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Granular Cell Tumor of The Inferior Rectus Muscle

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A 53-year-old woman complaining of vertical diplopia, presented with a localized swelling in the right lower lid. Magnetic resonance imaging studies demonstrated a relatively well-defined mass in the inferior rectus with similar signal characteristics to the muscle. Excisional biopsy of the mass revealed granular cell tumor composed of S-100 positive cells with acidophilic granular cytoplasm and a peripheral lymphocytic infiltration. Granular cell tumor, which is very rare in the orbit, should be considered in the differential diagnosis of tumors adjacent to or within the extraocular muscles, particularly in the inferior orbit.

Key Words: Extraocular muscle, Granular cell tumor, Inferior rectus, Magnetic resonance imaging, Orbit

Granular cell tumor, presumably of neural crest origin, may arise in any part of the body, most notably in the tongue, chest wall and upper extremities.[1,2] Less frequent sites are the larynx, gastrointestinal tract, breast, the pituitary stalk and the female anogenital region.[2] The name "granular" derives from the acidophilic granular appearing cytoplasm packed with lysosomes.[3] The tumor is diagnosed only seldom in the eye and in its adnexa and is usually seen between the third and sixth decades of life.[4,5] There appears to be a female preponderance.[4] Malignant forms are exceedingly rare and the tumor may be multicentric in 10-15% of cases.

We herein describe a woman with a granular cell tumor in the inferior orbit, initially presenting with vertical diplopia. Imaging features of this rare tumor are also emphasized.

Case Report

A 53-year-old woman presented with long-standing mild swelling of the right lower eyelid and a recent-onset diplopia. Her visual acuity was 20/20 in each eye. There was no axial proptosis but the right eye was slightly displaced upwards. On palpation, the mass was immobile, hard and painless. There was 2+ restriction of movement of the right eye all through the upper gaze positions accompanied by diplopia.

Magnetic resonance imaging (MRI) studies of the orbit showed a relatively well-circumscribed mass in close proximity to the right inferior rectus muscle (Fig. 1A). On T1-weighted images, the tumor was hypointense compared to orbital fat but slightly hyperintense with respect to the extraocular muscles. The mass only minimally enhanced homogeneously following gadolinium injection. On T2-

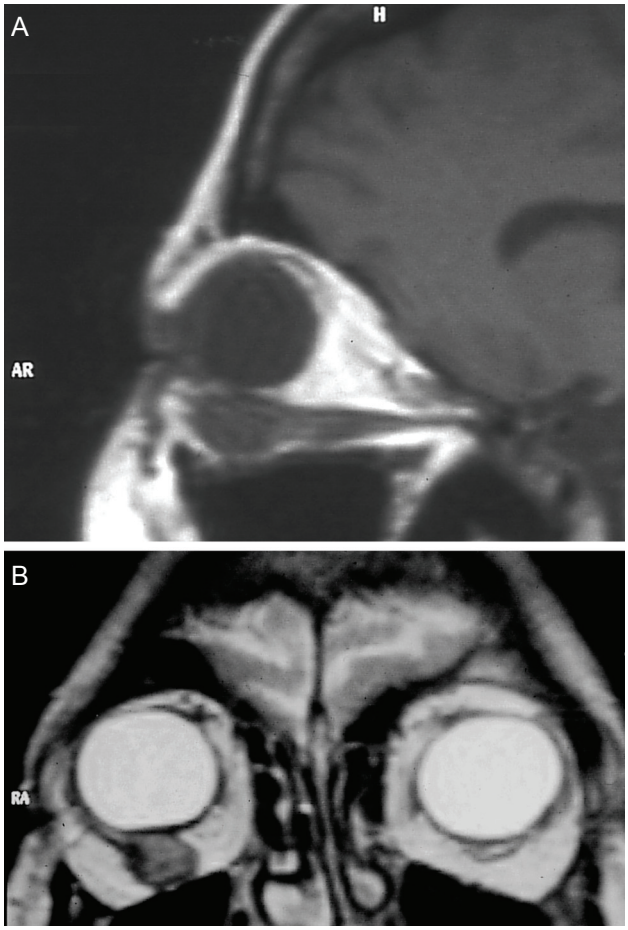

Fig. 1. (A) Sagittal T1-weighted MRI view of the right orbit showing the inferiorly located mass. (B) In this coronal T2-weighted MRI frame, distinction between the tumor and the inferior rectus muscle cannot be made.

