CYSTIC CRANIOPHARYNGIOMA: SURGICAL TREATMENT BY ENDONASAL APPROACH

Report of a Case

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CRANIOPHARYNGIOMAS are benign pituitary tumors that arise from Rathke's pouch. These tumors usually are cystic, but may be solid. Many other names have been given to these tumors, including *Rathke's pouch cysts or tumors, hypophyseal duct cysts or tumors, suprasellar cysts or tumors, adamantinomas,* and *epitheliomas.* Craniopharyngiomas need not be difficult to diagnose, but they always are difficult to treat.

This report presents the case history of a patient whose cystic craniopharyngioma was treated by endonasal sphenoidostomy and aspiration of the tumor mass.

Report of Case

A 35-year-old Negro man was first seen at the Cleveland Clinic in December 1947. His chief complaint was loss of libido, which he had first noted five years earlier. His history revealed that, approximately three years before examination, while serving in the Armed Forces in October 1944, he had noted loss of vision in the lateral area of the left eye. A brain tumor had been diagnosed, and a right frontal craniotomy had been performed in 1945 in an Army hospital. After surgery the sight in the left eye had improved, but vision in the right eye had decreased. He had been unable to read or to distinguish the features of people, but he had been able to get around by himself. When he was initially seen at the Clinic, the diagnosis of impotence probably secondary to suprasellar tumor was made. Testosterone propionate, 25 mg. three times weekly for three weeks, and 50 mg. three times weekly for three additional weeks, was administered, without improvement in his condition.

Two and one-half years later (June 1950) he returned to the Clinic to be examined in the Department of Ophthalmology because of total blindness of sudden onset ten days previously, allegedly following an upper respiratory infection. He said that he occasionally had had an ache in the right eye but no headaches. On examination, there were dilation of the pupils, absence of light reflex, bilateral paralysis of the sixth nerve, pale optic discs bilaterally, and total blindness. Recurrent suprasellar tumor was suspected and he was referred to the Department of Neurological Surgery. Except for the previously mentioned ocular findings, the neurological examination was negative. A spinal tap was not contributory.

Roentgenograms of the skull (Fig. 1) showed destruction of the sella turcica with pronounced depression of the floor. The dorsum sellae turcicae and the posterior clinoids were not visualized. A small calcification was present in the midline, posterior to the region of the sella turcica. No other intracranial calcifications were demonstrated. The

sphenoid and petrous ridges appeared to be normal, but there was a 1 by 2 cm. defect in the left sphenoid wing, probably representing bone erosion. A bone flap was noted in the right frontal temporal region.

In June 1950 a left frontal craniotomy was performed, revealing a large cystic tumor beneath and anterior to the optic chiasm. Aspiration yielded approximately 60 cc. of dark oily fluid. A circular section of the cystic wall removed for histopathologic examination was reported to be fibrous tissue.

Four years later, in July 1954, the patient was seen in the Department of Otolaryngology with symptoms of hoarseness, dysphagia, and a tendency to choke on liquids, of one month's duration. The findings on examination were: a mass filling the nasopharynx, paralysis of the right hypoglossal nerve, paralysis of the right side of the soft palate, paralysis of the right vocal cord, and diminished gag reflex.

