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Granular Cell Tumor in the Pituitary Stalk: A Case Report

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Granular cell tumors (GCTs) have been reported in various tissues, especially the skin and subcutaneous soft tissue of the head and neck. We report a 60-year-old man who presented with intermittent headache and dizziness for 3 months, but no other neurological symptoms. Magnetic resonance imaging (MRI) showed the presence of a mass in the pituitary stalk, and contrast-enhanced MRI showed nodular enhancement in this region. The lesion was completely excised microscopically via a frontotemporal (pterional) approach. On pathological examination, a final diagnosis of a typical GCT was made.

Key Words Granular cell tumor; Pituitary neoplasms.

INTRODUCTION

Granular cell tumors (GCTs) can develop in different parts of the body, but usually occur in the skin or the subcutaneous soft tissue of the head and neck. GCTs of the neurohypophysis or pituitary stalk are very rare [1-8]. They account for less than 0.1% of all primary brain tumors, and approximately 1–1.5% of adult brain tumors [9]. In most cases reported to date, GCTs have been found in the posterior pituitary gland. GCT of the neurohypophysis is difficult to diagnose preoperatively, owing to the lack of specific imaging features [10]. In this paper, we report the clinical, radiological, anatomical, and pathological findings of a patient with GCT of the pituitary stalk, along with a literature review.

CASE REPORT

A 60-year-old man presented to our clinic complaining of intermittent headache and dizziness for 3 months. The patient had no specific neurological or hormonal symptoms. Magnetic resonance imaging (MRI) showed iso-signal intensity in the pituitary stalk on T1-weighted imaging (T1WI), and contrast-enhanced MRI showed nodular enhancement in this region (Fig. 1). However, these findings were not conclusive, and the differential diagnoses included metastasis, lymphoma, and glioma. A pituitary function test revealed high levels of thyroid-stimulating hormone (22.02 μlU/mL). Other laboratory findings for hormones and infection markers were normal. The patient was diagnosed with hypothyroidism, and he was preoperatively administered 50 μg of levothyroxine sodium. Surgery was performed for pathologic confirmation.

The tumor adjacent to the pituitary stalk was completely excised via a frontotemporal (pterional) approach. Thinning of the pituitary stalk had been caused by the tumor (Fig. 2). Macroscopically, the tumor was a light gray, round, mass-like lesion, 1×1 cm in size. It was relatively solid compared to other pituitary gland adenomas. Histopathologic examination revealed a fragment of brain parenchyma, with dense fibrocollagenous tissue admixed with granular cell nests, and multifocal lymphocytic infiltration. The tumor cells had abundant granular cytoplasm, showed diffuse weak positivity for CD68, and diffuse, weak to strong positivity for S-100 (Fig. 3). These results were conclusive for the diagnosis of a typical GCT.

Transient diabetes insipidus occurred immediately post-surgery. However, the patient recovered, without the use of hormone replacement therapy, within 1 month of surgery. In addition, postoperative MRI revealed an intact pituitary stalk (Fig. 4).

DISCUSSION

Anatomically, the neurohypophysis consists of the posterior pituitary gland, pituitary stalk, infundibulum, and median eminence. The cellular elements include pituicytes, microglias, and the distal parts of nerve cells from anastomosed blood vessels and the hypothalamus. Pituicytes are considered to be modified neuroglial cells, and show positive staining for glial fibrillary acidic protein; they have been classified into five different types on the basis of their ultrastructural characteristics: major, dark, ependymal, oncocytic, and granular [11]. Granular pituicytes contain many granums. GCTs, the most common primary tumors that develop in the pituitary gland, have similar granums, and some studies have suggested that these tumors originate from granular pituicytes [3,12,13]. Primary tumors that develop in the neurohypophysis are rare, and are known by many different terms such as pituicytoma, infundibuloma, granular cell myoblastoma, choristoma, and GCT [14]. Of these, GCTs, granular cell myoblastomas, and choristomas are synonymous, and are composed of polygonal cells with finely granular, eosinophilic, strong periodic acid Schiff-positive cytoplasm. The cells show little nuclear pleomorphism and no mitotic figures. Tumor cells are reactive for S-100 and CD68 on immunohistochemistry, as observed in the present case [15]. These

