TUMORS OF THE OPTIC NERVE

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ABSTRACT

This is a review of primary and secondary tumors of the optic nerve. The emphasis
is on optic nerve gliomas and meningiomas.

Optic nerve gliomas are slowly growing astrocytic neoplasms of the anterior visual
pathways, the majority of which occur within the first two decades of life with
equal sex incidence in about 1 of 200,00 patients presenting with eye complaints.
The incidence is greater in neurofibromatosis. The typical presentation is visual im-
pairment in a verbal pre-school child with optic canal enlargement and optic atro-
phy. An intraorbital location of the tumor leads to axial, irreducible, non-pulsatile
proptosis. An intracranial location may disturb hypothalamic and pituitary
function and produce hydrocephalus. Ocular findings may also include limited
motility on a mechanical-restrictive basis, a papillary relative afferent defect,
nystagmus, and variable, non-specific visual field defects. Roentgenographic studies
may show concentric unilateral enlargement of the optic canal with preservation of
a well corticated margin, a fossa under the anterior clinoid process in continuity
with the optic canal ('J'-shaped sella), and findings of increased intracranial pres-
sure. On pathologic examination the tumor is a smooth, fusiform, intradural
enlargement of the optic nerve. Histologically there is proliferation of elongated
(pilocytic) astrocytes in reticulated patterns with intervening microcystic spaces
containing mucosubstance and surrounding reactive hyperplasia of the arachnoid.
Mitoses are not found. The diagnosis is clinical X-ray studies and brain scan should
be performed. The differential diagnosis is that of unilateral proptosis in a child and
includes acute ethmoiditis, hyperthyroidism, craniofacial, other neoplasms,
Hand-Schuller-Christian disease, and orbital hemorrhage due to trauma. Surgical
resection is performed in cases with unilateral optic nerve involvement, the surgical
approach being determined by tumor location. Bilateral or chiasmal cases are
treated with radiotherapy when progression occurs. Malignant optic nerve gliomas
and optic nerve hyperplasia are also discussed.

Optic nerve meningiomas arise from the nerve sheath and are to be distinguished
from orbital meningiomas arising from ectopic arachnoidal cells or those second-
darily involving the orbit by extension from adjacent sites. Up to 80% of orbital

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meningiomas occur in females, in two age peaks, 25% in the first decade, and the rest in the 5th decade. Meningiomas present with visual loss and may produce proptosis, papilledema and/or optic atrophy, retinal striae, opticociliary shunts, limitation of extra-ocular movements, and lid edema. Signs of von Recklinghausen's disease should be sought. X-rays are the mainstay of diagnosis. Orbital meningiomas are composed of cells in sheets or in whorls with some spindle shaped cells. Calcifications are typical. Usually the dura is penetrated and the orbit invaded. Primary orbital meningiomas are locally infiltrating but do not metastasize. Complete local excision en bloc is recommended. Local recurrences may require reoperation; however, long term survival is excellent.

Optic nerve head tumors are mostly benign hamartomatous proliferations of glial, melanocytic, or vascular tissue in the form of the astrocytic hamartomas of tuberous sclerosis, melanocytomas, and angiomatous malformations. There may also be primary or reactive proliferations of the juxtapapillary pigment epithelium. Metastatic lesions of the optic nerve are rare, breast and lung carcinomas accounting for most. The optic nerve may be infiltrated by neoplastic cells in the leukemias and lymphomas. The optic nerve may also be involved by inflammatory processes, notably sarcoidosis. Optic nerve medulloepitheliomas are highly malignant and must receive aggressive surgical treatment.