Giant craniopharyngioma has presented an unabating challenge in management. Throughout the literature, results are reported and statements are made in regard to increased surgical mortality, morbidity, and poor outcome with reduced success of total removal and higher recurrence rates. Master surgeons declared, “Experience has shown that the larger the tumor the greater will be the damage, both preoperatively and intraoperatively, to vital intracranial structures.”

Hence, this report on the radical microsurgical resection of craniopharyngioma in children is a most welcome contribution; the authors are to be congratulated for their experience and their thorough reporting. This report not only demonstrates that these giant tumors can be radically resected, but it also leads to the acceptance of radical resection as the management of choice for all craniopharyngiomas. Smaller craniopharyngiomas can then be totally removed with much less morbidity, mortality, and hence better outcome.

A heated controversy in craniopharyngioma management has been reignited with repeated recent calls for minimal surgical intervention. One recent article was even titled, “Neurosurgical treatment strategies in childhood craniopharyngiomas: is less more?” The application of endonasal endoscopic technique is likely to carry even less success with respect to total removal, particularly in giant and large tumors.

Almost all the patients who are treated with partial resection or aspiration technique are doomed to undergo adjunct radiation therapy in some form (conformal, radiosurgery, brachytherapy). The application of interferon or bleomycin has yet to gain wide acceptance. The side effects and complications of radiation, regardless of its form, are drastically underestimated, particularly in the young. The long-term side effects (visual, cognitive, hypothalamic) are frequent and serious. Radiation-induced meningioma, too, is an aggressive disease and a high price to pay for an attempt to control a benign tumor that is surgically curable, as with craniopharyngiomas of all sizes.

As the authors demonstrated, our experience corroborates that giant and even monstrous craniopharyngiomas can be safely removed with fine microneurosurgical technique with good outcome. We believe that skull base approaches are particularly helpful in achieving this goal because they provide a wide exposure, minimize brain retraction, provide multiple corridors for dissection, and shorten the distance, reducing the surgical morbidity related to size.

The most critical aspect of dealing with giant craniopharyngioma is the retrochiasmatic location, and since all the commonly used approaches have an anterior trajectory that poses risk to the optic pathways and the large vessels and perforators, we have found it greatly advantageous to overcome these obstacles by approaching the tumor through the petrosal approach.

References

Response

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We thank Dr. Al-Mefty for his comments on our experience with these very difficult and rare tumors—giant craniopharyngiomas. In addition to his excellent results in his series on giant tumors,2 he has reported success using the petrosal approach.1 We have tended to favor the more lateral (pterional) approach but the results from his center speak for themselves.

We also would like to commend Drs. Fahlbusch and Hoffman for their series of 16 cases involving patients with giant craniopharyngiomas (> 4 cm in maximal diameter) treated primarily by the bifrontal interhemispheric approach.4 In 4 patients, they utilized stereotactic cyst aspiration prior to surgical removal. As we suggested, this may help reduce the incidence of craniocerebral disproportion following the removal of such a large intracranial mass. They achieved complete resection in 10 or 16 patients (83%) and had no perioperative mortality. A single patient (6%) experienced neurological worsening and 2 patients had visual deterioration. As expected with aggressive surgical treatment of such large tumors, endocrinological deficits were common but not universal.

These results, and those of Dr. Al-Mefty, provide further support that giant craniopharyngiomas can be successfully and safely treated with microsurgery. We firmly believe microsurgery to be the best definitive treatment option for craniopharyngiomas of all sizes, if cure is the goal. We agree with Dr. Al-Mefty that irradiation should not be the primary therapy in children given the potential for late adverse effects in a population with such long life expectancy. However, curative surgical therapy will carry greater immediate morbidity and require long-term societal support for optimal quality of life. It is essential for the neurosurgeon to have a frank discussion with the parents of a child with a craniopharyngioma: the family must be well informed and the physician must assess the ability of the family to cope with the potential early and late sequelae of therapy.

While the popularity of endoscopic transnasal techniques is rising, such approaches are of limited utility in tumors with peripheral calcification, cases with significant solid disease in the suprasellar region, and tumors that extend lateral to the carotid arteries (which includes, most, if not all, giant tumors).3,5 Similarly, intracavitary therapies may be inadequate to provide lasting disease control for such tumors, given the presence of multiple compartments, loculated cysts, and varying amounts of solid tumor tissue.

Regardless of the surgical approach or treatment strategy, giant craniopharyngiomas remain some of the most difficult and potentially dangerous brain tumors to treat. For optimal outcomes and quality of life, we urge that consideration be given to referral of children with giant craniopharyngiomas to centers with experience in treating these difficult tumors. (DOI: 10.3171/2009.12.PEDS09508)

References