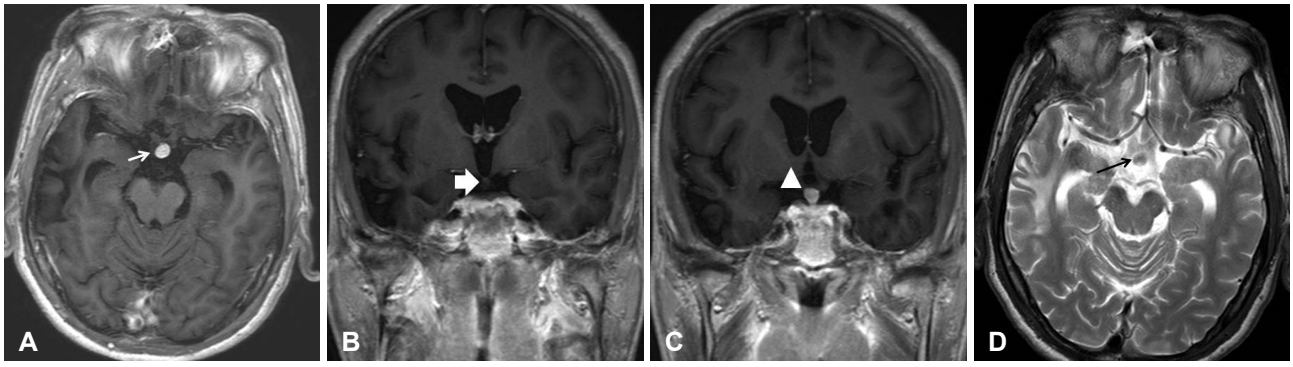

Fig. 1. Preoperative magnetic resonance imaging findings. A: A preoperative T1-weighted gadolinium-enhanced axial image shows a homogenous enhanced round mass (white arrow). B: A T1-weighted gadolinium-enhanced coronal image shows the pituitary stalk (white arrow). C: An anterior view of image B shows a round mass (white arrowhead). D: A T2-weighted axial image shows a mass (arrow) with low signal intensity.

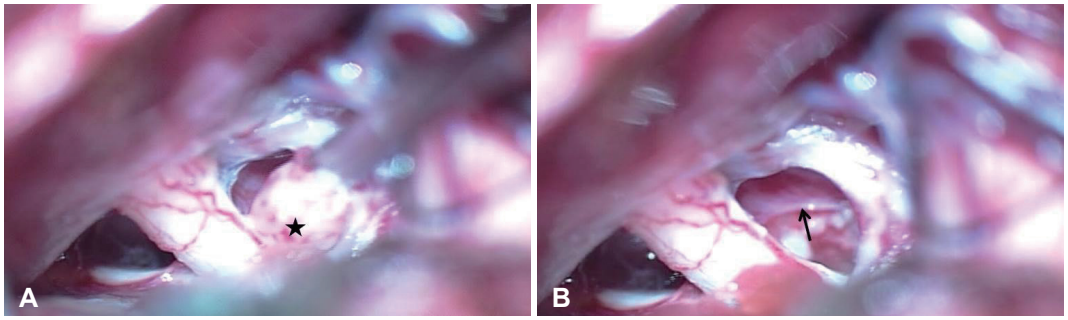

Fig. 2. Surgical findings. A: The tumor (asterisk) is round, and adjacent to the pituitary stalk. B: After tumor excision, the pituitary stalk (arrow) appears thinned, but remains intact.

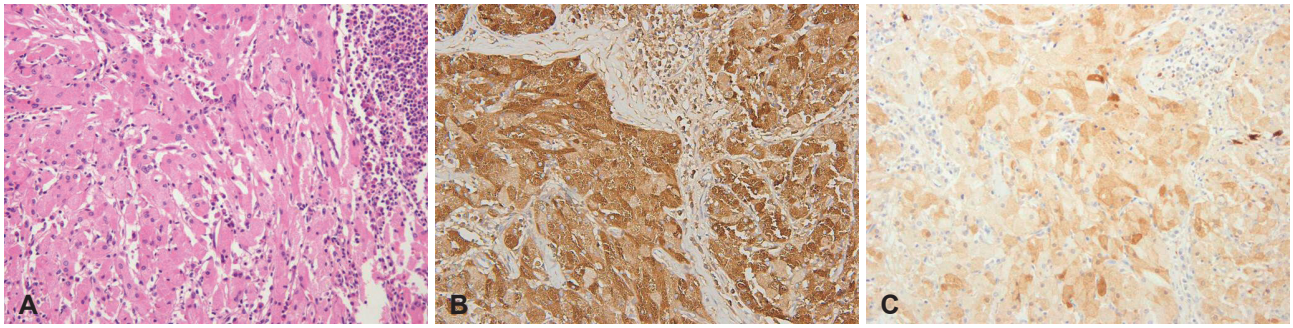

Fig. 3. Histopathologic findings. A: The tumor consists of large polygonal cells with ample granular cytoplasm and small, oval, eccentric nuclei (hematoxylin and eosin; original magnification ×100). B: The tumor shows dense fibrocollagenous tissue admixed with granular cell nests. Multifocal lymphocytic infiltration can also be observed (original magnification ×100). C: Immunostaining for S-100 shows diffuse weak to strong positivity (original magnification ×100).

