Suprasellar granular cell tumor: A rare case report

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ABSTRACT

Granular cell tumor (GCT) originating from the sellar and suprasellar regions, specifically from the neurohypophysis, is a rare neoplasm. Symptomatic GCT of the neurohypophysis is exceedingly rare, being <70 cases described so far. GCTs predominantly exhibit benign behavior, while the malignancy rate remains at 2%. Imaging is quite unspecific and diagnosis is difficult to establish preoperatively. Histopathology and immunohistochemistry serve as the definitive diagnostic approach for GCTs and help in distinguishing GCT from other pituitary tumors, including pituitary adenoma, pituicytoma, and spindle cell oncocytoma. These tumors are resistant to radiotherapy and chemotherapy, necessitating surgical resection as the primary treatment modality. Due to the potential absence of distinct tumor masses and local tissue infiltration by tumor cells, complete excision is crucial, with resection extent extending beyond areas of infiltration. Here, we present a rare case of GCT originating from the posterior pituitary in the suprasellar region. On immunohistochemistry, tumor cells expressed diffusely thyroid transcription factor-1, S-100 protein, and vimentin confirming the diagnosis of supra-sellar GCT. The reported case is noteworthy for the rarity of the clinicopathological entity.

Key words: *Granular cell tumor, Sellar suprasellar region, Pituitary, Thyroid transcription factor-1*

Pituicytoma, granular cell tumor (GCT) of the sellar region, and spindle cell oncocytoma form a distinct section in the 5th edition of the World Health Organization of Central Nervous System Tumors. These are low-grade, benign, and thyroid transcription factor-1 (TTF1)-positive neoplasms that arise from pituicytes of the posterior pituitary or infundibulum. Pre-operative diagnosis of GCT can be challenging due to its similarity to other sellar lesions such as adenoma, meningioma, chordoma, and teratoma [1]. Surgical intervention is the primary approach for treatment; however, achieving complete tumor excision is frequently hindered by factors such as tumor size, proximity to the optic chiasm, and high vascularity, leading to a significant recurrence rate [1]. In this report, we present a case of a 45-year-old woman with a GCT and provide a concise overview of the subject matter.

CASE REPORT

A 45-year-old female patient presented with headache decreased visual acuity and facial paralysis for 3 months. The symptoms gradually increased in severity and 1 week before, she presented with nausea, vomiting and decreased visual acuity which led her to a neurological evaluation. The patient had no significant past medical history and no family history of neoplasm or neurological disorder on investigation.

Radiological Investigation

Her magnetic resonance imaging (MRI) brain showed a well-defined lobulated homogeneously enhancing lesion appearing hypointense on T2Wt images in suprasellar region measuring about 15×14×12 mm and mildly bulging inferiorly into the sella turcica.

Histopathological Examination

We received a biopsy from suprasellar SOL. On gross examination, the biopsy showed multiple grayish fragments of tissue in aggregate measuring 1.3×1 cm, entirely submitted for histopathological examination.

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Microscopic Examination

Microscopic examination showed a tumor composed of densely packed polygonal cells with granular eosinophilic cytoplasm-forming sheets, nodules with whorls, and fascicular patterns. The tumor cell nuclei were mildly anisomorphic with few conspicuous nucleoli. Stroma shows some lymphoid cells and foci of hemorrhage and calcification. Significant mitosis or necrosis is not seen. Cytoplasmic granules in tumor cells were periodic acid–Schiff (PAS) positive and diastase resistant (Figure 1a-d).

On Immunohistochemistry

Tumor cells expressed diffusely TTF1, S100 protein, and vimentin and focally expressed glial fibrillary acidic protein. Tumor Ki67 proliferative index was low (1–2%). The tumor cells were immunonegative for PR, EMA, Olig 2, synaptophysin, and chromogranin (Figure 1e-h).

DISCUSSION

Reporting this case aims to enhance medical awareness and knowledge regarding this rare disease. GCTs were initially described in 1893 by Boyce and Beadles. They account for <0.5% of all sellar tumors, with <70 cases of symptomatic GCTs being reported in the literature thus far [2]. The majority of them are benign, with a low rate of malignant transformation estimated at 2%. Women are more commonly affected than men, with the peak incidence occurring in the fifth decade of life. Symptoms associated with GCTs include gradual onset of visual disturbances in up to 90% of cases, as well as headaches and endocrine changes such as amenorrhea, galactorrhea, infertility, impotence, and weight fluctuations in approximately 50% of cases. Hypopituitarism or hyperprolactinemia are frequently observed, although there have been rare cases of acromegaly. Radiological findings are not specific and may resemble other central nervous system lesions such as meningioma, pituitary adenoma, chordoma, or teratoma. Diagnosis of GCTs before surgery is challenging. In the present case, pre-operative diagnosis was pituicytoma or craniopharyngioma. The definitive diagnosis is typically established on histologic features through surgical excision [1].

