Orbital Hemangiopericytoma

Review of the Literature and Report of Four Cases*

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STOUT and Murray,21 in 1942, first described the hemangiopericytoma and demonstrated by tissue cultures17,21 that the cell of origin was the capillary pericyte of Zimmermann. As described by Zimmermann,22 the pericyte is a specific type of cell related closely to smooth-muscle cells but having no contractile fibers. Included in the initial report of Stout and Murray was a case of hemangiopericytoma arising intra-orbitally and extending into the orbit, and in a subsequent report by Stout20 2 more cases of orbital hemangiopericytoma were added to the literature, one starting in the ethmoid sinus and invading the orbit and the other beginning in the subcutaneous tissue over the middle of the infraorbital ridge. In the latter case, a single surgical procedure was performed; in the other multiple operations were done. In 1955, Goodman6 and Fox5 almost simultaneously reported the first hemangiopericytoma that was primary in the orbit; these patients also underwent multiple surgical procedures. Since then only 3 more orbital hemangiopericytomas have been reported,9,10,19 1 of them occurring in a child.10 All of these tumors were treated surgically by anterior orbitotomy, and 2 of them promptly recurred. MacCarty and Brown,12 in a report on 40 children with orbital tumors that were removed by the transcranial approach, did not list any hemangiopericytomas.

The following case reports involve hemangiopericytomas arising in the orbit and removed transcranially. Two of these patients had previously undergone operation by the anterior approach.

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Report of Cases

Case 1. A 63-year-old man was first seen in February 1960, at the age of 60, with a history of unilateral exophthalmos for 26 years. In 1934, a diagnosis of orbital tumor had been made elsewhere; the patient had refused operation but had received radiation therapy resulting in some recession of the protruding eye but never to normalcy. The proptosis began to increase in January 1939. In 1960 visual acuity was 20/70 on the left and 20/20 on the right. Hertel exophthalmometric measurements were O.D. 16 mm. and O.S. 25 mm. There was grade I paresis of ocular rotation in the left eye. The fundi were normal. Shit zest tension was 14.6 in each eye. Visual field was normal in the left eye but there was a scotoma in the right eye. Roentgenograms of the skull taken elsewhere showed enlargement of the left orbit, and a left carotid angiogram revealed a highly vascular spherical tumor behind the globe. The patient was unwilling to accept the risk of visual loss in the affected eye that could possibly arise secondary to operation and was therefore given a course of radiation therapy, which had no effect.

The patient returned to the Mayo Clinic in April 1963, at age 63, because of further increase in proptosis, pain in and around the eye, and gradual loss of vision to complete blindness in the left eye.

Examination revealed a pulsating exophthalmos; a blind left eye with normal visual acuity on the right; grade 2 limitation of ocular movements on the left; Hertel exophthalmometric measurements of O.D. 18 mm. and O.S. 32 mm.; Shitz tension of O.D. 10.2 mm. and O.S. 15.9; negative funduscoptic findings; and unchanged visual field of the right eye. Laboratory investigation was noncontributory.

A left carotid angiogram revealed a vascular tumor in the left orbit with feeding vessels from the internal carotid system and from an enlarged ophthalmic artery (Fig. 1).

Left frontal craniotomy and left blepharorhaphy were performed. The roof and the lateral wall of the orbit were very thin. The tumor, which appeared in the superior lateral aspect of the globe, was entered by many large vascular channels, and it measured 4.5X3X3 cm. The mass was removed totally (Fig. 2).
Postoperative course was uneventful, and the patient was dismissed 6 days after operation. The visual acuity in the left eye improved to 20/200 and 20/70 with a pinhole, remaining 20/20 in the right eye. Fundi were normal, although the left disk might have been a little lighter in color. Hertel measurements were O.D. 17 mm. and O.S. 26 mm., and rotations had returned to normal. The visual field of the left eye exhibited a temporal and central scotoma to the 2-mm. white bead at 1 meter and was full to the 10-mm. bead at 1 meter; the visual field of the right eye was unchanged.

Case 2. A 36-year-old woman had come to the ophthalmologic section in March 1947, at the age of 21, with the complaint of painless swelling of the left upper eyelid for 18 months. At that time, examination revealed that the globe was pushed downward, and a rather irregular, freely movable mass, measuring about 1×2 cm., with a wormy feeling to palpation, was present in the upper temporal portion of the orbit just beneath the orbital rim. Exophthalmometric readings were O.D. 19 mm. and O.S. 21 mm., and fundi were normal, as were the visual fields.

Subtotal removal through an anterior orbitotomy was performed by an ophthalmic surgeon. The tumor extended deeply into the muscle cone, and the posterior limits of the tumor could not be defined. The mass measured 2×3×4 cm.

The pathologic diagnosis at that time was malignant hemangioendothelioma, and a postoperative course of radiation therapy was administered.

The patient returned in May 1959, because of recurrence of the mass in the left upper lid.

A large mass was palpable, being movable anteriorly but fixed deeply. Palpebral fissures measured O.D. 9 mm. and O.S. 6 mm.; Hertel exophthalmometric readings were O.D. 16 mm. and O.S. 17 mm.; and extraocular movements were normal. There was change in the left eye indicative of early cataract, but fundi and visual fields were normal. Orbital roentgenograms were negative. It was felt that nothing more should be done at that time.

The previous pathologic diagnosis was amended to cellular hemangioma.

The patient returned again in April 1961, because of marked progression of the swelling of the left lid. The mass was now approximately 15×12 mm.; the left globe was 5 mm. lower than the right; palpebral fissures measured O.D. 10 mm. and O.S. 2 mm.; and Hertel exophthalmometric measurements were O.D. 17 mm. and O.S. 25 mm. Radon seeds were inserted, but the swelling again worsened in the fall of 1961.

The patient returned to the ophthalmologic section in February 1962, because of further swelling, but Hertel measurements were unchanged. There was an increase in the denseness of the posterior subcapsular cataract of the left eye. It was felt that further radiation therapy

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Fig. 1. Visualization by angiography of intraorbital hemangiopericytoma by means of the subtraction read-out technic.

Fig. 2. Tumor in superior lateral aspect of globe, 4.5×3×3 cm., was entered by many large vascular channels.