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Giant Cell Granuloma of the Orbit with Intracranial Extension

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An 8-year-old boy presented with acute proptosis of the right eye. Examination revealed 8 mm of exophthalmos, limitation of upward gaze, optic disc swelling, and normal visual acuity, but an inferonasal quadrantic visual field defect. Orbital ultrasound and computed tomographic scanning demonstrated a superotemporal tumor of the right orbit with intracranial extension. At operation, this proved to be a "reparative" giant cell granuloma. After partial resection, the remaining mass resolved spontaneously without further specific treatment. There had been no recurrence at the 1-year follow-up examination. (*Neurosurgery* 16:75–81, 1985)

Key words: Exophthalmos, Optic disc swelling, Intracranial extension, "Reparative" giant cell granuloma

During the past 3 decades, there has been an increasing awareness that not all giant cell lesions of the bone are benign giant cell tumors. In 1952, Jaffe pointed out the benign nature of so-called "reparative" giant cell granuloma, its predilection for the young, and its unusual histological pattern (7). In 1969, Friedberg et al. noted the frequency with which reparative giant cell granuloma is misdiagnosed as giant cell tumor (2). In 1974, Hirschl and Katz outlined specific criteria for discriminating between these two kinds of lesions on the basis of histopathology (5). These authors also pointed out the frequency with which reparative giant cell granuloma occurs outside the jaw bone.

There have been five reported cases of reparative giant cell granuloma with orbital involvement (2, 3, 6, 9). Intracranial extension was present in one case (6). The following case is presented because of the rarity of this disease in the orbit, especially with intracranial involvement, and because of the unique presentation and clinical course.

CASE REPORT

C.B. was an 8-year-old white boy who presented with the chief complaint of proptosis O.D. of 2 weeks duration. Examination of old photographs revealed that minimal proptosis O.D. had been present 3 years before presentation. There were no ocular, constitutional, or neurological symptoms. The medical and ocular histories were unremarkable.

Physical examination revealed an alert, cooperative, well-developed, well-nourished, right-handed boy in no apparent distress. His vital signs were normal. The visual acuity was 20/20 O.U. and color vision was intact O.U. There was 8 mm of proptosis O.D. (Fig. 1). Extraocular movements showed marked limitation in upward gaze O.D. There was no afferent pupillary defect. Slit lamp examination and intraocular pressures were normal. Fundus examination revealed swelling of the right disc (Fig. 2). Visual field testing demonstrated an

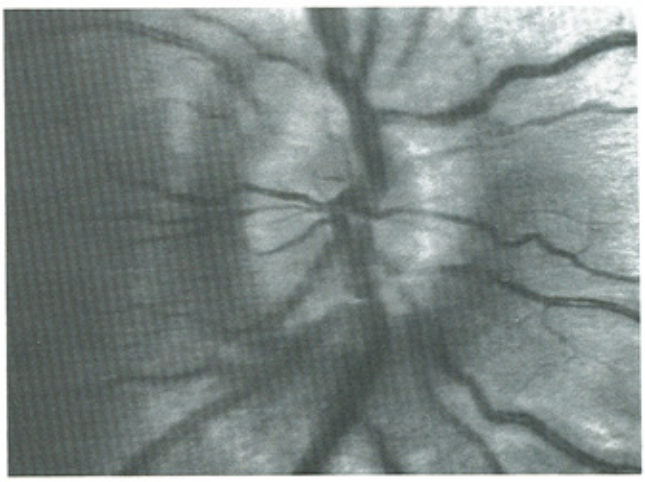

FIG. 2. Fundus photograph demonstrating swelling of the right optic disc.

