# Intraorbital Granular Cell Tumor Involving in Medial Rectus and Optic Nerve

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To the Editor: Here, we present a rare case whose medial rectus and optic nerve in the left eye are both affected by intraorbital granular cell tumor (GCT). Macroscopic size of the mass was around 4.3 cm × 2.2 cm × 1.8 cm.

A 39-year-old Chinese woman presented as progressive exophthalmos, diplopia, and photophobia in her left eye for the past one year. No history of ocular discharge or thyroid eye disease was recorded. Her best-corrected visual acuity was 1.0 and 0.15 in the right and left eyes, respectively. Intraocular pressure in the right eye was 15.2 mmHg (1 mmHg=0.133 kPa) and 12.3 mmHg in the left eye. Clinical examinations showed top-left displacement of the left eye [Figure 1a]. Abnormal protrusion of the left eye was found. Fundoscopy disclosed left optic disc swelling. Obvious limited adduction and abduction of the left eye globe were observed. B-scan showed a roundish dark lesion behind the globe wall [Figure 1b]. The neoplasm was confirmed by computed tomography (CT) scan, and the image disclosed a muscle density, well-defined, ovoid retrobulbar nodule measuring 2.8 cm × 1.9 cm between the optic nerve and medial rectus. The boundary between the lesion and medial recuts was obscure, and the optic nerve was squeezed outward by the neoplasm [Figure 1c]. No bony erosion was found [Figure 1d].

Surgery was performed under general anesthesia. The tumor was resected through a lateral orbital approach. The mass was carefully separated from the medial rectus and optic nerve, then completely removed. After the swelling went down, we found her visual acuity in the left eye dropped to no light perception. Eye globe movement was partly limited. CT-scan found no recurrence at her 7-month follow-up appointment [Figure 1e]. However, visual acuity and eye movement were not improved markedly.

Histological analysis was performed postoperatively. Macroscopic examination showed that the mass has the dimension of 4.3 cm × 2.2 cm × 1.8 cm [Figure 1f]. Light microscopy manifested that the cells had granular cytoplasm with ill-defined borders. A few fibrous connective tissues separated the tumor cells. Higher magnification showed that tumor cells were round to polygonal and the cytoplasm was fully eosinophilic. Tumor nuclei were homogeneous small, but no marked mitotic activity or cytologic atypia were observed [Figure 1g]. Immunohistochemistry showed the strong positive staining for S-100 in both nuclear and cytoplasmic areas [Figure 1h], but not for CD68. Then, diagnosis of orbital GCT was established by these histopathologic features.

GCT was first described by Abrikossoff as "myoblastoma" in 1926. This uncommon neoplasm can occur in any part of the body, but it rarely affects the orbit. It is estimated that only 3% arise in orbit in all GCT cases.[1] Until now, approximately 54 cases have been reported in English literature.[2] In 2011, Ribeir *et al*.[1] reviewed most of the literature. Furthermore, it can occur in the lacrimal sac, conjunctiva, uvea, and eyelids. For these intraorbital GCT cases, extraocular muscle involvement (79.3%) and diplopia (84.6%) are the most common features.[1] The tumor mostly located in the lower half of the orbit (58.3%) and the inferior rectus was the most commonly involved muscle (38.5%).[1]

Usually, GCTs are benign tumors, and complete surgical removal is the best choice. However, 2–9.68% of them are malignant.[3] Salour *et al*.[4] reported one case who rapidly recurred after subtotal excision of the tumor. The distinction between benign and malignant cases is based on histopathological features. Enlarged vesicular nuclei with prominent nucleoli, spindling of tumor cells, high nuclear-to-cytoplasmic ratio, nuclear pleomorphism, appreciable mitotic activity, and tumor necrosis should be suspicious of malignant GCT.[5]

In the past, usually GCT was considered as granular cell myoblastoma, but immunohistochemistry confirmed Schwann cell as the most likely cell of origin for the tumor in recent years. The close association between GCT and extraocular muscle might result from the multiply innervation of the muscle fibers.[1] Almost all cases showed positive staining for both S-100 and CD68.[1,4,5] The positive stain with CD68 can be explained by the intracytoplasmic accumulation of phagolysosomes and does not reflect a histiocytic origin. It is, therefore, not surprising that the case showed negative staining with CD68.

In summary, one rare case whose medial rectus and optic nerve were squeezed by intraorbital GCT was presented here. The typical

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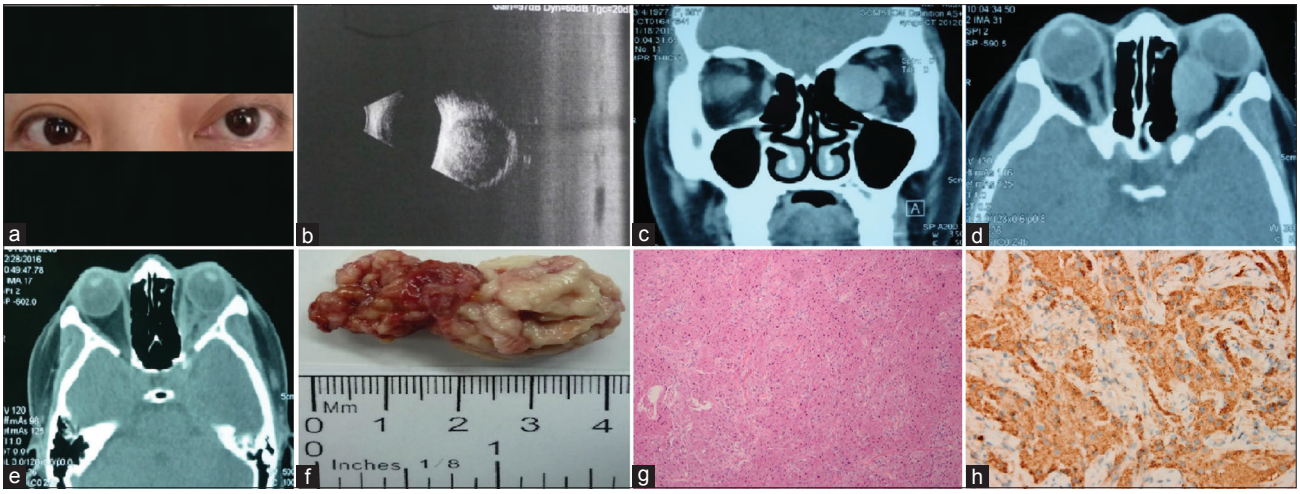


**Figure 1:** (a) Top-left displacement of the left eye; (b) B-scan revealed the roundish dark lesion behind the globe wall; (c) Pre-operative coronal computed tomography image; (d) axial computed tomography images; (e) Post-operative axial computed tomography image; (f) Macroscopic picture of the neoplasm; (g) Eosinophilic cells with abundant granular cytoplasm and small oval nuclei, with no marked mitotic activity or cytologic atypia (hematoxylin and eosin staining, original magnification ×100); (h) S-100 immunostaining (original magnification ×400).

histopathological feature is very helpful for the correct diagnosis and judgment of prognosis. Completely surgical excision of the tumor is one of the therapeutic choices.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.


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## Conflicts of interest

There are no conflicts of interest.

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