Granular cell tumor of the sellar region

AKA (infundibular) granular cell tumor (GCT). WHO grade I. Obsolete terms: choristoma, [1] granular cell myoblastoma, pituicytoma (this term is now reserved for a circumscribed glial neoplasm). A circumscribed tumor with nests of large cells has granular, eosinophilic cytoplasm due to copious lysosomes in the cytoplasm.

Epidemiology

While rare, GCTs are the most common primary tumor of the neurohypophysis and pituitary stalk/infundibulum [2] with a predilection for the stalk (these result in suprasellar extension). GCTs have been identified in the gastrointestinal tract, genitourinary tract, the orbital region as well as in other locations of the central nervous system with no connection to the pituitary gland or hypothalamus (e.g. spinal meninges [3]). Female: male ratio ≥ 2:1.

Pathology

Asymptomatic microscopic clusters of granular cells (tumorettes) are more common, with an incidence of up to 17% [4].

Clinical

They usually follow a slow progression with benign behavior. The most common presentation is with visual field deficits due to optic chiasm compression [5]. However, any symptom typical of a hormonally inactive sellar mass may occur.

Diagnosis

Imaging: may appear radiographically identical to adenomas. Rarely considered in the differential diagnosis pre-op. Isodense on CT and isointense on T1WI MRI, dense homogeneous enhancement on CT & MRI.

Treatment

If GTC is suspected pre-op, a transcranial approach is preferred over transsphenoidal because of the vascularity which has prevented total resection in 60–70% of reported cases [6]. XRT may be considered for subtotal resection [7].

Surgical resection is the standard treatment, more recently with transsphenoidal surgery when indicated. Surgical resection results in optimal outcome for patients [8].

Case series

A retrospective study of 11 patients with granular cell tumors. Information obtained from patients' medical records was supplemented by direct physician and patient telephone contact.

RESULTS: The study group consisted of 9 women and 2 men, with a mean +/- SD age of 50 +/- 15 years (range, 26-73 years). Seven patients were symptomatic, 3 of whom presented with visual complaints. Four patients presented primarily with endocrine dysfunction (2) or headaches (2). The duration of symptoms varied from 1 to 12 months (mean +/- SD, 6.0 +/- 4.5 months). Formal visual field testing revealed bitemporal hemianopsia in 6 and a right-sided visual field deficit in 2. The tumor appeared as a well-defined, relatively homogeneously (6) or heterogeneously (1) enhancing suprasellar mass on contrast infusion. Tumor size varied from 1.5 to 6.0 cm (mean +/- SD, 3.1 +/- 1.6 cm). At surgery, the mass was noted to be firm and vascular, and in at least 3 patients these features prevented gross total resection. Only 1 patient who had undergone biopsy of his lesion received adjuvant radiation therapy. Follow-up was obtained in all but 1 patient. Nine of these 10 patients are alive and are either free of disease or have had no disease progression. Follow-up in these 9 patients varied from 1 to 16 years (mean +/- SD, 6.5 +/- 53 years).

CONCLUSIONS: A granular cell tumor is generally a surprise finding in that it is seldom considered in the differential diagnosis of contrast-enhancing, demarcated sellar region masses. Granular cell tumors are benign and slow growing with no pronounced tendency for invasion or recurrence. Because of the indolent growth of granular cell tumors, the surgical goal of tumor decompression is a reasonable therapeutic approach [9].

Case reports

Three patients had a diagnosis of GCT of the sellar region occurring over an 18-year period. All three patients were followed postoperatively at our multidisciplinary pituitary center (median follow-up = 30 months; range 12-30 months). Hormonal disturbances, an incidental lesion requiring diagnosis, and neurological symptoms were indications for surgery in these patients. Two patients underwent a craniotomy and one underwent endoscopic transsphenoidal surgery. All three patients were free of tumor recurrence at last follow-up. In one case tested, positive thyroid transcription factor-1 (TTF-1) immunohistochemistry was observed.

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A 70-year-old female presented with progressive vision impairment found to have bitemporal visual field defects. Subsequent magnetic resonance imaging (MRI) revealed a 2.9 cm × 2.5 cm × 2.5 cm parasellar mass with extension into the third ventricle and causing optic tract edema (OTE). Right frontotemporal orbital craniotomy was performed and the tumor was partially removed to decompress optic nerves. Pathology identified the tumor as granular tumor of the sellar region. The patient's vision improved minimally after the surgery. Follow-up MRI after 3 months and 11 months showed stable left OTE.

GCTs were thought to be benign tumors with slow growth, but they could potentially possess aggressive features and invade into surrounding structures as described in this case. OTE can be a rare MRI finding of GCTs. Only one case of GCT-related OTE has been reported in literature to our best knowledge [11].

A 36-year-old neurologically normal woman with known MEN-1 underwent a screening magnetic resonance imaging (MRI) scan which revealed a 10 mm x 6 mm x 7 mm sellar/suprasellar lesion. She underwent endoscopic endonasal transsphenoidal resection. The subsequent neuropathological analysis was consistent with GCT of the pituitary gland. This is the first report of a GCT of the pituitary gland occurring in a patient with MEN-1 [12].

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