Long Term Survival with Craniopharyngioma

Report of Patient in 29th Year after Treatment, Seen for a Second Intracranial Tumor

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The study by Love and Marshall of 104 patients with craniopharyngioma shows that long-term survival of patients having this tumor is unusual. Twenty-one of their patients were alive 5 years after treatment, but only 5 survived 10 years, and one for 20 years. Radiation therapy was thought to be beneficial in relieving symptoms in 10 cases, but had no significant effect on longevity. The patient who survived 20 years was not included in the group receiving radiation. Kramer et al. showed favorable results in the treatment of 10 patients with craniopharyngioma by a combination of minimal surgery and supervoltage irradiation, but long-term follow-up of a large series of patients receiving this form of therapy is not yet available. Because of the long survival (29 years) we were prompted to report the following case of craniopharyngioma treated by aspiration and irradiation. Of additional interest was the occurrence of a second primary intracranial neoplasm diagnosed during evaluation of the patient for possible extension of the craniopharyngioma.

Case Report

A. K. (UVH #551420). A 47-year-old woman was admitted to the Neurosurgical Service of the University of Virginia Hospital with a chief complaint of failing vision.

History. In 1937, at the age of 18 years, the patient had complaints of amnorrhea, obesity, loss of visual acuity, visual field defects, and symptoms of generalized increased intracranial pressure. Roentgenograms of the skull had demonstrated the typical suprasellar calcification of a craniopharyngioma. Clinical improvement followed aspiration of a cyst. During the next 2 years she had had recurrent cyst formation requiring aspiration twice, and received 3 courses of conventional x-ray therapy to the suprasellar region. The hospital record does not include a notation of the amount of radiation given.

Twenty-eight years later she noticed progressive loss of vision. An ophthalmologist diagnosed early cataract formation; she was admitted to the neurosurgical service to determine whether her visual loss was caused by extension of the craniopharyngioma or cataracts. She had not complained of headache and had no other symptoms of increased intracranial pressure.

Examination. The patient was an alert, garrulous, obese white woman. She had excellent recall for her entire medical history, and her thought processes appeared perfectly normal. However, the following day, although she had the same excellent recall, she had no recollection that she had already given the history to at least 3 examiners. Consultation with her family physician confirmed the fact that a defect in recent memory and other personality changes had been present for approximately 1 year.

There was a divergent strabismus, and ptosis on the right. On ophthalmoscopic examination cataracts were seen. There was slight pallor of the optic discs, and no evidence of papilledema. There was no nystagmus. A right supranuclear facial weakness was present. There was no anosmia. The remaining cranial nerves were intact and no other neurological abnormalities were present.

Laboratory data. Routine hematologic studies, fasting blood glucose, blood urea, protein bound iodine and cholesterol were normal. Urinalysis was also normal. Roentgenograms of the skull showed a suprasellar calcification 3 cm. in diameter, and an unusual mottled appearance of the calvaria. A roentgenogram of the chest disclosed severe osteoporosis of the spine but no pulmonary parenchymal disease. Additional laboratory studies included serum calcium, phosphorous, total protein and a bone marrow examination, all of which were normal. The serum electrophoretic pattern indicated a relative increase in globulin and a decrease in albumin. The cerebrospinal fluid protein was 51 mg%. 

Course. An ophthalmological consultant observed the cataracts and demonstrated incongruous paracentral visual field defects. It could not be determined, however, if the decrease in visual acuity was entirely due to the cataracts. Because of the abnormal radiographic appearance of the calvaria, possible sites of metastatic disease were sought, and studies for multiple myeloma were made. None of these investigations showed abnormalities. A pneumoencephalogram (Fig. 1) showed that there had been no re-growth of the craniopharyngioma; the ventricular system was slightly enlarged, and there was some frontal atrophy. The frontal horn of the right lateral ventricle was distorted. The electroencephalogram gave evidence of a right anterior quadrant intracerebral destructive lesion. A brain scan (Fig. 2) demonstrated increased uptake of radioactive mercury in the medial right frontal lobe. At operation, a right frontal parasagittal meningioma was removed. The postoperative course was prolonged by saphenous thrombophlebitis, but was otherwise uneventful.

Discussion

In the initial stages of this disease, re-aspiration of the cystic tumor was necessary on several occasions but after 3 courses of conventional x-ray therapy, no recurrence was evident 29 years after the patient was first treated. The effectiveness of x-ray therapy in patients with craniopharyn-
Craniopharyngiomas have been suggested by Kramer et al. who had favorable results in 10 patients with craniopharyngiomas treated by aspiration, and in some cases, Torkildsen shunts, followed by supervoltage irradiation. Nine of their 10 patients were alive 6 or more years after treatment and the 10th died of intercurrent disease rather than the tumor. In addition, the apparent effectiveness of radioactive chromic phosphate in the treatment of recurrent cystic formation in craniopharyngioma has been suggested in the case reported by Overton and Sheff. On the other hand, there is evidence that a craniopharyngioma may remain latent for many years. In the series presented by Love and Marshall, 1 patient had a 31-year history of diabetes insipidus which subsided before symptoms of anterior pituitary insufficiency led to the diagnosis of craniopharyngioma.

The other unusual feature of this case is the occurrence of 2 primary intracranial neoplasms. Although the diagnosis of craniopharyngioma was not established by biopsy, the clinical syndrome, the suprasellar calcification, and the type of fluid aspirated from the cyst leave little doubt regarding the nature of the lesion. The meningioma, as shown in Fig. 3, was one of the psammomatous variety. An area of hyperostosis was present on the inner table of the frontal bone flap and was firmly attached to the dura overlying the meningioma. The tumor was also attached to the falx.

The sequence of events leading to the diagnosis of the meningioma is also interesting. The presenting problem, loss of vision, could not adequately be explained by the meningioma because there was neither an increase in intracranial pressure nor an extension into the optic system. The visual loss was most likely caused by the cataracts since the pneumoencephalogram showed no significant growth of the craniopharyngioma. The mottled appearance of the calvaria on plain x-rays of the skull suggested a diagnosis of metastatic disease or multiple myeloma; further studies then re-

**Fig. 1.** Antero-posterior (A) and lateral (B) views of the pneumoencephalogram demonstrate the suprasellar calcification, air in the basilar cisterns and ventricular enlargement with deformation of the frontal horn of the right lateral ventricle. The mottled appearance of the calvaria is also evident.

**Fig. 2.** Antero-posterior (above) and lateral (below) views of the Hg 197 brain scan show an area of greatly increased uptake in the right frontal pole. RLO: right lateral orbit; N: nasion; LLO: left lateral orbit; EAM: external auditory meatus.
revealed the presence of the right frontal lobe mass. A preoperative diagnosis of meningioma was considered because of the size and position of the tumor as well as the history of impairment of memory existing for at least 1 year. It was felt, however, that the most likely diagnosis was a metastatic carcinoma, probably from an occult carcinoma of the breast. Finding a curable lesion at operation was most gratifying.

Summary

We have reported the case of a patient surviving 29 years after treatment of a craniopharyngioma by aspiration and irradiation. The patient also had a second primary intracranial neoplasm, a meningioma.

References

