The role of surgery in optic pathway/hypothalamic gliomas in children.

Goodden J, Pizer B, Pettorini B, Williams D, Blair J, Didi M, Thorp N, Mallucci C.
Departments of Pediatric Neurosurgery.

Abstract
Object Optic pathway/hypothalamic gliomas (OPHGs) are generally benign tumors situated in an exquisitely sensitive brain region. The location and natural history of OPHGs has led to much debate about optimal treatment. This paper revisits the role of and optimal timing of debulking surgery in OPHG.

Methods This paper presents a series of cases managed by the neuro-oncology team at Alder Hey Children's Hospital and a single surgeon. Data were collected retrospectively for periods prior to 2009 and prospectively thereafter. Tailored treatment strategies were used, including observation and combinations of surgery, chemotherapy, and radiotherapy. Tumor control rates and outcomes are reviewed.

Results Forty-two patients were treated between 1998 and 2011. Their median age at diagnosis was 5 years 7 months. Nineteen patients were positive for neurofibromatosis Type 1 (NF1) and 23 patients were negative for NF1. The median duration of follow-up was 77 months (range 21.8-142.3 months). Presenting symptoms included visual impairment (in 50% of cases), headache (in 24%), and hypothalamic/pituitary dysfunction (in 29%). Twenty-two debulking procedures were performed in 21 patients. Four biopsies (3 open, 1 endoscopic) were also performed. The histological diagnosis was pilocytic astrocytoma in 21 patients and pilomyxoid astrocytoma in 2 patients. Ten patients (Group 1) had primary surgical debulking alone and were then observed. Four patients (Group 2) had surgical debulking, plus planned chemotherapy within 3 months. Seven patients (Group 3) required surgical debulking for progressive disease following a variety of treatments. Patient age had the greatest impact on subsequent tumor progression. In total, 13 patients received chemotherapy, 4 on initial presentation, 4 in combination with surgery, and 5 for further tumor progression. Five patients were treated with radiotherapy, 3 prior to referral to Alder Hey. Eleven patients required shunt insertion for hydrocephalus. Vision was stabilized for 74% of patients. The number of patients with hypothalamic/pituitary dysfunction increased from 12 at presentation to 16 by the end of treatment. The overall survival rate was 93%. Three patients died-1 from tumor progression, 1 from infective complications from tumor biopsy, and 1 from a spontaneous posterior fossa hemorrhage. NF1 was associated with improved outcome-fewer patients required active intervention and rates of visual impairment and/or hypothalamic/pituitary dysfunction were lower. Conclusions Good long-term survival and functional outcomes can be achieved in children with OPHG. Tumor control was achieved through an individualized approach using surgery, chemotherapy, or radiotherapy in varied combinations. The authors aim to limit radiotherapy to cases involving older children in whom other therapies have failed, due to the well-described and often devastating late effects associated with midline cranial irradiation. Surgery has a clear role for diagnosis, tumor control, and relief of mass effect. In particular, primary surgical debulking of tumor (without adjuvant therapy) is safe and effective. Recent advances in intraoperative MRI may add value and need further assessment.

PMID: 24138145 [PubMed - as supplied by publisher]
The role of surgery in optic pathway/hyp... [J Neurosurg Pediatr. 2013] ...