

# Granular Cell Tumor of the Orbit

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**Purpose:** To report a case of granular cell tumor as a rare orbital pathology.

**Case report:** A 50-year-old female presented with a 4-year history of diplopia, right ocular displacement and a firm nontender mass in her right lower lid. Computed tomography (CT) scan of the orbit disclosed a well-defined mass in the right inferior orbit involving the right inferior rectus. Subtotal excision of the mass was performed, and histopathologic and immunohistochemical studies revealed granular cell tumor. Subsequently, the tumor recurred and exenteration was required as multiple sessions of radiotherapy failed to prevent the residual tumor from growing.

**Conclusion:** Granular cell tumor, though very rare in the orbit, should be considered in patients with orbital masses especially in cases with involvement of the inferior rectus muscle. Infiltrative tumors may be impossible to completely resect and can rapidly recur following surgery.

**Keywords:** Orbit; Granular Cell Tumor; Radiotherapy; Exenteration

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## INTRODUCTION

Granular cell tumor (GCT), previously referred to as granular cell myoblastoma, is a rare benign soft tissue tumor that may involve the orbit, periocular skin, lacrimal sac, optic nerve, ciliary body, conjunctiva and caruncle, in addition to non-ocular tissues such as the skin, gastrointestinal, respiratory and genital tracts, peripheral nerves, and other organs.<sup>1,2</sup> This tumor was first described and denominated in 1926 by Abrikosoff as "myoblastic myoma".<sup>3</sup> Various tissues have been proposed as the origin of GCT including striated muscle, histiocytes, fibrocytes and mesenchymal cells. However, recent ultrastructural and immunohistochemical studies suggest Schwann cells as the probable origin of the tumor.<sup>4</sup> Although GCT is often described as a discrete benign mass which is

well-delineated from adjacent tissues in the orbit, there are some reports of malignant or infiltrating orbital GCTs.<sup>1,5</sup>

Herein, we present a case of infiltrating and progressive GCT leading to orbital exenteration.

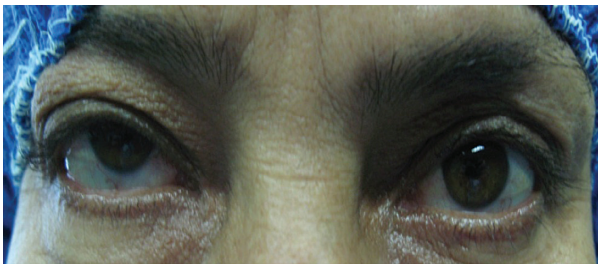
## CASE REPORT

A 50-year-old woman presented with a 4-year history of vertical diplopia and painless superior displacement of the right eye. She had no history of ocular discharge or thyroid eye disease. Biopsy of the right orbital mass three years before had yielded a non-specific diagnosis.

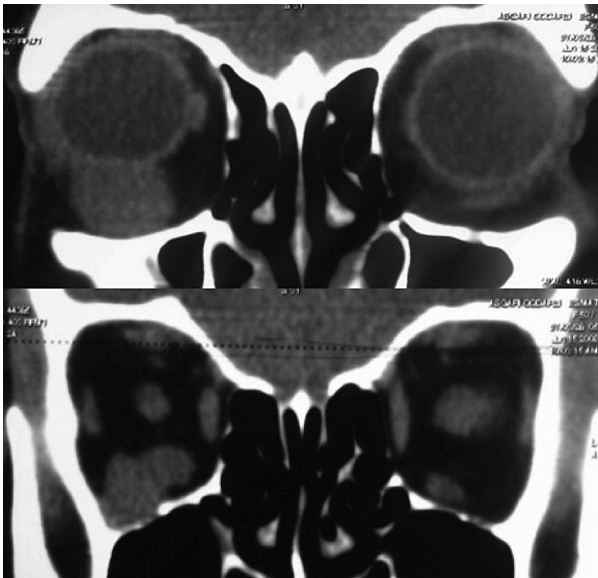
On admission, best corrected visual acuity (BCVA) was 20/30 and 20/20 in the right and left eyes, respectively; a mild afferent pupillary defect was also noted in the right eye. There was a firm non-tender mass palpable through the right

lower lid producing a small amount of proptosis and upward displacement. Exophthalmometric values were 18 mm and 14 mm on the right and left sides, respectively (Fig. 1). A significant reduction in ocular abduction and depression was noted without orbital bruits or pulsation. Slit lamp biomicroscopic examination was unremarkable but funduscopy disclosed right optic disc swelling nasally. Ishihara test revealed red-green color deficiency and visual field testing demonstrated a cecentral scotoma in the right eye. Intraocular pressure was 15 mmHg bilaterally.

Orbital computed tomography (CT) scan showed enlargement of the right inferior rectus muscle which was infiltrated by a muscle-density mass displacing the globe upward; maxillary sinus involvement and bone erosions were not observed (Fig. 2).