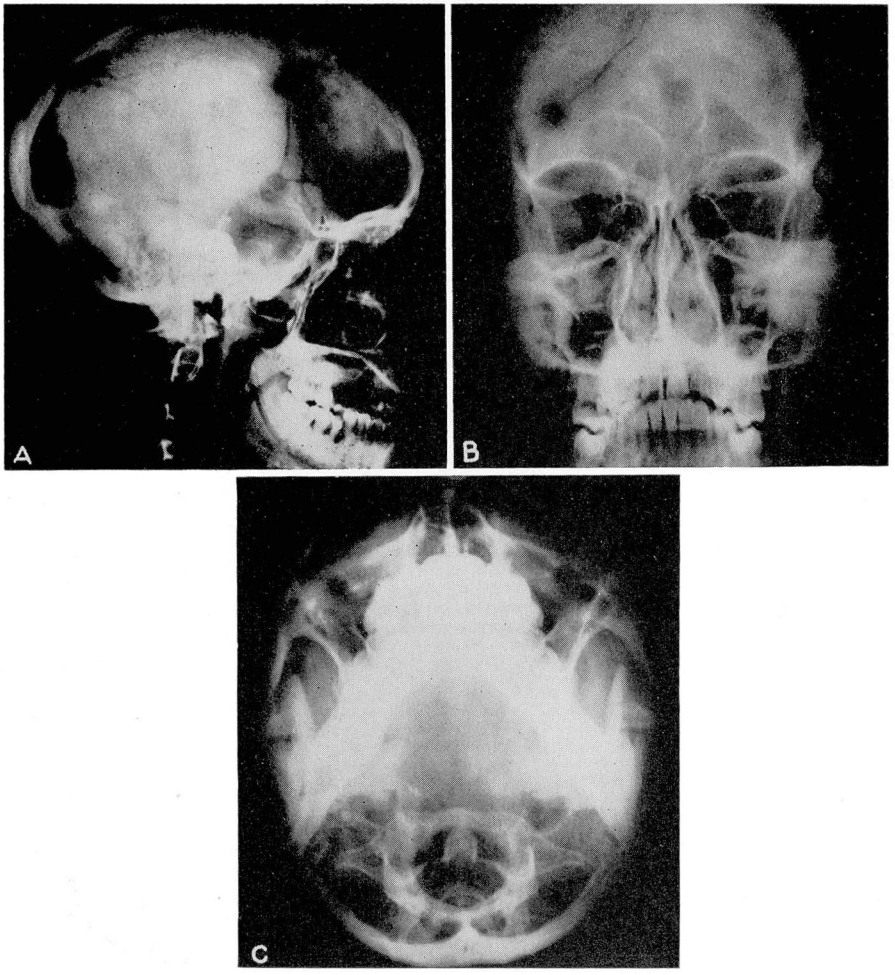

Fig. 1. Preoperative roentgenograms in 1950.

Roentgenograms of the skull at this time (Fig. 2) showed destruction of the floor of the middle fossa and calcification in a large area 5 cm. in diameter in the central region of the base of the skull. Part of this calcification was intracranial above the petrous tips, with the remainder in the nasopharynx extending down almost to the soft palate. The size of the nasopharyngeal tumor appeared to be greater than that indicated by previous roentgenograms in June 1950. The tips of the petrous apices were eroded.

In August 1954, an endonasal sphenoidostomy was performed. A transseptal approach was considered unnecessary because the nasal passages were large. The nasopharynx was well visualized by anterior nasoscopy after shrinkage and out-fracturing of

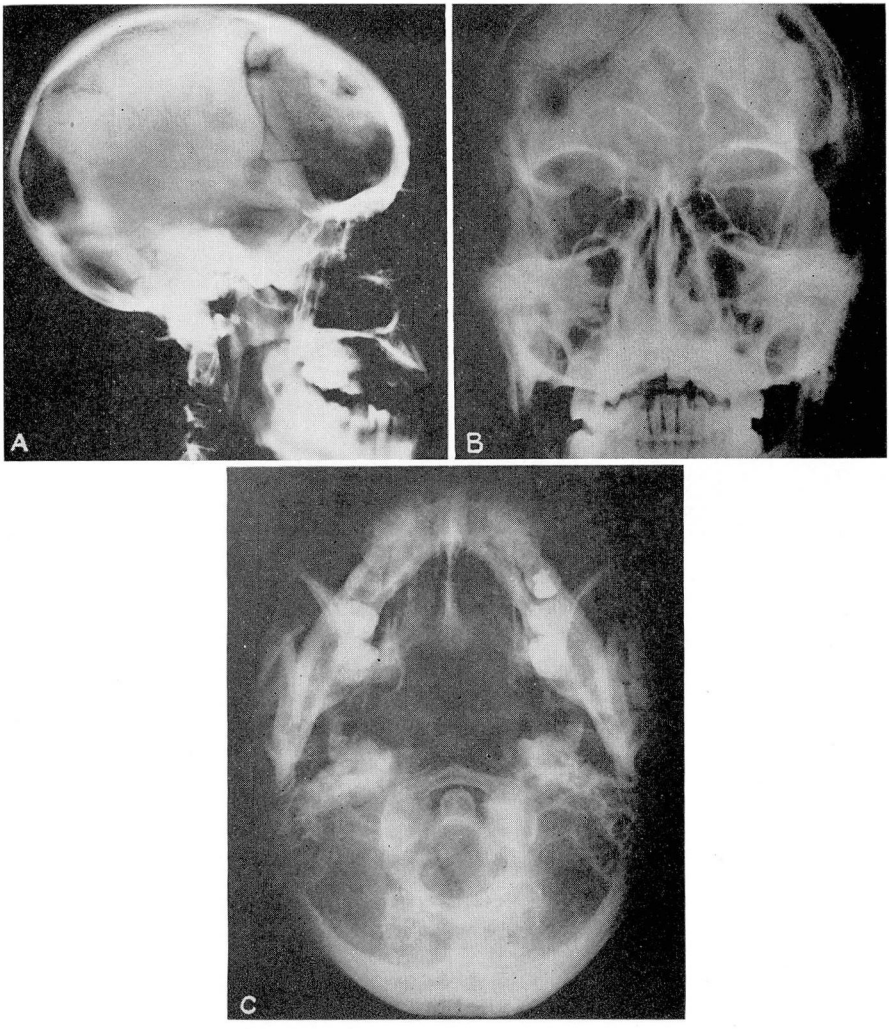

Fig. 2. Preoperative roentgenograms in 1954.

the turbinates against the lateral nasal wall. An incision was made (through the right nostril) into the tumor mass, releasing 70 to 80 cc. of dark-brown oily fluid. With a punch forceps the right nasal opening to the tumor was enlarged to approximately 2 cm. in diameter. A similar opening was then made through the left nostril, and the two openings were converted into a single large opening by removal of a portion of the posterior nasal septum (Fig. 3). (The procedure is similar to that described by Hirsch[1] which, however, entails submucous resection.)

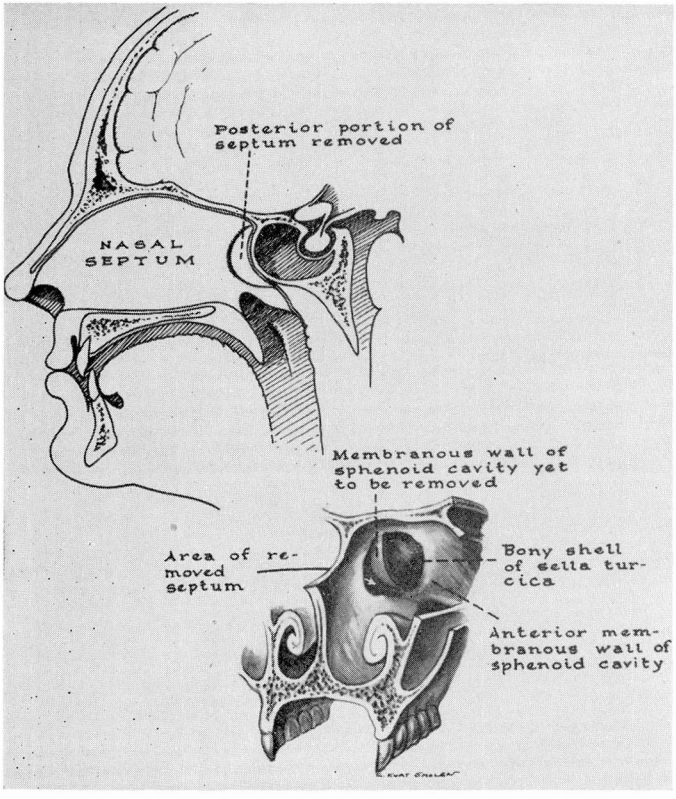

Fig. 3. Diagrams of operative technic.

The cavity was packed with 1-inch vaseline gauze. This pack was gradually removed during the next four days. On the fifth postoperative day roentgenograms obtained after instillation of 30 cc. of iodized oil outlined the cavity (Fig. 4). The immediately postoperative course was not remarkable.

Six months postoperatively, roentgenograms obtained after instillation of iodized oil showed a decrease in the size of the cavity (Fig. 5). In June 1955, nine months after operation, the patient said that his hoarseness had cleared and his tongue had been moving better during the past month. At that time he had normal movements of the tongue, a very active gag reflex (topical anesthesia had to be applied in order to visualize the larynx), normal movement of the soft palate, and good motion of the right vocal cord. The patient was last seen in December 1955, approximately 16 months after

surgery. The sphenoidal opening was patent and the patient stated that, except for his blindness and impotence, he felt better than he had in years.

