A 70-Year-Old Man With Diplopia, Nausea, and Vomiting

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A 70-year-old man presented with a 4-day history of progressive diplopia. Twenty-four hours prior to admission, he became nauseated and began to vomit. The neurological examination revealed no additional abnormalities, but a computed tomographic scan of the head demonstrated an expansion of the sella turcica with an area of increased intensity on the postcontrast study. There was no evidence of calcification within the mass or the sellar space. Axial gadolinium-enhanced T1-weighted magnetic resonance imaging (MRI) of the head revealed a large intrasellar mass with suprasellar extension (Figure 1). The mass measured approximately 3 cm in greatest dimension, superiorly displacing the optic chiasm. The lesion contained areas of decreased signal intensity consistent with foci of necrosis or cystic degeneration (Figure 1, arrowheads). The hormonal studies revealed slightly increased serum prolactin and reduced serum levels of thyroid-stimulating hormone and somatomedin C.

Two weeks later, the patient was admitted for transsphenoidal intracranial mass resection. During surgery, tumor erosion through the posterior wall of the sphenoid was noticed. The resected mass was cystic, pale, and tan-brown, grossly measuring 3 cm in greatest diameter (Figure 2). The unicameral cyst had a connective tissue wall and was lined by ciliated columnar epithelium (Figure 3). The intrasellar part of the cyst wall contained a 0.7-cm nodular area composed of uniform pituitary cells arranged in solid nests and sheets without lobular architecture (Figure 4). Immunohistochemical stains for prolactin, follicle-stimulating hormone, leuteinizing hormone, thyroid-stimulating hormone, human growth hormone, and adrenocorticotropic hormone were all negative. Postoperatively, the patient was found to be alert and oriented with voluntary movement of all extremities noted.

What is your diagnosis?
Pathologic Diagnosis: Rathke Cleft Cyst Concomitant With Pituitary Adenoma

Abstract

A 70-year-old man who presented with diplopia was found to have an intrasellar unilocular cyst lined by ciliated columnar epithelium. The cyst showed suprasellar extension and measured 3 cm in greatest dimension. The intrasellar part of the cyst wall contained a pituitary adenoma. These findings are characteristic of Rathke cleft cyst with concomitant pituitary adenoma, an entity that must be distinguished from other intrasellar and suprasellar lesions.

Rathke cleft cyst (RCC), first described by Luschka in 1860, is thought to arise from incomplete obliteration of the lumen of Rathke pouch.1 This embryonic structure develops as a rostral outpouching of the stomodeum or primitive oral cavity during the third or fourth week of gestation.2 It has an anterior and posterior wall, as well as a central embryonic cleft. The anterior wall of the pouch proliferates to form the anterior lobe of the pituitary gland and the pars tuberalis; the posterior wall becomes the pars intermedia. The residual lumen of the pouch is reduced to the narrow Rathke cleft, which usually regresses completely. It is the persistence and enlargement of this cleft that is said to be the cause of symptomatic RCC.3 Other theories on the origin of RCC, such as neuroepithelial tissue, remnants of embryonic endoderm, or metaplastic anterior pituitary cells that have changed into ciliated cells,4 seem to be less plausible.

The cells of the anterior pituitary lobe, from which pituitary adenomas develop, are also derived from the cells of Rathke pouch. Although RCC and pituitary adenoma have a shared ancestry, they rarely occur coincidentally; we only found 9 cases reported in the literature.5 Small, clinically asymptomatic RCCs are found incidentally in 11% to 33% of random postmortem examinations.2 Larger cysts, like the one described in this article, are less common and are usually associated with symptoms related to compression of adjacent structures and increased intracranial pressure. Such manifestations are indistinguishable from those caused by other sellar or suprasellar masses, such as pituitary adenoma or craniopharyngioma.4 Headache is encountered in 44% to 81% of patients, visual loss in 11% to 67% of patients, and endocrinological symptoms in 30% to 60% of patients.5,6

When RCC and pituitary adenoma are found together, symptoms of mass effect may be accompanied by hormonal changes caused by the functioning tumor.7 Our patient had no endocrine-related symptoms, even though he had a slightly elevated serum prolactin concentration. The hyperprolactinemia was most likely related to compres-

References