Case report of granular cell myoblastoma arising within the medial rectus muscle

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SUMMARY  A rare case of granular cell myoblastoma is reported in a 37-year-old man where the tumour was situated within the medial rectus muscle. Histological, cytochemical and ultrastructural examination showed it to have features typical of granular cell myoblastoma. It also reacted with chorioembryonic antigen antisera, a sign of primitive cell origin. The findings add further weight to the idea of the histiogenesis of these tumours from primitive mesenchymal cells.

Since Abrikossoff1 first described granular cell myoblastoma (GCM) in 1926 this tumour has been found in many parts of the body, most commonly the tongue. Cases have been reported involving the lids,2,4 eyebrow,5 lacrimal sac,6 and the ciliary body.7 Orbital GCM is rare, and only 11 cases have previously been recorded.2,5,6,10-16 This case is presented for 3 reasons: first, the rarity of granular cell myoblastoma in the orbit; secondly, the unusual presenting feature of visual obscuration related to eye position; and lastly the good recovery of eye movements and visual acuity following the resection of a large tumour from within the orbit, particularly as it was within one of the extraocular muscles.

PHYSICAL SIGNS
On examination visual acuity was 6/5 unaided in the right eye and 6/12 unaided in the left eye. Extraocular movements showed slight limitation of abduction and some left incycloduction and left sursumduction, with left over right in the primary position. The left eye was proptosed 4 mm axially and displaced 2 mm laterally (Fig. 1). Anisocoria was present with the left pupil larger than the right. On prolonged laevoversion an afferent pupillary defect was noted which disappeared on regaining the primary position. Fundal examination showed left optic disc swelling, with venous engorgement and choroidal folds running through the macular area.

Fig. 1  Left proptosis and lateral displacement of the globe at the time of presentation.

Case report

HISTORY
A 37-year-old man complained of mild pain around his left frontal area for 12 months. For the latter 8 months of this time he had also noted diplopia on laevoversion and increasing visual distortion in the left eye. As well as diplopia colour vision was reduced, and he had visual obscurations after prolonged laevoversion which cleared on regaining the primary position. At the time of the presenting symptoms he had a daughter of 5½ years who was undergoing treatment for cerebral astrocytoma. The patient’s father had died from cerebral glioma.

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INVESTIGATIONS
Plain radiographs of the orbit were normal. CT scans showed a large cigar-shaped mass which was indistinguishable from the medial rectus muscle and displayed contrast enhancement (Figs. 2, 3). Carotid angiography was performed, but no abnormal circulation could be detected. Fluorescein angiography showed a reduced blood flow in the retinal circulation when the left eye was held in abduction (Fig. 4).

TREATMENT
A lateral orbitotomy was performed. The medial rectus muscle was approached across the intracanal space, and the optic nerve was displaced downwards. A large pale tumour was found in the medial rectus muscle. The muscle was split, longitudinally on its inner surface, posteriorly between the 2 branches of the third nerve supplying the muscle and anteriorly to its insertion on the globe. The tumour was shelled out from the muscle fibres within which it lay and was totally removed.

Postoperatively vision in the left eye was reduced to 6/18 and adduction was absent. After 10 weeks the vision had returned to 6/6, with disappearance of the choroidal folds, adduction of the eye had almost fully recovered, and there was no proptosis or displacement. After 6 months he had equal vision in
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both eyes (6/5) and diplopia only in extreme laevoversion, there being a 20% reduction in adduction and abduction. After 18 months adduction had totally recovered, but there was an approximate 10% reduction in abduction in the left eye. There is no sign of recurrence (Figs. 5, 6).

**PATHOLOGY**

The surgical specimen measured 27×15×17 mm and was hard, pale grey, and encapsulated. Light microscopy showed long strap-like cells, which tended to form a syncytium, within a connective tissue stroma (Fig. 7). The nuclei were small, and mitotic figures were not seen. Frozen sections stained for lipid showed a markedly granular staining pattern (Fig. 8). Histochemical analysis of the granules showed them to consist of glycoprotein, with some bound lipid, indicating a probable lysosomal origin.

Electron microscopy showed prominent basal laminae, organelles packed with granules, some myelin figures, and areas of degenerate mitochondria; long-spaced collagen was also present (Figs. 9, 10).

**Discussion**

Abrikossoff's original assertion that the tumour arises from striated muscle cells has been challenged. Evidence has been put forward for a histiocytic origin either as a hyperplasia, a granulomatous response to parasites, or as a storage disorder. Others have drawn attention to similarities between granular cell...
myoblastoma and neural tissue, suggesting an affinity with neurofibroma,\textsuperscript{22,23} perineural fibroblastoma,\textsuperscript{24} or Schwann cells.\textsuperscript{25,26} More recently, owing to the great similarity between the ultrastructure of schwannomas and granular cell myoblastoma, it has been postulated that both may arise from a pluripotential mesenchymal cell. Sobel et al.\textsuperscript{27} put forward a schema to show the evolution of fibroblasts, smooth muscle cells, and granular histiocytes at sites of muscle trauma from such cells. This evidence, coupled with the positive choroiembryonic antigen antisera test, suggests that there is a strong case for assigning the genesis of both schwannomas and granular cell myoblastoma to a primitive pluripotential cell line. Furthermore, the finding that granular cell myoblastoma is similar to recognised tumours of Schwann cells and melanocytes in containing S-100 protein\textsuperscript{28} is putative evidence of a neural crest origin. The location of the tumour in the present case within the medial rectus muscle could be taken to support a myoblastic origin, but in the absence of confirmatory evidence from histochemical and ultrastructural study this conclusion is not really warranted.

A presentation of visual obscuration caused by the

\[\text{Fig. 8 Frozen sections reveal this lipid content of the granules within the tumour cell cytoplasm. (Oil red O, } x270)\]

\[\text{Fig. 9 The cytoplasmic granules are membrane-bound and contain fine amorphous material of variable density. (EM, } x10000)\]
mechanical effects of the tumour in different positions of gaze is unusual, though we have seen this with 2 other patients at Moorfields who have had optic nerve meningiomas. In this case as in the others it could be shown that the visual symptoms were due to the reduction of blood flow in the retinal circulation on holding the eye in the abducted position, the mechanical action of the tumour in this position causing compression of the optic nerve and its vascular supply. After the operation there were no signs of fluctuating circulation with the eye position.

The almost total lack of postoperative problems in this patient was due to a wide exposure by means of a lateral orbitotomy approach and a careful microsurgical dissection carried out under hypotensive anaesthesia. We can foresee no other approach which could give such a good result considering the difficult anatomical site of the lesion.

Previously this tumour has been reported fixed to the extraocular muscles\(^\text{15}\) and to the optic nerve,\(^\text{1}\) but, as far as we are aware, it is unique for this neoplasm to be contained within an extraocular muscle.

According to the literature this tumour is usually benign, and only 2 reports of malignancy in the orbit have been published.\(^\text{2,16}\) One of these\(^\text{16}\) was a metastasis from a primary granular cell myoblastoma of the ovary which metastasised to the eyelid. The other, a primary orbital tumour,\(^\text{2}\) recurred after exenteration and spread regionally and metastatically over a 2-year period. At present our reported case has been followed up for 18 months with no signs of recurrence.

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References


