

Aggressive Surgical Management of Craniopharyngiomas in Children

Hoffman et al, from the Hospital for Sick Children in Toronto, report 14-years experience in total excision of craniopharyngiomas in 50 children (22 girls and 28 boys, from 1 year 10 months to 17 years 7 months in age). Headache with a duration of 2 weeks to 4 years was the most common presenting complaint (68%). Thirty-three patients (66%) had some endocrine abnormality at presentation (14%, hypothyroidism; 40%, short stature; 24%, diabetes insipidus; 18%, obesity; and 14%, delayed secondary sexual development). One patient presented with precocious puberty. Visual abnormalities were present in 30% to 60%, with 48% having a field defect, the most common being bitemporal hemianopia.

Computed tomographic (CT) evidence of tumor calcification was observed in all 50 patients. Eighty percent of the tumors had some form of cyst formation. Sellar enlargement and/or blunting of the dorsum sellae was noted in 40%. Half the tumors were prechiasmatic. Hydrocephalus was present in 24 of the 50 patients at the time of surgery and in 74% of the patients with retrochiasmatic tumors. The most common surgical approach was right frontal craniotomy. Forty-five patients, or 90%, were considered by the surgeon to have undergone total tumor excision at the time of surgery.

Follow-up was obtained on 46 patients. One died in the immediate postoperative period, and 2 others died 9 years after initial surgery. Follow-up was from 1 to 14 years and 39% for at least 5 years. Thirty-four percent (16) have experienced tumor recurrence, a third of which were asymptomatic and discovered on routine neuroimaging. Eight presented with headaches, 5 had deterioration in visual acuity, and 1 had an increased need for desmopressin. Among 13 patients with tumor recurrence, 5 had normal postoperative CT findings, but 8 demonstrated either calcification or residual tumor.

Nine had improvement in visual fields, but 16 of those who had no field defect before surgery had deterioration of visual fields. Endocrine deficiencies were observed in all 46 follow-up patients postoperatively. Over 90% required desmopressin, 89% cortisone, 83% thyroid hormone, 31% sex steroids, and 20% growth hormone. Seventy-four percent

required a combination of thyroid hormone, cortisone, and desmopressin. Fifty-two percent were obese at follow-up, but almost 30% of these had been obese prior to surgery.

Twenty-seven children had a formal psychometric evaluation at follow-up. Twenty-six of 27 had intelligence levels at or above average levels. However, memory was impaired in 16 of the 28 children tested. Twenty-four of 39 assessed for educational status were attending regular school.

Quality of life was assessed by categorizing patients into 3 groups based on morbidity. Those in the first group, or those with the "good quality of life," had no tumor recurrence or, if the tumor recurred, it was adequately managed with surgery. In addition, these patients had good control of their endocrine deficiencies and were attending or had attended regular school, and displayed no behavioral or eating disturbances. Sixty-four percent of patients fell into this category. The severely handicapped group included patients with unstable tumor recurrence and poorly controlled endocrine status. They were not attending school because of behavioral problems or major psychological disturbances. Only 9% fell into this group. The remaining 27% fell into the intermediate group.

Hoffman HJ, De Silva M, Humphreys RP, et al. *J Neurosurg* 1992;76:47-52.

Editor's comment: *This is an interesting and very well compiled follow-up report regarding the outcome of microsurgical management of craniopharyngiomas evaluated by CT. The data have been carefully assessed and should provide useful information for pediatric endocrinologists who are often responsible for diagnosing this tumor and who need to discuss the outcome and prognosis with their patients and their families. It is interesting to note that radiation therapy was used only in patients with tumor recurrences. In 1992 most patients will receive radiation in addition to surgical therapy. Therefore, statistics using data from combined therapy should be improved over those presented here.*

William L. Clarke, MD