A Cyst of Rathke's Cleft

B. FAIRBURN, F.R.C.S., AND I. M. LARKIN, M.D.
Oldchurch Hospital, Romford, Essex, England

Cases of compression of the optic chiasm form an important section of a neurosurgeon's practice. In the majority of cases, such compression is caused by adenomas of the pituitary gland or by suprasellar cysts (tumours of Rathke's pouch). A very rare cause of chiasmal compression is that resulting from the presence of a cyst within the sella turcica arising from Rathke's cleft.

This type of cyst has a distinctive histological appearance. It is lined by cuboidal or columnar epithelium which is often ciliated in places and may include mucous goblet cells. The clinical picture produced by such a cyst may be indistinguishable from that produced by a chromophobe adenoma. Only 3 cases of successful surgical treatment of this condition have been recorded.4,7,9 A fourth case is here reported.

Case Report

I.C.R., aged 43, a married woman with three children, was first admitted to Oldchurch Hospital on Sept. 24, 1958. She gave a history of attacks of violent headache since May, 1955. These attacks would come on suddenly and for no apparent reason. The headache was felt in both temporal regions and radiated into the occiput. It was described as continuous, violent and throbbing and it was aggravated by stooping. The headache would last for several weeks and she would vomit repeatedly during the attack. At times her vision was blurred during the attack and on one occasion she lost the vision in her right eye completely for a few days. Between attacks she felt quite well. The worst of these attacks had occurred shortly before her admission to hospital and was associated with a marked mental disturbance.

Her previous medical history was clear apart from thrombophlebitis of the right leg at the time of her third pregnancy. There was nothing relevant in the family history. Her periods had ceased in May, 1957.

Examination. She was alert and orientated. Her fundi were normal and her visual fields were full. There were no abnormal neurological signs. Investigations showed: Hemoglobin 87 per cent (12.85 gm. per cent), white blood cells 9,000 per c.mm. (normal differential), erythrocyte sedimentation rate 24 mm. in 1 hr., Wassermann reaction and Kahn negative. Cerebrospinal fluid pressure 160 mm., protein 20 mg. per cent, and no excess of white blood cells. Roentgenograms of skull showed pituitary fossa on the large side of normal. A ventriculogram showed no abnormality of the ventricular system. She was discharged home on Sept. 29, 1958.

Course. She remained well until January, 1959, when her headaches recurred. They gradually became more frequent and severe and at the beginning of April, 1959, her vision began to deteriorate, especially in the left eye.

2nd Admission, April 26, 1959. She was alert and rational. There was some pallor of the left optic disc. Visual acuity was 6/9 in the right eye. Vision in the left eye was reduced to counting fingers. There was an almost complete temporal hemianopia of the right eye with some constriction of the nasal field. A similar but more severe defect was present in the left eye. Neurological findings were otherwise negative. Her blood pressure was 90/68. Her skin was dry and the axillary and pubic hair were sparse.

The following investigations were carried out: Hemoglobin 79 per cent (11.05 gm. per cent). Slight anisocytosis and polychromasia of red blood cells. White blood cells 3,800 per c.mm. (polymorphonuclear cells 53 per cent, lymphocytes 29 per cent, monocytes 7 per cent, eosinophils 11 per cent). Serum chlorides (as NaCl) 151 mg. per cent, serum sodium 270 mg. per cent, serum potassium 18 mg. per cent, and plasma cholesterol 196 mg. per cent. Urinary neutral ketosteroids 1.3 mg., 17 ketogenic ketosteroids nil. Sugar tolerance curve was normal. Electroencephalogram was normal. Cerebrospinal fluid: normal pressure and constituents. Roentgenograms of skull revealed slight enlargement of pituitary fossa with thinning of dorsum sellae. Lumbar air encephalogram showed a rounded swelling arising out of the pituitary fossa and projecting into the basal cisterns.

Operation. On May 12, 1959, a right frontal osteoplastic flap was turned down and the pituitary region was approached by elevating the right frontal lobe. The anterior part of the optic chiasm and both optic nerves were seen to be stretched over a rounded tumour which projected upwards as a pink dome-shaped structure between the flattened optic nerves. The tumour felt moderately tense. After preliminary aspiration had shown that the swelling was not an aneurysm, the capsule was incised. Some 2 ml. of thick, purulent-looking material escaped. When this was cleared away a smooth-walled cystic cavity about 12 mm. in diameter was revealed. It appeared to occupy practically the whole of the pituitary fossa. A piece of the capsule was removed for section. With the evacuation of the cyst, the chiasm and optic nerves were seen to be lying in their normal anatomical position and free from tension. The wound was closed without drainage.

Pathologic Report. The cystic fluid showed a few epithelial cells but no organisms. Culture was sterile. Histological sections of the cyst showed that is had a simple fibrous wall and was lined by a single layer of tall columnar ciliated cells with occasional mucus-secreting goblet cells (Fig. 1).

Subsequent Progress. Postoperative recovery was uneventful apart from a transient mild diabetes insipidus. Her bitemporal hemianopia showed evidence of recovery within a few days and her fields eventually became full. Visual acuity improved to 6/6 in each eye (J1 on the Jaeger types). Her headaches have been completely relieved. She has required regular cortisone 12.5 mg. t.d.s. and thyroxin 0.5 mg. since operation. She now

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Discussion

Mucoid epithelial cysts of the pituitary body, although uncommon, are a well established pathological entity, and quite distinct from the squamous tumours and cysts which occur in the same region. They may, however, have a similar origin.

The anterior lobe of the pituitary body derives in the embryo from ectoderm of the roof of the stomodaeum. A diverticulum appears at this site (the pouch of Rathke). It extends up in front of the cephalic end of the notochord and comes into contact with the undersurface of the forebrain. In man it is constricted off to form a closed vesicle but remains for a time connected to the ectoderm of the stomodaeum by a solid cord of cells. In some of the lower chordates (cyclostomes) where the elements of the pituitary body first make their appearance, the anterior lobe of the pituitary body is represented by the hypophyseal duct, which remains as a simple tube in the myxinoids, lined by a single layer of ectodermal cells.

Mott and Barratt\(^4\) developed the idea that epithelial tumours in the region of the hypophysis are derived from remnants of the craniopharyngeal duct.

Duffy\(^5\) described a pituitary cyst lined by ciliated columnar epithelium which was found in a negro patient in association with a large pituitary adenoma. He distinguished this clearly from the commoner squamous type of cyst and considered that it derived from Rathke’s cleft, i.e. the cleft that persists into human postnatal life between the pars anterior and the pars posterior of the pituitary. It is then distended with gelatinous material and is obvious to the naked eye.\(^6\) Sometimes it persists in this state into adult life but causes no functional disturbance. The cleft is lined with cuboidal or columnar epithelium which is often ciliated in places and may include mucous goblet cells.

Rasmussen\(^6\) examined several hundred human pituitary glands in a search for ciliated columnar and mucus-secreting cells and he thus summarised his findings. “It appears that tall columnar cells with numerous delicate cilia occasionally occur in the epithelium lining the residual lumen and cysts in the region of the pars intermedia of human hypophyses that otherwise are essentially normal. Some of these cells apparently secrete a mucus-like material or undergo mucoid degeneration. Their origin is uncertain but probably they should be looked upon as unusual differentiations of hypophyseal tissues or as migrations of nasopharyngeal elements during early stages of development.”

Bailey,\(^1\) writing on tumours of the hypophyseal duct, stated that they may be divided into three types: i) mucoid epithelial cysts; ii) simple squamous epitheliomas, and iii) adamantinomas. Of

* The use of this term in this context is not today generally acceptable.