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E F Downey, Jr and Z R Weinstein

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Unusual Case of Orbital Encephalocele

Edward F. Downey, Jr.,[1,2] and Zelig R. Weinstein[1,2]

Retinoblastoma is the most common intraocular neoplasm of childhood (frequency of 1:17,000–34,000 births) with bilateral lesions occurring in 25%–33% of cases [1, 2]. Other associated anomalies or tumors of the orbit have not been reported with retinoblastoma. We present a case of retinoblastoma in one globe and a presumed encephalocele in the opposite orbit located in an unusual area.

Case Report

A 1-year-old boy had ophthalmologic findings of a retinoblastoma in the right eye. Noncontrast and contrast computed tomography (CT) demonstrated a characteristic calcified lesion of retinoblastoma in the right globe and a nonenhancing, well circumscribed, extraconal mass in the superolateral aspect of the left orbit (fig. 1A). This mass, measuring 18 H, displaced the optic nerve medially. An area of hyperostosis was demonstrated in the lateral bony wall of the left orbit adjacent to the lesion, and there was widening of the superior orbital fissure (fig. 1B). Plain radiographs, including Caldwell views, were normal.

Ten days after the initial scan, 4.5 ml of metrizamide (concentration, 170 mg I/ml) was introduced into the basilar cisterns via a lumbar puncture, and routine axial and coronal CT scans were obtained. The study demonstrated a rim of metrizamide surrounding the soft-tissue mass, the density of which was unchanged from the earlier scan (figs. 1C and 1D). CT scans 4 hr later showed disappearance of the contrast material from the rim of the mass, with no increase in the overall density of the lesion. On the basis of this study, a diagnosis of encephalocele was made. Follow-up scans 19 months later demonstrated no change in the appearance or size of the left orbital lesion.

Discussion

This case is unusual in that the patient had a retinoblastoma with an incidental, yet significant, presumed encephalocele of the opposite orbit. Another possible explanation for this lesion would be an arachnoid cyst. There are reports of optic nerve arachnoid cysts occurring alone or in association with gliomas, neurofibromas, empty sellae, or porencephalic cysts [3, 4]. These cysts communicate with the subarachnoid spaces of the brain and optic nerve and would be expected to fill with contrast material during a metrizamide cisternogram [5]. However, in our case, the lesion was completely separate from the optic nerve, and no adjacent intracranial cyst was present.

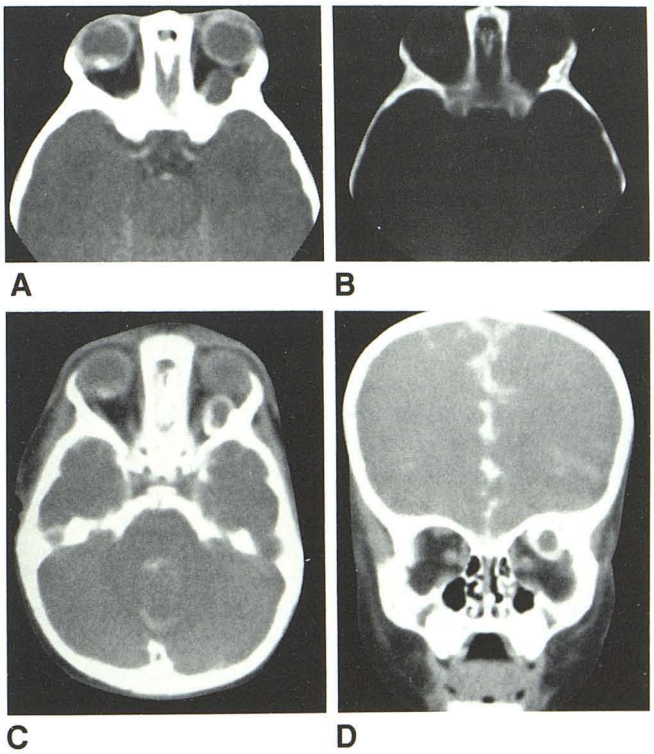

Fig. 1.—**A**, Axial CT scan. Calcified lesion of retinoblastoma in right globe. Low-density (18 H) mass in superolateral left orbit displacing optic nerve medially. **B**, Same scan with bone density setting. Hyperostosis of lateral orbital wall adjacent to lesion. Slight widening of superior orbital fissure. **C**, Axial metrizamide CT cisternogram. Rim of contrast around left orbital lesion. **D**, Coronal view. Lesion completely separate from optic nerve.

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The opinions expressed herein are those of the authors and are not to be construed as reflecting the views of the Navy Department, of the Naval Services at large, or of the Department of Defense.

[1] Department of Radiology, National Naval Medical Center, Bethesda, MD 20814.

[2] Department of Radiology/Nuclear Medicine, Uniformed Services University of Health Sciences, Bethesda, MD 20814. Address reprint requests to E. F. Downey.

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Reports of meningoceles and encephaloceles of the orbit have stressed the point that anterior encephaloceles are usually seen in the region of the glabella, medial orbit, or nasal cavity [6, 7]. More lateral orbital defects are extremely rare with only two cases reported to date [8]. In many cases of orbital encephaloceles there is a defect in osseous development that prevents proper ossification of bone. Bony fusion is incomplete with true herniations of intracranial contents through deformed or enlarged bony apertures such as the optic canal or superior orbital fissure [9]. In our case, only widening of the superior orbital fissure was demonstrated, and the precise point of communication between the orbit and intracranial contents was not seen. In some cases, neural dysgenesis with failure of normal separation of the surface ectoderm from neuroectoderm may be the cause of an orbital encephalocele [9]. Because of this faulty separation, a part of brain may be pinched off, completely separate from the intracranial contents. Other encephaloceles may retain a thin connection with the intracranial contents by a cord of tissue that may include meninges [9]. Our case appears to be an example of this type of maldevelopment. Although the pathway by which the metrizamide entered the encephalocele from the intracranial subarachnoid space could not be demonstrated with certainty, the connection is presumably through the superior orbital fissure.

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