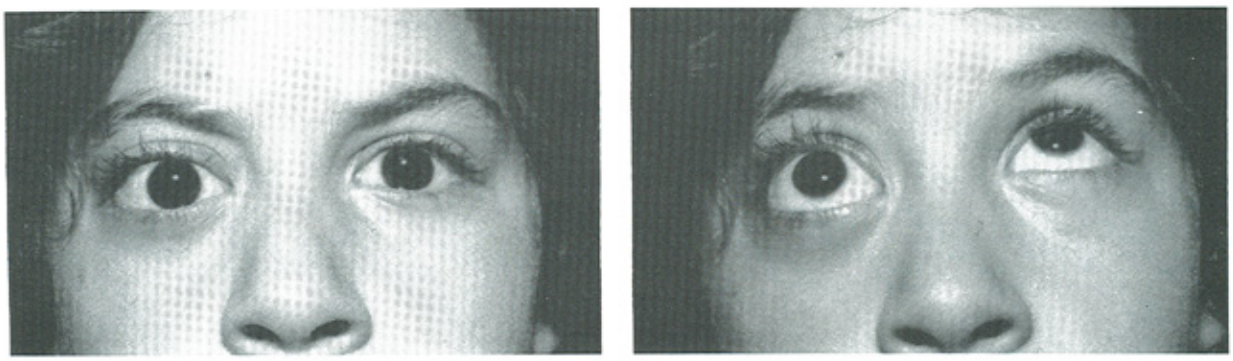

FIG. 1. This 8-year-old boy has 8-mm proptosis and limitation in upward gaze O.D.

inferonasal quadrantic defect O.D. Ultrasound showed a well-demarcated mass in the superotemporal orbit with heterogeneous internal echoes (Fig. 3). Polytomographic x-ray films revealed erosion of the lesser wing of the sphenoid and the upper portion of the greater wing of the sphenoid. There was also erosion of the posterior portion of the orbital roof. A computed tomographic (CT) scan revealed a superior orbital mass with intracranial involvement superiorly and laterally (Fig. 4).

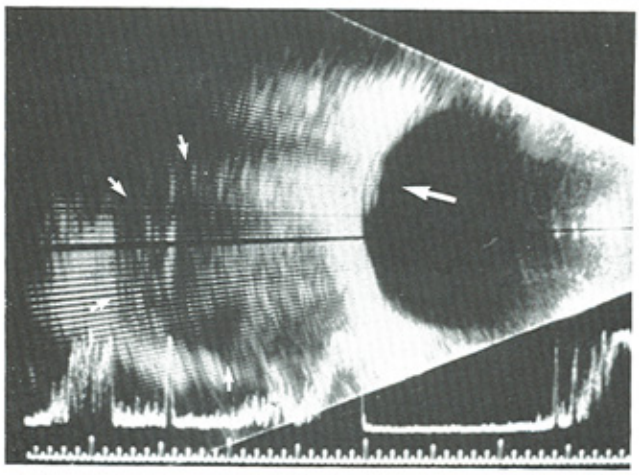

FIG. 3. Orbital ultrasound showing the retrobulbar tumor (*small arrows*) with heterogenous internal echoes. Elevation of the optic disc was also noted (*large arrow*). (Courtesy of Dr. Peter Lou.)

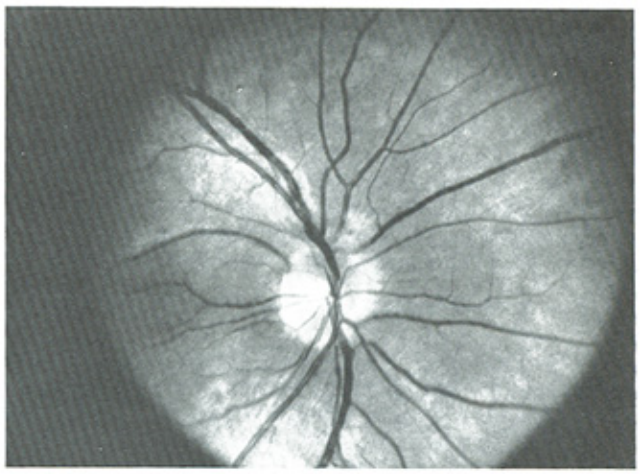

FIG. 6. Postoperative resolution of optic disc swelling.

