metaplasia of anterior hypophyseal cells (as opposed to true embryonic cell rests) may be the source of these foci.\textsuperscript{10,11,25,26}

A clear understanding of the nature of these tumors is complicated by the difficulty in differentiating unequivocally between epidermoid cysts in the sellar
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FIG. 3. Left: Photomicrograph of the cyst contents showing plates of keratin. H & E, × 250. Right: Photomicrograph showing an atrophic pituitary gland (arrows) adjacent to the layer of squamous epithelium. H & E, × 100.

region and craniopharyngiomas. Some authors believe that this distinction is unwarranted; however, others contend that it is justified.

In a review of over 400 cases of craniopharyngiomas, Petito, et al., concluded that no purpose is served by making such a differentiation. They stated, "Most squamous-lined cysts of the suprasellar or parasellar region are probably variants of the craniopharyngioma rather than an 'epidermoid cyst' distinct from craniopharyngiomas because transitions between squamous and adamaninomatous areas can be seen and keratohyaline granules can be found, albeit rarely [in craniopharyngiomas]. Epidermoid cysts elsewhere ... usually show more regular keratohyaline granules, thus producing the pattern of normally keratinizing skin. This is a point that tends to differentiate between ordinary epidermoid cysts and the squamous-lined cysts that occur in the suprasellar region as part of craniopharyngioma." Lever and Schaumberg-Lever pointed out that older epidermoid cysts may lack the stratum granulosum, and thus may not exhibit prominent keratohyalin granules. However, the presence of classic horny material arranged in laminated layers, as observed in our patient's cyst, supports the designation of epidermoid cyst.

Russell and Rubinstein challenged the concept, most recently restated by Yoshida, et al., and Matsushima, et al., that craniopharyngiomas originate from remnants of Rathke's pouch. In agreement with Petito, et al., Russell and Rubinstein believed that there are no valid criteria to support the traditional distinction between suprasellar epidermoid cysts and craniopharyngiomas. Finding no significant differences between these tumors with respect to their clinical presentation, behavior, or histology, the latter authors seemed to posit a common origin of these tumors from squamous cell rests derived from the metaplasia of anterior hypophyseal cells.

In this view, craniopharyngiomas must be considered unrelated to adamantinomas of the jaw, which they can resemble histologically. The increasing number of reports of "odontogenic" craniopharyngiomas containing well developed teeth, however, tends to support the concept that these tumors do indeed originate from derivatives of the stomodeum, or the stomodeum plus pharyngeal endoderm or ectoderm. Landolt suggested that the "enamel organ of developing teeth contains the same cell type as the craniopharyngioma." Relative to histological differences between epidermoid cysts and craniopharyngiomas, adamaninomatous areas have not been reported either in otherwise typical intracranial epidermoid tumors or in teratomas — tumors frequently associated with bone and tooth formation. Similarly, the regions of solid tumor, the rare presence of astrocytes, and the mixtures of epithelium commonly observed in craniopharyngiomas have not been reported in cases of epidermoid tumors.

Additional confusion may arise when considering Rathke's cleft cysts and similar cystic lesions of the sellar region that contain ciliated cuboidal epithelium. Shuangshoti, et al., believed that these latter cysts are, or at least are often, indistinguishable from cysts of neuroepithelial origin. Yoshida, et al., and Matsushima, et al., in a more traditional vein, conclude that Rathke's cleft cysts and craniopharyngiomas have a common origin from Rathke's pouch remnants.

Russell and Rubinstein, in their discussion of several dumbbell-shaped sellar cysts, considered the
role of metaplasia in the formation of squamous and other types of epithelium observed in these lesions. In these rare tumors that they describe, the suprasellar portion was lined with squamous epithelium, whereas the intrasellar portion was lined with ciliated or simple cuboidal epithelium. They suggested that the suprasellar portion of these "dumbbell cysts" may represent an area of epithelial metaplasia, in that similar foci of squamous metaplasia have been observed in the persistent nonpathological Rathke's cleft. An argument along similar lines might be proposed to explain the transitional regions in craniopharyngiomas where adamantinomatous areas merge into areas of squamous epithelium, which could also be the result of metaplasia. Independent of the mode of origin, the demonstration of cuboidal cells and goblet cells in the cysts described above and in craniopharyngiomas contrasts with their absence from intracranial epidermoids.

Our patient presented with hypopituitarism, which is unusual in cases of epidermoid cysts, but characteristic of lesions that are primarily intrasellar. Business management, such as therapies to manage tumor growth, was unnecessary, and the tumor was not adherent to the pituitary gland. In a craniopharyngioma, we have not observed the caseous white semisolid laminated material (typical of epidermoids) found in the tumor we report here. Cyst contents, however, do vary considerably in both tumor types. Landolt suggested that the horny material representative of the final step of keratinization is not found in craniopharyngiomas.

Kernohan and Sayre pointed out differences in the characteristics of growth between craniopharyngiomas and epidermoid cysts that may have clinical relevance. The slow progression of the tumor in our case is consistent with the behavior of epidermoid cysts outlined by Alvor and others. Based on his evaluation of growth rates, biological characteristics, and clinical behavior, Alvord concluded that epidermoids are actually congenital malformations, whereas craniopharyngiomas are true neoplasms. Liszczak, et al., studying the biological characteristics of craniopharyngiomas in cell culture, showed that these tumors had two distinct tissue populations, a finding that seems to correlate with their clinical behavior. One population, although exhibiting epithelioid features, also had features characteristic of neoepithelial transformation. Clinically, one is often impressed with a greater tendency for recurrence in craniopharyngiomas, particularly following subtotal resection, which leads some authors to recommend postoperative irradiation. Such a course of postoperative care contrasts sharply with that recommended for other cystic sellar lesions. Another notable clinical difference between epidermoid cysts and craniopharyngiomas is that intracranial epidermoids tend to present during the third to fifth decades of life, in contrast to the predominance of craniopharyngiomas in the first to second decade. Unfortunately, no detailed comparison of craniopharyngiomas, epidermoid cysts, and other sellar cysts based on clinical or tissue-culture studies has been reported.

The origins of and complex relationship between the cystic sellar region tumors cannot be resolved here. In light of the complex embryological development of the sellar region and the varied manifestations of epithelial lesions occurring in this region, it seems an oversimplification to claim one pathogenesis for all of them. The tumor reported here exhibited features so characteristic of epidermoid cysts observed elsewhere in the CNS that a brief review of this lesion's relationship to craniopharyngiomas and other intrasellar cysts seemed warranted. We suggest that a continued distinction between these lesions is appropriate, and that some intrasellar "craniopharyngiomas" may in fact prove to be distinct epidermoid cysts.

References

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