**Figure 1.** Right lower lid mass together with vertical ocular displacement preoperatively.



**Figure 2.** A well-defined mass in the right orbit with inferior orbital muscle involvement on computed tomography.

Due to close proximity of the tumor with inferior orbital muscles, a subciliary approach was employed to excise the tumor. Under general anesthesia, the anterior portion of the mass was excised as much as possible and sent in 10% formalin solution for histopathologic evaluation. Complete excision of the mass was not feasible due to infiltration of the tumor within the rectus muscle and other orbital structures.

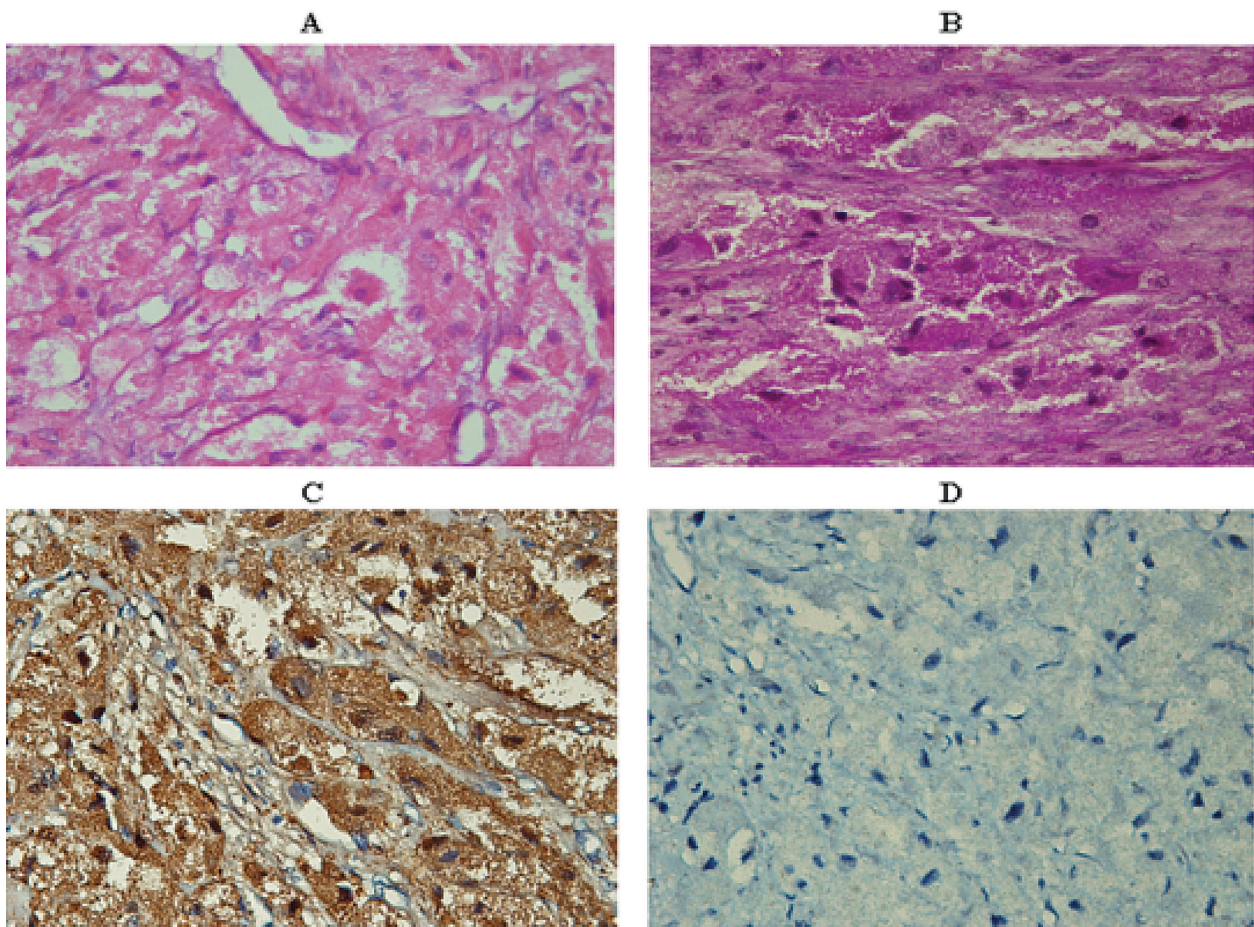
Gross examination revealed a cream-colored to brownish firm mass measuring 20×16×12mm. After tissue processing and embedding in paraffin blocks, thin sections were stained with hematoxylin and eosin (H&E) and periodic acid-Schiff (PAS). Microscopic examination disclosed a partially encapsulated mass composed of lobules and cords of polyhedral and spindle-shaped cells with bland nuclei and intracytoplasmic PAS-reactive granules (Figures 3A and 3B); however, mitosis was not a feature. The tumor cells were strongly immunoreactive for S100 in both nuclear and cytoplasmic areas but not for HMB45 and pan-cytokeratin immunostaining (Figures 3C and 3D). These histopathologic features were consistent with GCT.

During a six-month postoperative course, BCVA of the right eye decreased gradually and multiple sessions of radiotherapy, summing to 40 Gy in 2 Gy fractions, failed to control the growth of the residual tumor. Eventually vision dropped to no light perception and exenteration of the right orbit was performed to prevent extension of the tumor to the optic canal.

