Letters to the Editor


Periodic fever: an unusual manifestation of a recurrent Rathke's cleft

Rathke's cleft cysts are generally asymptomatic intrasellar cysts. Occasionally they grow and compress surrounding structures and this can lead to various neurological, neuro-ophthalmological and endocrinological symptoms. The report deals with a recurrent Rathke's cleft cyst which was associated with periodic fever; permanent drainage gave remarkable relief of symptoms.

A 30 year old woman was referred in December 1977 with a 17 month history of visual impairment, occasional headaches, and irregular menses. She was slow of thought, otherwise physical examination was normal. Perimetry showed bitemporal hemianopia. Routine blood and urine analysis gave normal results. Endocrine evaluation revealed hypocortisolaemia and hypogonadotropism of pituitary origin. Moderate hyperprolactinaemia was also found. A contrast-enhanced CT scan of the brain showed a well-circumscribed intra- and suprasellar homogeneous low density lesion (15 x 15 x 18 mm) with capsular enhancement. No evidence of calcification was found.

In May 1984 a craniotomy was performed. This showed a cystic tumour with compression of the optic chiasm. Needle aspiration yielded 4 ml sterile fluid containing leukocytes. The histological features were consistent with a Rathke's cleft cyst accompanied by signs of chronic inflammatory reaction. Postoperatively she remained well and her visual fields improved, but panhypopituitarism made replacement therapy necessary.

From April 1986 she suffered from attacks of fever, which recurred at two week intervals and lasted six hours. These attacks occurred during her sleep and were associated with feelings of cold and discomfort, pilo-erection, and frequent chills. The rectal temperature reached to 39°C. This was followed by a three hour period of excessive sweating during which the body temperature gradually returned to normal. In December 1986 she was admitted to hospital because of severe bifrontal headache and vomiting. Routine laboratory investigation revealed no abnormalities. A CT scan of the brain showed a homogenous, low-density lesion (20 x 20 x 20 mm), situated in the midline above the sella turcica. Both lateral ventricles were dilated. A bifrontal ventriculostomy was performed with a Holter-Hauser normal pressure valve was installed. The cerebrospinal fluid (CSF) pressure was considered elevated (50 mm H2O); the cisterns were normal.

This operation produced only slight improvement of the headaches and had no effect on the fever attacks, which now recurred about once a week at random times during the day. Magnetic resonance imaging (MRI) of the brain was performed on a Gyrocscan S5 operating with a field strength of 0.5 Tesla. Coronal and sagittal T1 and T2 weighted spin echo pulse sequences were used (TR/TE 400-600; 30 and 2000/50-100, respectively). This clearly demonstrated suprasellar extension of the cyst, which reached to the level of the foramen of Monro and caused compression of hypothalamic structures. The cystic lesion was non-homogeneous and inintense to slightly hyperintense on the T1-weighted image compared with the signal intensity of CSF and slightly hyperintense on the T2-weighted image. Adequate drainage of the ventricles was established (fig). Under CT guidance, drainage of the cyst was accomplished by stereotactic introduction of a drain tip into the cyst. Aspiration yielded 4 ml of fluid.

Microscopically, this fluid contained necrotic material. Cultures were again negative. A subgaleal reservoir was mounted and connected to the ventriculostrial system. MRI of the brain performed one month later showed that the cyst had almost disappeared. Almost two years after this operation the patient is still doing well. The headaches ceased immediately and the fever attacks became much less frequent and less severe within one month after drainage of the cyst. About once or twice a month she wakes up for a few minutes during the night feeling cold and shivering slightly without any change in body temperature.

In the few reports of Rathke's cleft cysts that have produced symptoms, the most common features are headache, hypopituitarism, visual symptoms, and hydrocephalus. Fever has rarely been reported in association with Rathke's cleft cyst. In our patient, the periodic attacks of fever developed more than one year after the first operation, and eight months before the obstructive hydrocephalus became symptomatic. Because large oscillations of intracranial pressure occur during the paradoxical stage of sleep, the focal occurrence of fever attacks might represent related phenomena of elevated intracranial pressure. However, after ventriculostomy the shunting the fever attacks persisted and only permanent drainage of the cyst resulted in rapid clinical improvement, suggesting that local compression of the hypothalamus by the cyst is more essential than elevated intracranial pressure in general. The initial two week cycle of fever attacks and suprasellar extension of the cyst further suggest a hypothalamic disturbance.

Two other possible causes of fever in Rathke's cleft cyst should be considered. In the first place, the signs of chronic inflammatory reaction in the cyst wall suggest the presence of cytokines, which may act on the hypothalamus to induce fever. This is, however, contradicted by the frequently documented microscopic signs of inflammation in the cyst wall without the development of fever. Second, the contents of the cyst may leak into the subarachnoid space and cause aseptic meningitis. This seems unlikely because our patient did not show signs of meningitis and the CSF was normal.

In conclusion, suprasellar extension of a Rathke's cleft cyst may cause signs and symptoms of hydrocephalus related to hypothalamic dysfunction. This case illustrates the occurrence of periodic fever as a hypothalamic feature of suprasellar extension of a Rathke's cleft cyst.

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Figure: Coronal T1 and T2 weighted images demonstrate the suprasellar component. Note the flow void phenomenon in the ventriculostrial system bifrontally.

References: