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Case Report

Aggressive granular cell tumor of the neurohypophysis with optic tract edema and invasion into third ventricle

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ABSTRACT

Background: Granular cell tumors (GCTs) of the neurohypophysis are parasellar tumors arising from pituicytes in the neurohypophysis and are generally considered benign slow-growing tumors. We present a case of sellar GCT with aggressive features.

Case Description: 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.

Conclusion: 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.

Keywords: Granular cell tumor, Optic tract edema, Parasellar tumor, Third ventricle invasion

INTRODUCTION

Granular cell tumors (GCTs) of the neurohypophysis are benign tumors arising from pituicytes in the neurohypophysis. It was first described by Boyce and Beadles as an infundibular mass in 1893 and was identified as a tumor with granular cell histology by Sternberg 30 years later.[2] Most patients with GCTs are asymptomatic because tumors are usually not large enough to cause symptoms.[2] In contrast to asymptomatic GCTs, which are relatively common, symptomatic GCTs are very rare with <100 cases reported in literature.[5] Symptoms observed in patients with GCTs include visual impairment, headaches, dizziness, and manifestations of partial or complete hypopituitarism.[4] GCTs are typically considered benign and indolent tumors despite occasional invasion and recurrence.[4] Here, we describe a case of suprasellar GCT with optic nerve edema and invasion into the third ventricle, suggesting that this benign tumor could potentially possess invasive features.

CASE REPORT

A 70-year-old female with progressive vision impairment and headaches of at least 6 months duration presented to her ophthalmologist for eyeglasses replacement. She was found to have bitemporal visual field defects, left worse than right. She had no relevant medical history and physical exam was unremarkable except impaired vision. The patient had no significant symptoms of hypopituitarism and pituitary function was not evaluated by a neuroendocrinologist at this time.

Magnetic resonance imaging (MRI) of the brain with contrast revealed a 2.9 cm × 2.5 cm × 2.5 cm well-defined parasellar mass in the region of the hypothalamus and optic chiasm. The lesion was isointense on precontrast T1 and fluid-attenuated inversion recovery (FLAIR), hypointense on T2 [Figure 1a] with homogeneous enhancement [Figure 1b]. The mass was inseparable from the superior aspect of the optic chiasm which was displaced inferiorly. It extended superiorly into the inferior margin of the third ventricle [Figure 1c] and was closely abutting the superior aspect of the pituitary infundibulum. There was no evidence of cystic component or calcification. There was increased signal along bilateral optic tracts and visualized a portion of the optic chiasm particularly on the left side, suggesting optic tract edema (OTE) [Figure 1d].

Based on these findings, the initial differential diagnosis included hypothalamic chiasmatic glioma, choristoma and less likely lymphoma. The patient underwent right frontotemporal orbital craniotomy with intraoperative neurophysiologic monitoring and image-guided neuronavigation for partial resection of the tumor and decompression of the bilateral optic nerves. Intraoperatively, a large suprasellar mass extending over the optic chiasm was identified. The optic chiasm was almost inseparable from, hard to define from the tumor. Frozen section histology was consistent with possible GCT or less likely plasmacytoma. Given the pathologic diagnosis, debulking of the tumor was completed in order to decompress the optic nerves.

The formal pathology report identified the tumor as GCT of the sellar region, World Health Organization (WHO) Grade I. The tumor consisted of epithelioid to vaguely spindled cells with round to ovoid nuclei and abundant granular eosinophilic cytoplasm [Figure 2a and b]. The tumor cells displayed mild to moderate nuclear pleomorphism with occasional distinct to prominent nucleoli. No mitotic figures or necrosis was seen. The periodic acid-Schiff (PAS) stain largely highlighted the cytoplasmic granularity [Figure 2c]. The tumor showed diffuse positive nuclear staining for thyroid transcription factor-1 (TTF-1) [Figure 2d] and weak, patchy staining for S-100 and glial fibrillary acid protein. There was a lack of staining for cluster of differentiation 1a and synaptophysin. The Ki-67 labeling index was approximately 1% indicating a low proliferative index.

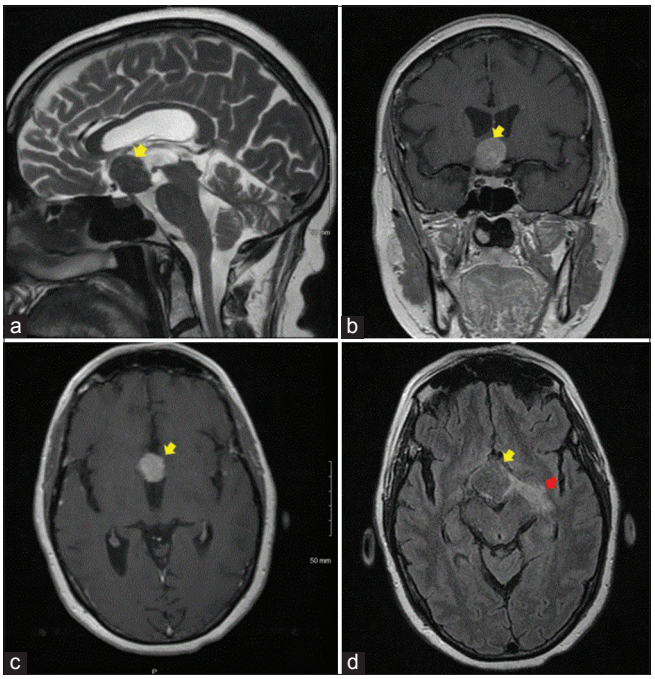

Figure 1: MRI findings of the granular cell tumor (yellow arrow). The lesion was hypointense on T2 image (a). It was enhanced homogeneously (b). The lesion invaded into the third ventricle (c) and caused optic tract edema (d, red arrow).

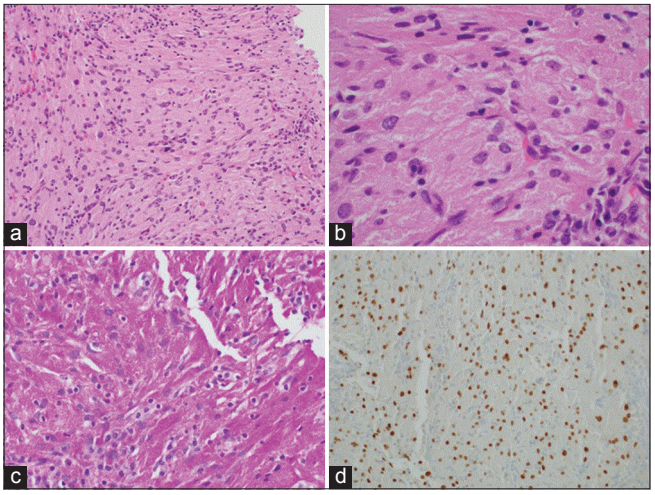

Figure 2: Pathology findings. H & E stain demonstrated epithelioid to vaguely spindled cells with round to ovoid nuclei and granular eosinophilic cytoplasm (a, b). PAS stain highlighted the cytoplasmic granularity (c). Positive nuclear staining for thyroid transcription factor-1 (d).

Immediate postoperative MRI showed a 2.6 cm × 2.4 cm × 2.4 cm anteriorly resected mass in close proximity to the optic chiasm. The patient was discharged home on

postoperative day 3 with no new neurologic deficits. Her vision improved minimally after the surgery. MRI 3 months and 11 months after the surgery showed that the residual mass was stable in size. There was persistent abnormal T2-FLAIR signal hyperintensity in the left optic radiation consistent with persistent edema along the left optic tract. She did not develop diabetes insipidus after the surgery but she complained of excessive fatigue at 5 months follow-up. Evaluation of pituitary function at the time showed a low normal free thyroxine at 0.63 ng/dL (normal range: 0.61–1.76 ng/dL) and a low normal morning cortisol at 8.9 μg/dL (normal range: 6.7–22.6 μg/dL). Her insulin-like gorwth factor-1, thyroid-stimulating hormone, and adrenocorticotropic hormone (ACTH) was normal. She was placed on levothyroxine and hydrocortisone replacement due to the concern of central hypothyroidism and adrenal insufficiency. Hydrocortisone was discontinued after 3 weeks due to a normal ACTH stimulation test.

DISCUSSION

Symptomatic GCTs of the neurohypophysis are rare tumors of the sellar region. GCT of the neurohypophysis was classified as a benign tumor according to the 2016 WHO classification of the central nervous system tumors.[9] Asymptomatic GCTs of neurohypophysis could be relatively common. In one study, 9% of 100 pituitary autopsies were found to contain GCTs.[15]

Symptomatic GCTs are very rare with <100 cases reported in literature.[5] Cohen-Gadol *et al*.[4] reviewed 50 cases of symptomatic GCTs of the neurohypophysis reported in literature and found out 90% patients had visual impairment or visual field defects due to compression of the optic chiasm. Other common symptoms include headache, decreased libido, infertility, panhypopituitarism, and amenorrhea.

Covington *et al*.[5] and Han *et al*.[6] summarized imaging features of GCTs of neurohypophysis. Although nonspecific, GCTs of neurohypophysis tend to be well-circumscribed, hyperattenuated on computed tomography (CT), hypointense on T2-weighted MRI, and enhance homogeneously. Cystic lesions are rare and calcifications are extremely rare. Approximate 60% of GCTs were suprasellar and 40% were both intrasellar and suprasellar.[5,9] The purely intrasellar lesion was not reported. In our patient, CT and MRI images demonstrated features compatible with GCT.

In addition to typical MRI features of GCTs, increased signal along bilateral optic tracts and visualized portion of the optic chiasm consistent with OTE was observed in this patient. OTE was thought to be specific to craniopharyngiomas only, but it is currently considered to be caused by various parasellar and sellar lesions. The mechanism underlying the development of OTE is not totally clear though a few hypotheses were proposed. In patients with craniopharyngioma, leakage of cyst contents could potentially cause local inflammation and edema, whereas in patients with other types of sellar or parasellar lesions, local edema, and OTE were attributed to direct tumor invasion and retention of interstitial fluid in perivascular spaces caused by tumor blockage.[10] To the best of our knowledge, OTE was only reported in one case of GCT of neurohypophysis where the edema diminished 90 days after subtotal resection of the tumor.[13] Although our patient had slight improvement in her vision after the decompression surgery, follow-up MRI 3 months and 11 months after the surgery showed stable left OTE. The relationship between OTE and visual outcomes was assessed in a few studies, but results were inconsistent.[1,3,11] Using optical coherence tomography, a recent study evaluated retinal nerve fiber layer thickness in addition to visual acuity and visual field in patients with sellar and suprasellar lesions. The study demonstrated worse visual function and poor visual improvement postoperatively in patients who had OTE.[7]

The exact diagnosis of GCTs depends on pathological examination.[9] GCTs, spindle cell oncocytomas, and pituicytomas are thought to potentially arise from the same origin, pituicytes which are specialized glial cells of the neurohypophysis. All of the three tumors stain positive for TTF-1. The distinction of GCTs from spindle cell oncocytoma and pituicytoma is mostly based on morphologic features. In GCTs, the PAS stain largely highlights the cytoplasmic granularity, which does not really occur in pituicytomas or spindle cell oncocytomas.

GCTs are generally considered benign and slow-growing tumors with no pronounced tendency for invasion or recurrence.[4] Although rarely reported, GCTs could potentially possess aggressive features such as mitotic figures,[14] third ventricle invasion,[12] and cavernous sinus invasion.[8] Another uncommon feature of GCTs, as seen in our patient, is tumor extension into the third ventricle suggesting tumor aggressiveness.

Surgical resection remains the mainstay of treatment to reduce the compression of surrounding structures. However, gross total resection is often prohibited by the firm and vascular nature of the GCTs along with lack of obvious dissection plane between and tumor and normal brain.[4] Transsylvian, subfrontal, transsphenoidal, and transcallosal approaches have been described in literature to resect GCTs based on the location of the tumor. The availability of assertive intraoperative histologic diagnosis is paramount for real-time intraoperative decision-making for goals of surgical resection.

CONCLUSION

We describe a patient with symptomatic GCT of neurohypophysis with OTE and third ventricle invasion,

indicating that this benign tumor could potentially possess aggressive features.

Declaration of patient consent

Patient's consent not obtained as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Baggenstos M, Chew E, Butman JA, Oldfield EH, Lonser RR. Progressive peritumoral edema defining the optic fibers and resulting in reversible visual loss. J Neurosurg 2008;109:313-7.
2. Barrande G, Kujas M, Gancel A, Turpin G, Bruckert E, Kuhn JM, *et al.* Granular cell tumors. Rare tumors of the neurohypophysis. Presse Med 1995;24:1376-80.
3. Bussat A, Proisy M, Bruneau B, Bouzillé G, Chappé C, Riffaud L, *et al.* Edema of the optic tract in patients with tumors of the sellar region: Clinical and visual implications in the pediatric population. J Neurosurg Pediatr 2018;21:516-22.
4. Cohen-Gadol AA, Pichelmann MA, Link MJ, Scheithauer BW, Krecke KN, Young WF Jr., *et al.* Granular cell tumor of the sellar and suprasellar region: Clinicopathologic study of 11 cases and literature review. Mayo Clin Proc 2003;78:567-73.
5. Covington MF, Chin SS, Osborn AG. Pituicytoma, spindle cell oncocytoma, and granular cell tumor: Clarification and meta-analysis of the world literature since 1893. AJNR Am J Neuroradiol 2011;32:2067-72.
6. Han F, Gao L, Wang Y, Jin Y, Lv Y, Yao Z, *et al.* Clinical and imaging features of granular cell tumor of the neurohypophysis: A retrospective analysis. Medicine (Baltimore) 2018;97:e9745.
7. Ju DG, Jeon C, Kim KH, Park KA, Hong SD, Seoul HJ, *et al.* Clinical significance of tumor-related edema of optic tract affecting visual function in patients with sellar and suprasellar tumors. World Neurosurg 2019;[Epub ahead of print].
8. Liu HL, Huang BY, Zhang MS, Wang HR, Qu YM, Yu CJ, *et al.* Sellar and suprasellar granular cell tumor of neurohypophysis. Chin Med J (Engl) 2017;130:741-3.
9. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, *et al.* The 2016 world health organization classification of tumors of the central nervous system: A summary. Acta Neuropathol 2016;131:803-20.
10. Saeki N, Nagai Y, Matsuura I, Uchino Y, Kubota M, Murai H, *et al.* Histologic characteristics of normal perivascular spaces along the optic tract: New pathogenetic mechanism for edema in tumors in the pituitary region. AJNR Am J Neuroradiol 2004;25:1218-22.
11. Saeki N, Uchino Y, Murai H, Kubota M, Isobe K, Uno T, *et al.* MR imaging study of edema-like change along the optic tract in patients with pituitary region tumors. AJNR Am J Neuroradiol 2003;24:336-42.
12. Schaller B, Kirsch E, Tolnay M, Mindermann T. Symptomatic granular cell tumor of the pituitary gland: Case report and review of the literature. Neurosurgery 1998;42:166-70.
13. Shizukuishi T, Abe O, Haradome H, Fukushima T, Katayama Y, Sugitani M, *et al.* Granular cell tumor of the neurohypophysis with optic tract edema. Jpn J Radiol 2014;32:179-82.
14. Shuangshoti S, Chantra K, Navalitloha Y, Charoonwatanalaoha S, Shuangshoti S. Atypical granular cell tumor of the neurohypophysis: A case report with review of the literature. J Med Assoc Thai 1998;81:641-6.
15. Tomita T, Gates E. Pituitary adenomas and granular cell tumors. Incidence, cell type, and location of tumor in 100 pituitary glands at autopsy. Am J Clin Pathol 1999;111:817-25.

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