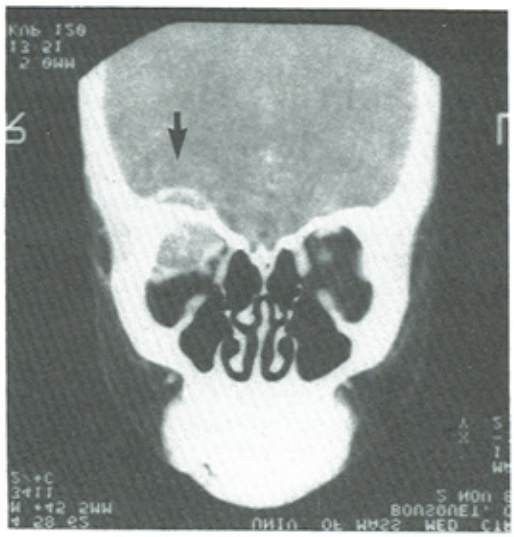

FIG. 4. Computed tomographic scan demonstrating the orbital tumor with a contrast-enhancing periphery. Coronal views revealed the presence of intracranial extension (*arrow*). (Courtesy of Dr. A. Weber.)

FIG. 5. Postoperative resolution of proptosis and gaze paresis.

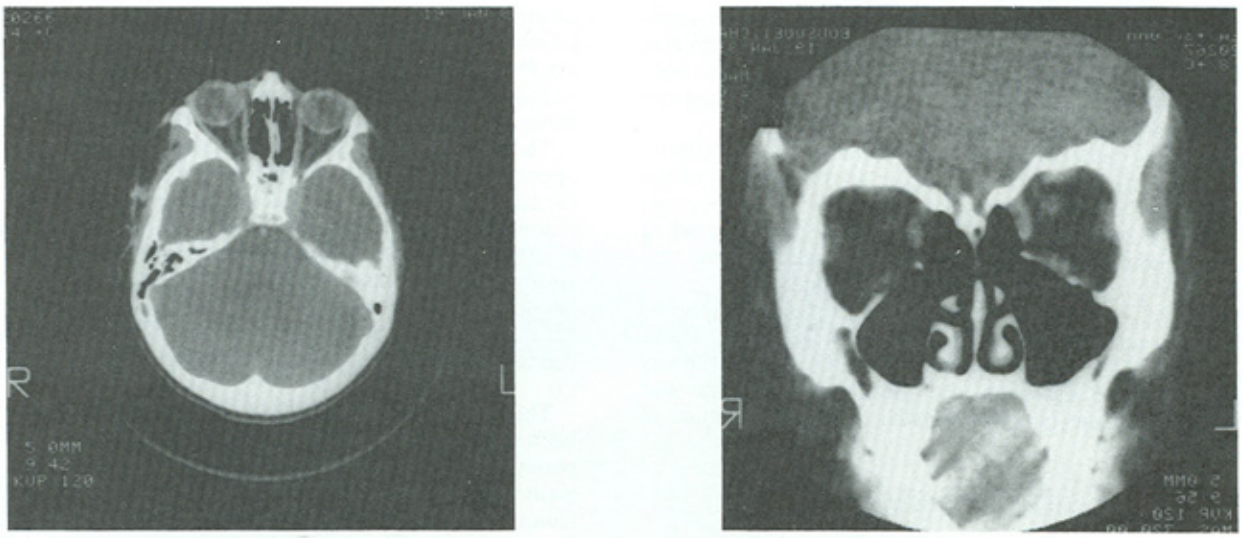

FIG. 7. Computed tomographic scan performed 2 months postoperatively demonstrating the absence of an orbital tumor.