Histopathological examination of GCTs reveals neoplastic cells that exhibit both spindle and polygonal shapes, with eosinophilic granular cytoplasm. These cells are diffusely positive for certain markers such as PAS, S100, CD68, vimentin, and TTF1 but negative for pituitary hormone expression. These characteristics differentiate GCTs from other oncocytic tumors of the sellar region, such as pituicytoma and spindle cell oncocytoma [3]. In addition, a comprehensive analysis of expression profiles in three sellar tumor types (spindle cell oncocytoma, pituicytoma, and GCT) revealed varying patterns of marker expression, providing valuable insights into their distinct characteristics. Furthermore, the discussion considers the ultrastructural variants of normal pituicytes and raises the possibility that certain regions might

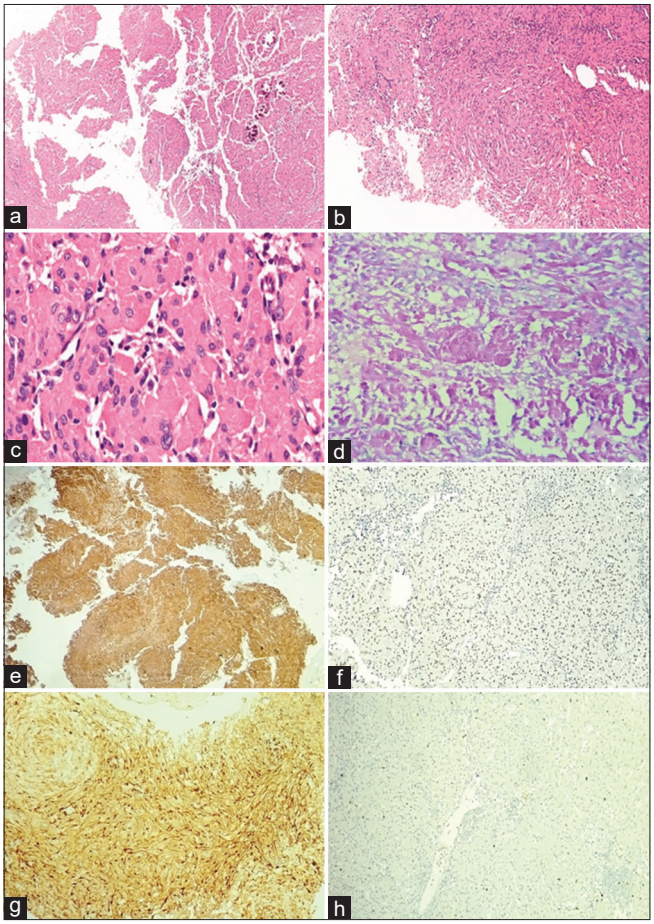

Figure 1: (a) H and E ×10–tumor composed of densely packed polygonal cells forming sheets and nodules, (b) H and E ×10–tumor composed of densely packed polygonal cells forming whorls, (c) H and E ×40–tumor cells showing granular eosinophilic cytoplasm, (d) PAS diastase–cytoplasmic granules in tumor cells are PAS positive and diastase resistant, (e) Vimentin: ×10–diffuse positive in tumor cell, (f) TTF1 ×10–diffuse nuclear expression in tumor cells, (g) S100 ×10–diffuse positive in tumor cells, (h) KI 67 proliferative index low (1–2%) in tumor cells. PAS: Periodic acid-Schiff, TTF1: Thyroid transcription factor-1

represent ependymal pituicytes or ependymal differentiation within an oncocytic pituicytoma. Literature also explores the impact of anatomic location on TTF-1 expression, suggesting that folliculostellate cells may share a common origin with pituicytes but exhibit differential TTF-1 expression based on their migration and developmental context. This multifaceted exploration contributes to the evolving understanding of sellar tumors and underscores the complexity of their histopathological features [4].

Treatment is surgical and should be based on clinical judgment and pituitary adenoma guidelines. Surgical removal can be performed through a transnasoseptal-sphenoidal approach (microscopic or endoscopic technique) or by pterional (frontotemporal) craniotomy depending on the tumor size and extension. These tumors are difficult to resect due to their tough and highly vascular structure and tendency to adhere to adjacent brain structures (optic pathway, hypothalamus, among others) [2]. Surgical outcomes are variable and usually only subtotal removal is possible if preservation of adherent normal brain structures is

desired. To achieve complete resection, sacrifice or damage of the pituitary stalk and other structures is frequent and carries a higher risk of endocrine sequelae such as diabetes insipidus and hypopituitarism. Radiotherapy as a sole or adjuvant treatment strategy is controversial and cannot be routinely recommended. Cases with anaplastic features, progressive clinical course, or inoperable recurrence may constitute indications for adjuvant radiotherapy. The prognosis is largely dependent on the surgical removal [5]. Metastasis has not been reported [6].

CONCLUSION

GCTs of sellar and suprasellar regions are rare neoplasms predominantly and exhibit benign behavior. Histopathology serves as the definitive diagnostic approach. These tumors are resistant to radiotherapy and chemotherapy, necessitating surgical resection as the primary treatment modality. Due to the potential absence of distinct tumor masses and local tissue infiltration by tumor cells, complete excision is the gold standard of treatment. Meticulous recognition of the clinical manifestations, MRI features, and comprehensive assessment of immunohistochemical analyses are imperative to achieve precise diagnosis and optimal treatment strategy for GCTs located in the sellar region.

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