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MR Appearance of Intraorbital Granular Cell Tumor: A Case Report

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MR imaging of the orbit has proved useful for diagnosing a number of lesions, including neurinomas, meningiomas, gliomas, cavernous hemangiomas, pseudotumors, and lymphomas [1-5]. We describe the MR appearance of a rare intraorbital granular cell tumor in a patient with diplopia and exophthalmos, and discuss its characteristic MR features.

Case Report

A 32-year-old woman was admitted to our hospital because of diplopia of 17 months duration and proptosis of 7 months duration. Neuroophthalmologic examination on admission showed a slight decrease in visual acuity (18/20) as well as mild restriction of supraduction and exophthalmos. There was neither family history of neurofibromatosis nor contributory medical history. Thyroid functions were within normal limits. Plain skull films revealed no abnormalities. CT scans demonstrated a well-defined, ovoid, retrobulbar mass in the left orbit, which was slightly high in density compared with temporal muscle (Fig. 1A) and diffusely enhanced with administration of IV contrast medium (Fig. 1B). On MR images, the intraorbital tumor had an isointense signal relative to gray matter on T1-weighted studies (500/22) (Fig. 1C) and low signal on T2-weighted studies (2000/120) (Fig. 1D). MR imaging was performed with a 1.5-T superconducting magnet. With IV administration of gadopentetate dimeglumine, the tumor demonstrated diffusely speckled enhancement (Fig. 1E).

At surgery, a whitish, firm, and well-circumscribed tumor adhered tightly to the superior oblique and the superior palpebral levator muscles as well as to the nasociliary nerve. The optic nerve was displaced laterally.

Histopathologic examination disclosed a granular cell tumor showing closely packed polygonal tumor cells, which were diffusely eosinophilic and granular (Fig. 1F). S100 protein (Fig. 1G), myelin-associated glycoprotein Leu 7, and neuron-specific enolase were all immunohistochemically positive in tumor cells. The postoperative course after total removal of the tumor was uneventful. Left visual acuity as well as diplopia and proptosis were normal 4 months after the operation.

Discussion

Granular cell tumors, first described as granular cell myoblastomas by Abrikossoff [6], are benign, nonencapsulated growths that occur in a variety of organs [7], but only 10 cases have been reported in the orbit [8-17]. Abrikossoff's opinion that granular cell tumors arise from striated muscle is not widely accepted today, and their histogenesis has been a matter of controversy. It has been suggested that these tumors are derived from Schwann cells [18-21], perineural fibroblasts [22], primitive mesenchymal cells [23], or histiocytes [24]. The intraorbital granular cell tumors reported have been attached to extraocular muscles of the optic nerve [8, 11, 14-16]. The present intraorbital granular cell tumor demonstrated a tight attachment to a nasociliary nerve in addition to extraocular muscles at surgery. In addition, the presence of S100 protein, Leu 7, and neuron-specific enolase in tumor cells was shown immunohistochemically. These immunohistochemical findings in granular cell tumors support their neural, possibly Schwann cell, derivation [19-21, 25].

The present intraorbital granular cell tumor was of slightly high intensity relative to temporal muscle, and enhanced diffusely on CT scans. The signal was isointense with gray matter on T1-weighted MR images and of low signal intensity on T2-weighted images. Cone et al. [26] reported two cases of granular cell tumors in the neurohypophysis. These showed high density on plain CT scans and intense homogeneous enhancement with the addition of IV contrast material. In addition, MR imaging in one case demonstrated a suprasellar mass isointense with gray matter on T1-, proton-density, and T2-weighted images.

Intraconal orbital lesions previously reported to have isointense signal relative to gray matter on T1-weighted images include meningiomas, gliomas, cavernous hemangiomas, pseudotumors, and lymphomas [1, 4, 5]. The former three tumors, however, usually show isointense or high signal on T2-weighted images [4, 5], while the latter two tumors may demonstrate signal similar to fat on T2-weighted images [1]. Intraorbital neurinomas usually exhibit low signal on T1-weighted images and high signal on T2-weighted images [3]. The MR appearance of our intraorbital granular cell tumor is different from most other intraorbital tumors because of its low T2 signal intensity. This should be taken into consideration in the differentiation of intraorbital lesions.

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Fig. 1.—32-year-old woman with diplopia and proptosis.
A and B, Axial CT scans show ovoid intraorbital mass in medial half of left retrobulbar space. Tumor is well-defined, showing slight hyperdensity on plain CT scan (A) and diffuse enhancement after IV contrast injection (B).
C–E, On MR images the signal is similar on both T1-weighted (500/22) (C) and T2-weighted (2000/120) (D) studies. The signal enhances in a speckled manner with IV contrast administration (E).
F and G, Histologic specimens show polygonal tumor cells arranged close together and diffuse eosinophilic and granular cytoplasm (F). The cytoplasm and nuclei are immunohistochemically positive for S100 protein (G). (H and E, original magnification ×200)

REFERENCES