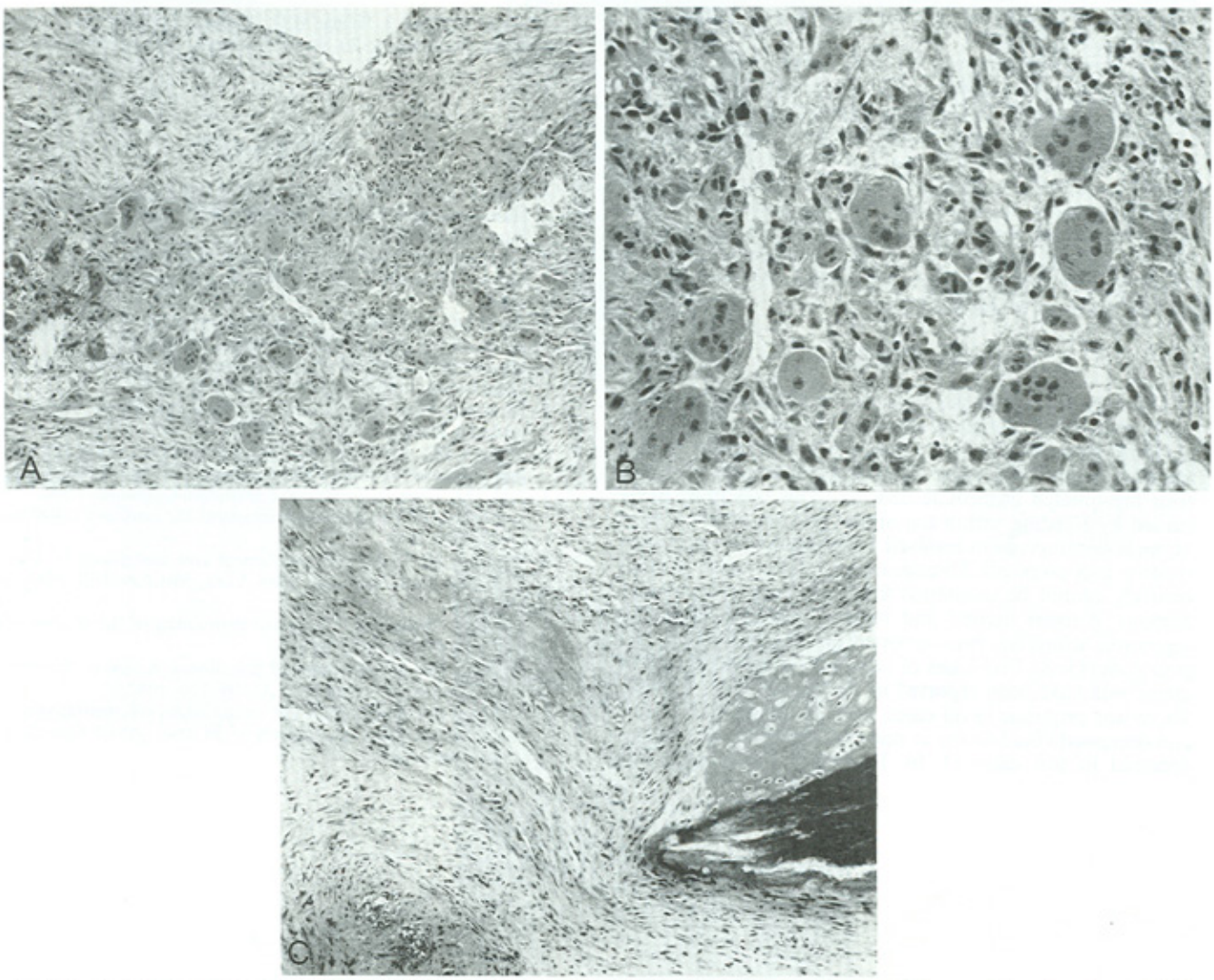

FIG. 8. *A*, multinucleated giant cells surround a focus of hemorrhage (*left*) within a fibroblastic stroma. (Hematoxlin and eosin, × 160.) *B*, giant cells, activated fibroblasts, and scattered lymphocytes. (Hematoxlin and eosin, × 400.) *C*, new bone formation and fibrous tissue. A focus of hemorrhage is at the *lower left*. (Hematoxlin and eosin, × 160.)