## DISCUSSION

Granular cell tumors are uncommon benign soft tissue tumors with the basic histopathologic feature of a granular cytoplasm which may occur anywhere in the body.<sup>1</sup> Presence of chorioembryonic antigen as a sign of primitive cell origin, immunoreactivity of tumor cells for both S100 protein and myelin basic protein, and presence of basement membrane material around tumor cells on electron microscopy indicate a modified Schwann cell origin for these lesions.<sup>6</sup>

Approximately, 3% of GCTs arise in the orbit.<sup>7</sup> In a review of 31 cases with orbital and periorbital GCT, average age was 40 (range 3 to



**Figure 3.** A, the tumor was composed of cords of polyhedral cells with bland nuclei and prominent intracytoplasmic granules (hematoxylin and eosin stain,  $\times 400$ ); B, note periodic acid-Schiff (PAS)-reactive intracytoplasmic granules (PAS stain,  $\times 400$ ); C, strong reactivity of the tumor cells for S100 immunostaining ( $\times 400$ ); D, lack of reactivity for HMB45 immunostaining ( $\times 400$ ).

74) years with no gender preference but varying duration of signs and symptoms, from weeks to years.<sup>4</sup> Our patient was a middle-aged woman with a four year history of diplopia and ocular displacement due to an orbital GCT with typical histopathologic and immunohistochemical features of this tumor. Furthermore, our patient showed evidence of optic nerve compression such as reduced vision, afferent pupillary defect, optic disc swelling and abnormalities in color vision and visual fields.

The most common reported location for an orbital GCT is the inferior half of the orbit with extraocular muscle involvement, especially the inferior rectus muscle, as a prominent feature<sup>8,9</sup> which was observed in our patient. In a literature review by Ribeiro et al including 40 cases of GCT, ocular dysmotility was present in 79% of

patients and the inferior rectus was the most commonly involved muscle with diplopia present in most cases.<sup>8</sup>

Although orbital GCT is usually described as a circumscribed lesion, there are reports of infiltrating lesions.<sup>5</sup> In our patient, the tumor seemed rather localized on orbital CT; however, we encountered a diffuse and infiltrating tumor within the inferior rectus muscle and orbital tissues rendering complete resection of the tumor impossible.

Tumor excision is recommended for orbital GCTs. Nevertheless, this might be difficult in cases with close association of the tumor with an extraocular muscle due to possibility of permanent muscle dysfunction. In our patient, the condition continued to progress after the operation leading to optic nerve involvement

and visual loss. In addition, radiotherapy was performed for the patient in a standard fashion but was ineffective. There are a few reports in which radiotherapy had not proven helpful for the management of GCTs.<sup>5,10,11</sup>

Eventually, we decided to perform orbital exenteration to prevent intracranial extension of the tumor. The reported local recurrence rate of GCT is about 7%<sup>5</sup>, however there is no report describing orbital exenteration for management of GCT.<sup>5</sup> The current case may demonstrate that an infiltrative GCT which is not completely resectable, may rapidly recur and diffuse within the orbit.

In summary, we present a rare case of orbital GCT with progressive growth and rapid recurrence following incomplete excision, eventually necessitating orbital exenteration.

#### Conflicts of Interest

None.

#### REFERENCES

1. Callejo SA, Kronish JW, Decker SJ, Cohen GR, Rosa RH Jr. Malignant granular cell tumor metastatic to the orbit. *Ophthalmology* 2000;107:550-554.
2. Ordóñez NG, Mackay B. Granular cell tumor: a review of the pathology and histogenesis. *Ultrastruct Pathol* 1999;23:207-222.
3. Abrikosoff A. Über myoma ausgehend von der quergestreiften willkürlichen muskulatur. *Virchows Arch Pathol Anat* 1926;260:215-233.
4. Jaeger MJ, Green WR, Miller NR, Harris GJ. Granular cell tumor of the orbit and ocular adnexae. *Surv Ophthalmol* 1987;31:417-423.
5. Dolman PJ, Rootman J, Dolman CL. Infiltrating orbital granular cell tumour: a case report and literature review. *Br J Ophthalmol* 1987;71:47-53.
6. Mukai M. Immunohistochemical localization of S-100 protein and peripheral nerve myelin proteins (P2 protein, P0 protein) in granular cell tumors. *Am J Pathol* 1983;112:139-146.
7. Moseley I. Granular cell tumor of the orbit: radiological findings. *Neuroradiology* 1991;33:399-402.
8. Ribeiro SF, Chahud F, Cruz AA. Oculomotor disturbances due to granular cell tumor. *Ophthalm Plast Reconstr Surg* 2012;28:e23-27.
9. Poyraz CE, Kiratli H, Söylemezoğlu F. Granular cell tumor of the inferior rectus muscle. *Korean J Ophthalmol* 2009;23:43-45.
10. Karcioğlu ZA, Hemphil GL, Wool BM. Granular cell tumor of the orbit: case report and review of the literature. *Ophthalmic Surg* 1983;14:125-129.
11. Kubacz GJ. Malignant granular cell myoblastoma: report of a case. *Aust N Z J Surg* 1971;40:291-295.