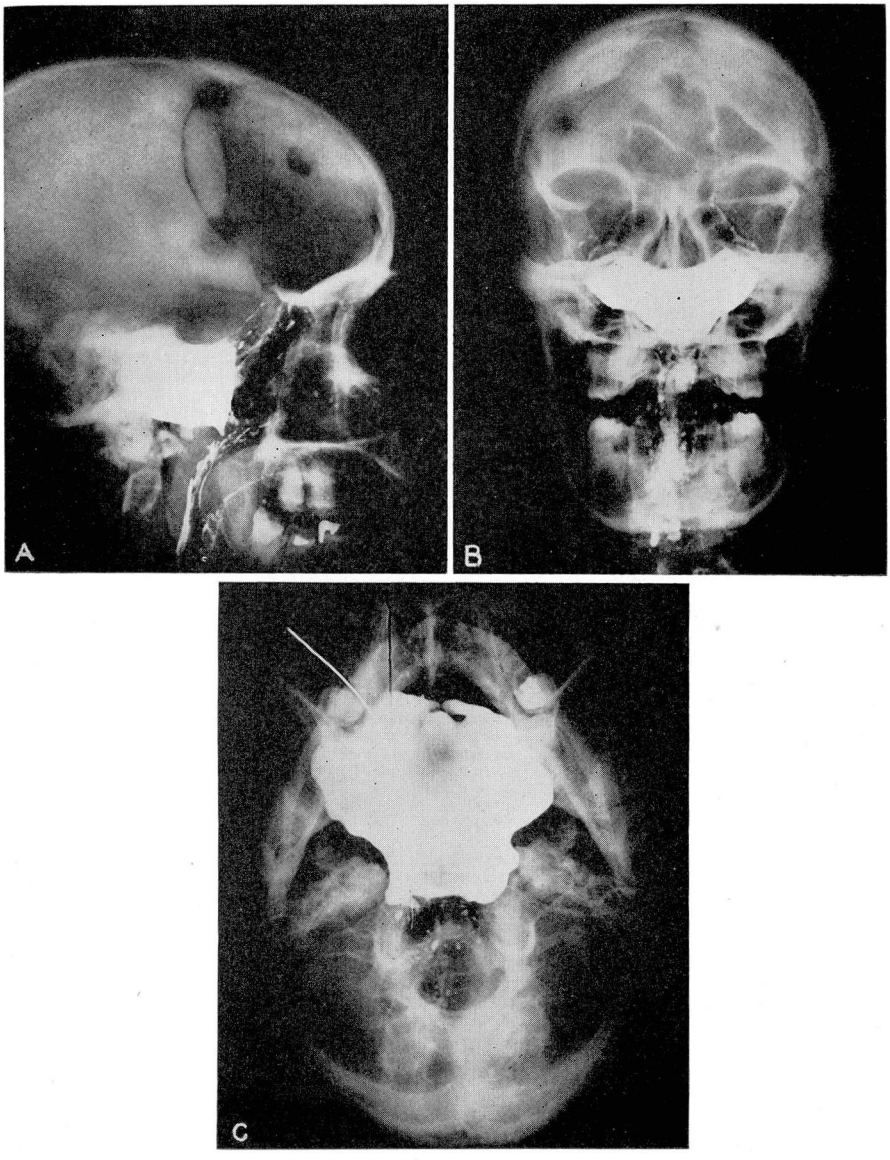

Fig. 4. Roentgenogram of cavity after instillation of iodized oil immediately after operation.

