A 44-year-old man with a 3-year history of bilateral vision deterioration presented for medical attention because of severe retroocular headaches. Physical examination revealed amaurosis of the right eye and temporal field blindness in the left eye; funduscopia showed atrophy of the optic disc borders bilaterally. No other signs or symptoms were present. A computed tomography scan demonstrated a hypodense intrasellar and suprasellar lesion. A T1-weighted magnetic resonance image revealed a large suprasellar and intrasellar cystic lesion that dorsally displaced and compressed the hypothalamus (Figure 1, arrow). No abnormalities of pituitary hormone levels were apparent. The patient underwent a subfrontal craniotomy, the serous cyst content was drained, and the wall of the cyst was partially excised. The patient died of respiratory failure that developed after the operation.

The autopsy revealed multiple foci of pneumonia as the cause of death. Unfortunately, no cultures were performed, so the etiologic agent was not identified. The ventral surface of the brain showed a 5-cm suprasellar cystic cavity that compressed the optic chiasm (Figure 2) and a softening due to a subacute infarction in the territory of both the middle and right anterior cerebral arteries, a change associated with the surgical procedure and more evident in the coronal sections (Figure 3). The hypophysis was not identified grossly. Histologic examination of the cystic lesion revealed a nonstratified cuboidal to columnar ciliated epithelium with round to oval basal nuclei (Figure 4) and some islands of normal adenohypophysis and neurohypophysis. The brain tissue near the cyst showed prominent gliosis.

What is your diagnosis?
Pathologic Diagnosis: Rathke Cleft Cyst

Rathke cleft cysts, such as craniopharyngiomas, are generally believed to originate from remnants of the Rathke pouch, although some authors believe there may be a different origin or that Rathke cysts may originate from the Rathke pouch at a different stage of development. Rathke cysts are incidental findings in approximately 33% of routine autopsies. Rarely, these cysts become large enough to cause symptoms. Since Goldzieher first described the entity in 1913, at least 179 cases have been reported. They usually present in the fourth or fifth decade of life, and there is a 2:1 female predominance. The most common symptoms are visual field defects, hypopituitarism, and headaches. On computed tomography scans and magnetic resonance images, the cysts are usually intrasellar and/or suprasellar, without calcifications, and range from hypo-, iso-, or mixed density or intensity, respectively. This last feature is due to the variable amount of mucus in the cyst. T1-weighted images are hyperintense when there is a high content of protein, cholesterol, or mucopolysaccharide in the cyst. In imaging studies, the main differential diagnosis is cystic craniopharyngioma, cystic or hemorrhagic pituitary adenoma, arachnoid cyst, pars intermedia cyst, ependymal cyst, epidermoid cyst, cysticercosis, empty sella, mucocele, and intrasellar aneurysm.

The cysts may become symptomatic when they attain a diameter of 1 cm. They have a thin wall, and the cyst contents are variable with a serous, mucoid, gelatinous, or motor oil appearance. The epithelial lining of the cyst is usually cuboidal or columnar and may be ciliated or contain goblet cells. Squamous metaplasia frequently occurs, which has been attributed to proliferating reserve cells beneath the surface epithelium, although some authors think that it is a secondary change caused by inflammation. When squamous metaplasia is present, it may be difficult to distinguish craniopharyngioma from Rathke cleft cyst, especially in small surgical specimens. Xin et al. found that cytokeratins 8 and 20 are positive in Rathke cleft cysts and negative in craniopharyngioma. This immunohistochemical profile may be useful diagnostically in difficult cases.

The management of this benign lesion is surgical excision. Some recurrent cases have been reported, and in some the recurrence was associated with sellar packing with organic materials such as bone, fat, or cartilage.

References