Hematological and serum chemistry studies were unremarkable except for an alkaline phosphatase value of 86 IU/litre (17–60 is normal) and an inorganic phosphorous value of 4.8 mm/100 ml. Bone marrow biopsy and cerebrospinal fluid analyses were unremarkable.

The patient underwent a right frontotemporal craniotomy with orbital exploration. In the region of the junction of the roof and the superolateral wall of the orbit, a fleshy, red, somewhat hemorrhagic tumor was encountered extending into the epidural space. The mass was partially resected and a small cavity containing old blood was evacuated.

Postoperatively there was resolution of proptosis (Fig. 5) and disc swelling (Fig. 6) with recovery of full visual fields. Repeat CT scanning performed 2 months postoperatively showed complete regression of the tumor (Fig. 7). One year postoperatively the patient was well, with no evidence of recurrence.

HISTOPATHOLOGY

Several fragments of soft hemorrhagic tissue were obtained at operation. Microscopic examination showed scattered groups of multinucleated giant cells set in predominantly fibrous stroma with foci of hemosiderin and woven bone formation (Fig. 8*A*).

The giant cells contained up to 12 nuclei with indistinct nucleoli (Fig. 8*B*). Several contained phagocytosed hemosiderin. The giant cells occurred in groups with a granulomatous appearance. Between the giant cells were stromal cells of moderate size with round to oval nuclei. Although trichrome stains showed slight collagen formation around giant cell aggregates, this was markedly less than that seen in the fibrous areas. The fibrous areas were composed of spindle cell fibroblasts and dense collagen. Scattered throughout were spicules of woven bone (Fig. 8*C*). These are the histological characteristics of giant cell granuloma.

DISCUSSION

This tumor was a benign, chronic, "reparative" granuloma of the orbit with intracranial extension. In view of slight, long-standing proptosis, the tumor had presumably been present for a few years. Sudden and substantial increase in the proptosis precipitated immediate concern. This was most likely caused by bleeding within the orbital portion of the tumor. Surgical decompression resolved the compressive optic neuropathy and proptosis. Disappearance of the tumor postoperatively cannot be accounted for by the relatively modest amount of tissue excised and must represent spontaneous regression, which has been reported previously for giant cell granuloma (5, 8). Five cases of orbital "reparative" giant cell granuloma have been reported in the literature (2, 3, 6, 9). There was proptosis in all cases, gaze paresis in three cases, and decreased visual acuity in one case. Multiple recurrences occurred in two cases (3, 6). In one of these cases, two recurrences were unsuccessfully managed with operation. This lesion was treated with radiotherapy and ultimately resolved (3). However, several authors advise the use of only low doses of radiation because of the risks of sarcomatous and carcinomatous transformation after radiotherapy (1, 4).

The absence of a history of trauma, infection, or previous operation in this case underscores the inappropriateness of the term "reparative." As others have suggested (6, 11), such lesions are more accurately named giant cell granuloma. The cause of this curious tumor remains obscure. Although some authors thought that the lesion was caused by trauma, a history of trauma was not elicited in many cases. Hirshl and Katz have proposed that local chronic inflammation giving rise to microhemorrhages may cause the "reparative process" (5).

The origin of the giant cells likewise remains unclear. It has been suggested that osteoclasts, undifferentiated stromal cells, pericytes, and macrophages may serve as progenitors of the giant cells and that a lymphokine called "macrophage fusion factor" from T-cell lymphocytes may stimulate cell fusion to form giant cells. This subject has been reviewed by Singh (10).

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