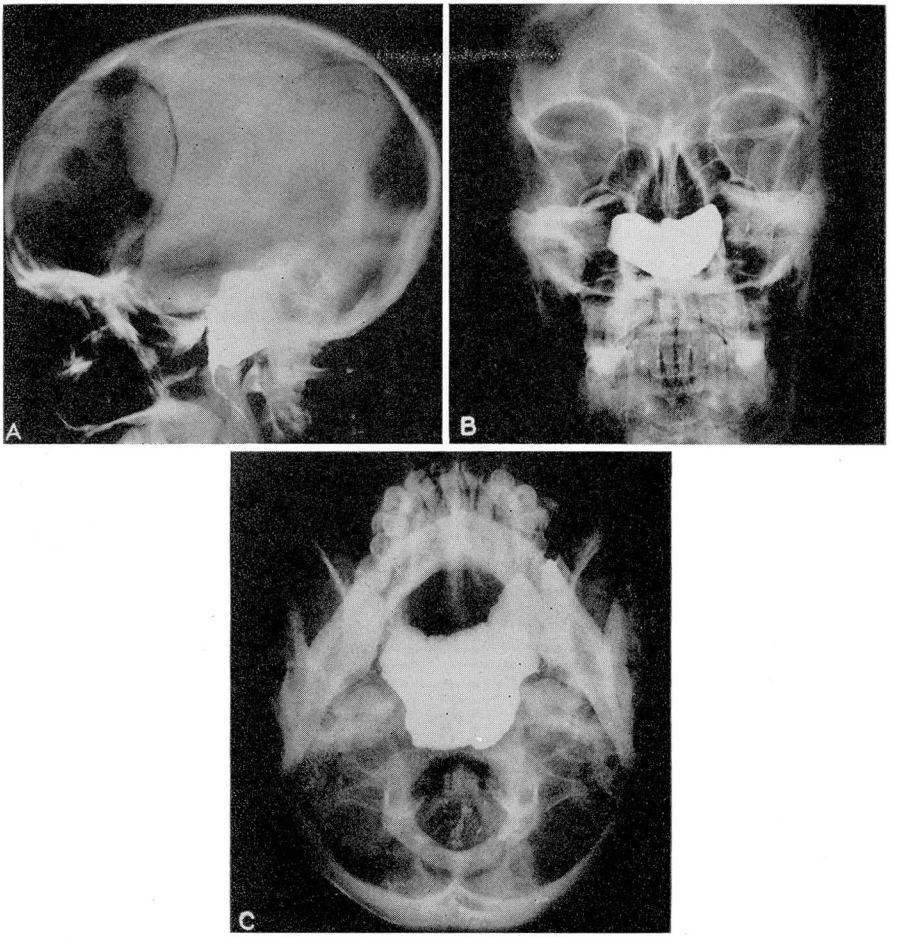

Fig. 5. Roentgenogram after instillation of iodized oil six months postoperatively.

Discussion

Diagnosis. The diagnosis of craniopharyngiomas like that of other pituitary tumors is made on the basis of endocrinologic, ophthalmologic, and roentgenographic findings. In the case presented, hemianopsia was the first symptom that caused the patient to seek medical aid, although he had noted a loss of libido two years earlier. Total blindness and paralysis of the extraocular muscles developed approximately five and one-half years after the onset of the hemianopsia. Roentgenograms of the skull at that time showed destruction of the sella turcica by a tumor mass with a partially calcified periphery, and thus probably indicative of a craniopharyngioma.

The occasional difficulties in diagnosing craniopharyngioma are exemplified by the results reported for one series of 96 patients who were operated upon for suspected craniopharyngioma and in 6 of whom no tumor was found.[2] Endocrine disturbances may not be apparent in children, who comprise a large proportion of persons having craniopharyngiomas, [2,3] or in menopausal or in hysterectomized women. An enlarged sella turcica is not in itself diagnostic since hydrocephalus or glioma in the region of the sella may cause the enlargement. Sphenoid mucocele,[4] a rare condition, also must be considered in the differential diagnosis. Calcifications considered characteristic of craniopharyngiomas also may be found in aneurysms, chondromas, osteomas, and sclerotic carotid arteries. Roentgenograms with contrast air filling and arteriograms may be of diagnostic aid.

Treatment. Craniopharyngiomas may be treated medically, surgically, and by irradiation. Medical management generally is instituted for the control of the endocrine disturbances. Irradiation alone has been considered of no value in treatment,[3] although postoperative irradiation has been reported to alleviate symptoms that had not been relieved by surgery or that had recurred postoperatively.[5] The surgical treatment employs either of two approaches: the transfrontal craniotomy, or the endonasal sphenoidostomy.

Gordy, Peet, and Kahn[3] reported a mortality of 41 per cent in a series of 51 patients having craniopharyngiomas operated upon by the transfrontal approach. They remarked on the pessimism among neurosurgeons concerning the results of surgery for craniopharyngiomas, occasioned by the high operative mortality and the many cases in which total removal is manifestly impossible. Love and Marshall[2] reported a mortality of 40 per cent in 96 patients after transfrontal craniotomy for craniopharyngioma.

Many variations of the transsphenoidal approach have been performed, but the one used most frequently, most successfully, and for the longest period of time is the transseptal-sphenoidal approach (nasal submucous resection route) devised by Hirsch.[1] From 1910 to 1951, Hirsch employed this procedure in the treatment of 383 pituitary tumors of various types, such as the chromophobic and eosinophilic adenomas where there was enlargement of the sella turcica, with about 35 per cent being cystic or semicystic tumors (craniopharyngiomas or cystic chromophobic tumors). Although he did not classify his data according to types of tumors, the over-all mortality for his series was significantly low—5.4 per cent. A very recent report by Hirsch,[6] analyzing a total of 413 patients with pituitary tumors, reveals a 1.5 per cent mortality in the antibiotic era since 1945. On the basis of these figures it would appear that selected pituitary tumors where there is enlargement of the sella turcica could be readily approached through the endonasal route.

Summary

A case report is presented of a patient with a cystic craniopharyngioma that had been treated by two transfrontal craniotomies without preventing continued

enlargement of the tumor and cranial nerve involvement. Aspiration of the neoplasm through endonasal sphenoidostomy resulted in recovery of the involved nerves. It is believed that the endonasal approach has definite application in the treatment of cystic craniopharyngiomas such as that in the case presented.

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