

Case 4

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Clinical Abstract:

This boy was first seen at 3½ years of age with a 2½ month history of inturning of the left eye, ataxia and headache. The optic nerve heads were pale and grey, but the visual fields and acuity could not be assessed. Skull x-rays showed suture separation and parasellar calcification. On pneumoencephalography the third ventricle was not visualized. A subtotal excision of a craniopharyngioma was carried out since the tumor was adherent to the optic nerve.

At 5½ years of age he received cobalt 60 radiation (4970 rad) because of recurrent vomiting and, one year later, a ventriculo-peritoneal shunt was placed. At 8 years of age he became ataxic, had frequent falls, and became deaf first in the left ear and subsequently bilaterally. He died at 13 years of age.

At autopsy there was fibrous thickening of the meninges about the optic nerves and chiasm which were flattened and encased in the scar tissue, but no gross or microscopic residual tumor was seen. However, keratin debris was plentiful in this region. At the level of the mammillary bodies, fibrosis and a partially calcified, grumous tumor were present.

In each cerebellopontine angle there was a small, dark, cystic mass. On the left side the eighth nerve was adherent to this mass, and on the right the seventh and eighth nerves were splayed over the mass.

MATERIAL SUBMITTED: One trichrome-stained section of tissue from cerebellopontine angle

Points for Discussion:

1. How often does this happen?
2. What is the relation of these tumors to epidermoid tumors?