Rathke’s cleft cyst associated with hypophysitis: MRI

Abstract We report a symptomatic Rathke’s cleft cyst associated with hypophysitis in a 61-year-old woman. We demonstrate the MRI features and discuss the pathophysiology. To the best of our knowledge this is the first description of a Rathke’s cleft cyst shrinking after high-dose steroid therapy.

Key words Cyst, Rathke’s cleft · Hypophysitis · Magnetic resonance imaging

Introduction

Rathke’s cleft cysts are non-neoplastic lesions thought to be derived from remnants of Rathke’s pouch. They are found between the anterior and posterior lobes of the pituitary in 11–33% of routine autopsies [1, 2]. They may also develop in the suprasellar area, along the pituitary stalk. They are usually asymptomatic, but some are large enough to compress the pituitary gland, optic chiasm, optic nerves or hypothalamus [3–7]. The symptoms and signs are generally caused by compression of these structures [3–7]. We report an extremely rare case of a Rathke’s cleft cyst with hypophysitis, which we think caused hypophysitis.

Case report

A 61-year-old woman was admitted with a history of polyuria and polydipsia for a year and gradually decreasing visual acuity for 2 months. Examination revealed decreased visual acuity and bitemporal hemianopia. Routine laboratory examinations were normal. Endocrine investigations revealed hypopituitarism, with low levels of growth hormone, thyroid-stimulating hormone, cortisol and antidiuretic hormone. Plasma prolactin was slightly increased. The cerebrospinal fluid (CSF) showed high protein, 132 mg/dl (normal range 10–40 mg/dl), and slightly elevated glucose, 100 mg/dl (50–80 mg/dl). The CSF contained 46 nucleated cells mm³, 94% of which were mononuclear.

MRI revealed intra- and suprasellar cystic masses which ballooned the sella turcica and compressed the optic chiasm. The intrasellar mass gave high signal on T1-weighted images and slightly lower signal than CSF on T2-weighted images, while the suprasellar one gave slightly high signal on T1-weighted images and was isointense with CSF on T2-weighted images (Fig.1a, b). The optic nerves were swollen bilaterally and their central portion gave high signal on T2-weighted images. The hypothalamus gave mildly increased signal on T2-weighted images, suggesting oedema (Fig.1b–d). The bright posterior lobe of the pituitary was not seen on T1-weighted images (Fig.1e). There was thick rim contrast enhancement of the masses (Fig.1f). No calcification was seen on CT. To prevent further visual impairment, high-dose steroid therapy was started. CT 5 days later demonstrated slight shrinkage of the masses and MRI after 13 days revealed further reduction in size of the intra- and suprasellar cystic masses (Fig.2).
**Fig. 1** a Coronal T1-weighted image shows intra- and suprasellar cystic masses that compress the optic chiasm (arrow). b On a coronal T2-weighted image the intrasellar portion gives high signal but slightly lower than CSF, while the suprasellar mass gives markedly high signal. c The optic nerves are swollen bilaterally and their central portions (arrow) give high signal. d Mildly increased signal is seen in the hypothalamus. e Sagittal T1-weighted image: no bright posterior lobe is observed. f After contrast medium, thick rim enhancement of the suprasellar cystic mass is seen. Intense contrast enhancement is observed anterior to the intrasellar cyst in an area proven histologically to show adenohypophysitis.

**Fig. 2** Sagittal contrast-enhanced T1-weighted image 13 days after high-dose steroid therapy: the intra- and suprasellar masses have reduced in size.

**Fig. 3a, b** Histology of the specimen (haematoxylin and eosin: a original magnification, ×20; b original magnification, ×200). In the magnified view the cyst wall is seen to be lined by ciliated columnar epithelium, with focal squamous metaplasia and occasional goblet cells (arrow). The cyst wall (small arrow) shows a focal dehiscence, which appears to have been repaired by fibrin tissue (arrowhead). The pituitary tissue abutting the cyst is heavily infiltrated by inflammatory cells (asterisk), mainly lymphocytes and plasma cells.
The intrasellar mass was excised via a transphenoidal approach 16 days after the steroid treatment. A slightly yellow cyst was observed behind the anterior lobe, and white creamy contents poured out when its wall was incised. The cyst wall and a small specimen of the adenohypophysis were sent for histological examination. There was no attempt to remove the suprasellar mass.

The cyst wall was lined with ciliated columnar epithelium, focal areas of squamous metaplasia and occasional goblet cells. The epithelium exhibited a focal dehiscence. The pituitary tissue abutting the cyst was heavily infiltrated by inflammatory cells, mainly lymphocytes and plasma cells. No organisms were identified, and no granuloma was found. The histological diagnosis was Rathke’s cleft cyst and adenohypophysisis (Fig.3).

Another course of high-dose steroid therapy was given after surgery, and the suprasellar cystic eventually disappeared during follow-up. The patient is doing well on replacement therapy with oral cortisone, levothyroxine sodium, and nasal vasopressin 2 years after surgery.

Adenohypophysitis in our patient was probably induced by the cyst fluid from the ruptured Rathke’s cleft cyst, since a focal defect in the wall was observed microscopically and inflammatory cells strongly infiltrated the pituitary tissue in the vicinity of the cyst.

Although no histologic confirmation was obtained from the suprasellar mass, the presentation with diabetes insipidus and the disappearance of the suprasellar mass after steroid therapy strongly suggest that the mass was not only related to adenohypophysitis, but also to “neurohypophysitis”. Sumida et al. [11] reported a case of “hypophysitis” associated with a Rathke’s cleft cyst. The pituitary surrounding the cyst enhanced markedly with contrast medium, but no image was presented. It is difficult to assess whether there was “neurohypophysitis”. It seems that ours is the first report of the MRI features of adenohypophysitis and “neurohypophysitis” associated with a Rathke’s cleft cyst. It was difficult to detect the adenohypophysitis on MRI. The marked shaggy enhancement anterior to the intrasellar cyst presumably indicated adenohypophysitis (Fig.11).

An interesting aspect is the decrease in size of the cyst after high-dose steroid therapy. To our knowledge, this is the first report of this phenomenon. Although the pathophysiology is not clear, we assume that the steroids had an effect on the secretion or absorption of cyst fluid. Our finding suggests that steroid therapy may be useful in some cases of Rathke’s cleft cyst with inflammatory change.

**Discussion**

Rathke’s cleft cyst associated with hypophysitis is extremely rare, only four cases having been reported [8–11], two Rathke’s cleft cysts associated with chronic hypophysitis. One of these appeared to have been a reactive response to the cyst contents. The other two were Rathke’s cleft cysts associated with granulomatous hypophysitis, the latter thought to have been induced by rupture of cyst and leakage of the contents.

**References**