

5. Treatment should be deferred until clear evidence of progression. Chemotherapy is first-line therapy. Radiation is not recommended. Surgery is considered with excessive proptosis and blindness. (Listernick R, Ferrier RE, Liu GT, Gutmann DH. Optic pathway gliomas in neurofibromatosis-1: Controversies and recommendations. **Ann Neurol** March 2007;61:189-198). (Respond: David H Gutmann MD PhD, Department of Neurology, Washington University School of Medicine, 660 South Euclid Ave, Box 8111, St Louis, MO 63110).

COMMENT. In a prospective, longitudinal study at Children's Memorial Hospital, Chicago, OPGs were found in 19% of 176 children with NF-1 who had CT or MRI at a median age of 4.1 years (Listernick R et al. **J Pediatr** 1994;125:63-66). OPG had not developed at follow-up in those not receiving an MRI. Eye lesions such as proptosis or glaucoma were most common in younger children (median age 1.9 years). OPG was asymptomatic at time of diagnosis in 76%, and eye findings were normal in 64%. At follow-up of 0.2-8 years, only 3 (9%) showed progressive tumor growth on MRI or deteriorating vision after diagnosis. Patients with symptomatic OPG were all diagnosed before age 6 years. Tumor growth after 6 years is unusual. Progressive abnormalities and precocious puberty occurred only with chiasmatic OPG. In accord with the present recommendations, serial eye exams in young children with NF-1 but not MRI were advocated in this earlier report.

BRAIN MALFORMATIONS

CHIARI TYPE I MALFORMATION AFTER RADIATION THERAPY

The development of a Chiari I malformation and cervical syringomyelia, 1.5 years after radiation for a malignant, rhabdoid tumor of the neck at 3 years of age, was diagnosed by MRI after the patient had 2 episodes of unresponsiveness. The radiation therapy followed chemotherapy and surgical exploration of the mass that involved the cranial base. The tumor could not be resected because it surrounded the internal carotid artery. The Chiari malformation was treated by suboccipital decompressive craniectomy and C1 laminectomy with duraplasty, and no further syncopal episodes occurred during a follow-up of 5.5 years. Postoperative MRIs indicated reconstitution of the CSF at the foramen magnum, ascent of the cerebellar tonsils, and resolution of the presyrinx state of the cervical cord. One previous case report in the literature describes a Chiari I malformation in a child after fractionated radiation therapy to the anterior cranial base. (Hoffman CE, Lis E, Wolden SL et al. Symptomatic Chiari I malformation after radiation therapy in an infant: Case report. **Neurosurgery** April 2007;60:E782).

COMMENT. Known causes of acquired Chiari I malformation cited by the authors include lumboperitoneal shunts, craniosynostosis, rickets, supratentorial mass, spinal drainage, and acromegaly. A case of a 13-year-old girl whose Chiari I malformation presented after head trauma is also worthy of note (Mampalam TJ et al. **Neurosurgery** 1988;23:760-762). Radiation exposure is an additional acquired cause. Children who undergo radiation for medulloblastoma or brainstem tumor might also be observed for Chiari I malformation. Syncope described in the above patient is mentioned as cough syncope syndrome in a previous publication on Chiari malformations (Ireland PD et al. **Arch Neurol** 1996;53:526-531).