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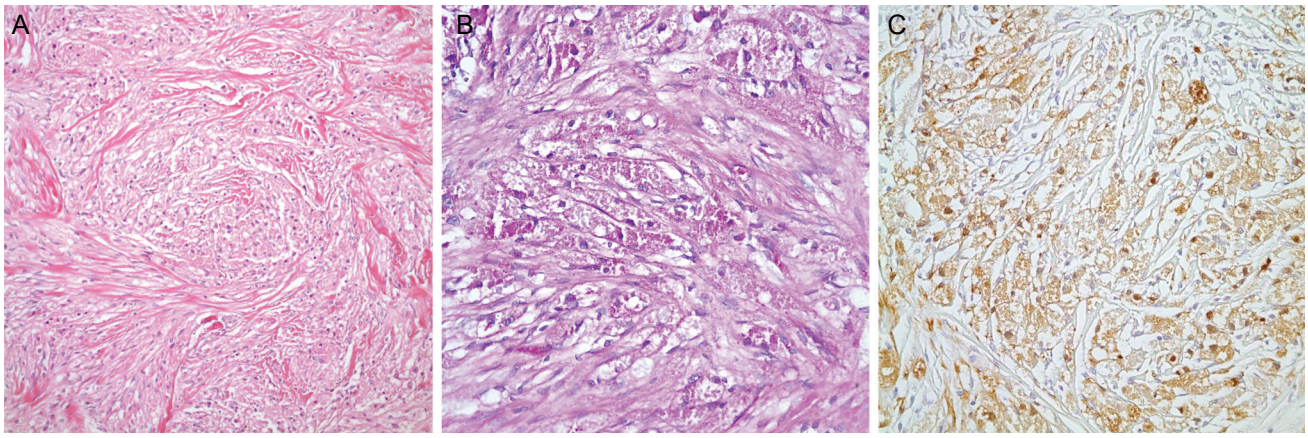

Fig. 2. (A) The tumor consists of nests of polygonal cells with prominent round to oval nuclei in a collagenized stroma with a fascicular development pattern (H&E, original magnification ×60). (B) Positive staining with periodic acid-Schiff reagent (original magnification ×100). (C) Tumor cells stain strongly positive for S-100 protein (S-100, original magnification ×100).

weighted sequences, the tumor was hyperintense relative to the orbital fat (Fig. 1B).

The tumor was approached via inferior cul-de-sac orbitotomy. Unexpectedly tight adhesions to the inferior rectus muscle and adjacent tissues were present. The tumor was stripped from the muscle in its entirety with sharp dissection. There was no recurrence during 4 years of follow-up. However, her vertical diplopia persisted and fresnel prisms were prescribed to alleviate her symptoms during the first year postoperatively.

Histopathological Findings

On gross examination, the tumor was well-demarcated and unencapsulated. The lesion was tan to gray in color and measured 2.3×1.5×1.5 cm. The tumor was composed of closely packed polygonal cells that had conspicuous, abundant granular cytoplasms (Fig. 2A). Several sections of peripheral nerve bundles were encountered within the tumor. There was mononuclear cellular infiltration at the periphery of the tumor. No muscular elements were seen. Staining with the periodic acid-Schiff reagent was strongly positive (Fig. 2B) and this was resistant to diastase digestion. Immunohistochemical studies showed significant positivity for S-100 protein (Fig. 2C) (S-100, Zymed, 1:50) but negative result for myoglobin.

Discussion

Not long ago, granular cell tumor or myoblastoma was classified as a mysterious tumor and interpretation with aproper diagnosis was regarded as a matter of opinion.[6] Since then, many more sporadic cases were recognized in the orbicularis muscle, lacrimal sac, lids, caruncle, conjunctiva and the ciliary body.[5,7] Orbital involvement continues to be rare however. When present, anteriorly located tumors tend to occupy the inferior orbit whereas more posterior tumors usually have an inferotemporal location.[4] While most of the granular cell tumors are discrete and well-delineated from the surrounding tissues, some may be infiltrative, moulded to the globe, invade the sclera or surround the optic nerve sheath.[1,2,8] A striking feature of the tumor is the involvement of extraocular muscles in 43% of cases.[3] Associations with the medial rectus,[8,9] inferior oblique,[8,10] inferior rectus[3,11-14] and the lateral rectus[2] muscles were clearly documented. Diplopia was almost invariably the leading symptom in these patients.

According to a recent report describing the MRI features, granular cell tumor appeared homogeneously isointense with respect to extraocular muscles on T1-weighted images and hypointense relative to the orbital fat on T2-weighted frames, enhancing slightly with gadolinium-DTPA.[10] The boundary between the tumor and the inferior oblique muscle could not be determined by these authors. We had similar observations on MRI on which the tumor appeared to arise from within the muscle whereas it was found adhered to the muscle during surgery.

Another interesting but perhaps underreported feature of granular cell tumor is the presence of lymphocytes at the tumor periphery. Apart from our case, this peculiar finding was also observed in two other patients.[2,15] The significance of peripheral lymphocytic accumulation is not clear.

Our experience with this patient tends to suggest that a solitary and well-delineated mass adjacent to or within an extraocular muscle and inferior rectus in particular, with the complaint of diplopia should remind the possibility of granular cell tumor in the differential diagnosis.

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