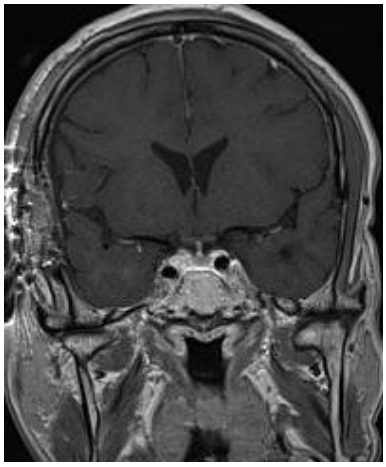

Fig. 4. Postoperative magnetic resonance imaging findings. A postoperative T1-weighted gadolinium-enhanced coronal image shows resection of the tumor adjacent to the pituitary stalk. The pituitary stalk remains intact.

tumors can be differentiated from pituitary adenomas by using endocrine markers such as chromogranin A, growth hormone, and adrenocorticotropic hormone [16].

A previous study reported that the number of cases of GCT involving the posterior pituitary gland was small, and 50 of these cases were symptomatic GCTs. Furthermore, all these cases were clinically benign.

GCTs of the neurohypophysis are difficult to diagnose preoperatively, owing to the lack of specific imaging features [10]. However, despite this, computed tomography (CT) and MRI are useful for visualizing tumor expansion to the sellar or suprasellar region. CT usually shows hyperdense tumors with contrast enhancement. MRI shows high signal intensity on T1WI for normal posterior pituitary gland, while gadolinium diethylenetri-aminepentaacetic acid-enhanced MRI shows iso-intense tumors with uneven contrast enhancement on T1WI, indicating fat components or neural hormones, as observed in this case. The radiologic findings in this case suggested the possibility of tumor, inflammatory pseudotumor, infection, or autoimmune disease. Infection, inflammatory pseudotumor, and autoimmune disease were excluded on the basis of the laboratory findings, and it remained only to distinguish between different tumor entities using pathological studies. As the imaging findings could not differentiate between metastases, lymphoma, and glioma, pathologic confirmation using a surgical specimen was necessary. It is rarely possibility to diagnose metastatic tumors on the basis of negative tumor marker results, and pathologic confirmation is indispensable for establishing treatment plans for lymphomas. Pathological examination to determine the degree of cell differentiation and malignancy is also necessary to distinguish between high- and low-grade gliomas.

The clinical features of GCTs in the posterior pituitary gland are mainly non-specific. The tumors are usually small, and have no space-occupying effects. Furthermore, they are usually asymptomatic; granular cell nests or pinhead-sized GCTs are observed in 6.4−17% of autopsy cases. Their frequency is similar to that found in the pituitary stalk and the posterior hypothalamus [1]. In rare cases, the tumors can be large, and the large size can result in headaches, visual defects, and endocrine problems such as hypocortisolism and acromegaly [2,17]. The current case was not associated with hormonal or ophthalmologic symptoms because of the small tumor size. Therefore, complete tumor resection was possible before the tumor became symptomatic and began to destroy the pituitary stalk.

Little is known about the natural progression of GCTs, and significant research using large cohorts of patients with GCT has not been performed. Most GCTs are benign, slow-growing tumors. However, they are occasionally associated with invasion or recurrence, and therefore, complete surgical resection is the treatment of choice. However, because of the benign, indolent nature of the tumor, partial resection is recommended when there is risk to major blood vessels or vital structures. In the present case, the tumor was located in the pituitary stalk, and in order to minimize stalk damage, a biopsy was performed for the differential diagnosis. Biopsy findings revealed a solid tumor, and safe surgical resection was deemed possible. In cases where stalk injury is likely, further resection would not be performed, and the tumor would remain untreated, as radiation therapy is not effective for GCTs.

Here, we report a rare case of GCT of the pituitary stalk, which is difficult to diagnose without pathologic confirmation. The GCT was completely resected, and the patient recovered fully from transient post-operative diabetes insipidus. Long-term follow-up is necessary in cases such as this, as the natural history of GCTs is poorly understood.

Conflicts of Interest

The authors have no financial conflicts of interest.

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