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Case Reports & Case Series

Clinical features and endoscopic findings of pituicytoma in the sellar region: A case report and review of the literature

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ABSTRACT

We report a rare case of pituicytoma occurring in the sellar region resected by endoscopic transsphenoidal surgery (ETSS). We suggest useful indicators for the accurate diagnosis and pitfalls for the surgical procedure. A 38-year-old man was admitted to our hospital with polyuria, erectile dysfunction and bitemporal hemianopsia. Neuroimaging revealed pituitary tumor in the sellar region, and the enhanced anterior pituitary gland was displaced anteriorly. Gross total resection was achieved using ETSS. Histological findings revealed bipolar spindle cells staining strongly for thyroid transcription factor 1 (TTF-1). We diagnosed the tumor as pituicytoma originated from posterior pituitary. Pituicytoma is difficult to diagnose on routine neuroimaging and pathological analysis, so accurate diagnosis requires identification of the forward deviation of anterior pituitary gland and analysis of immunohistochemical studies using TTF-1.

1. Introduction

Pituicytoma is a rare neoplasm originating from the neurohypophysis or pituitary stalk. Histologically, this tumor arises from pituicytes, which are specialized glial cells in the neurohypophysis and infundibulum, and the tumor most commonly occurs in the sellar and suprasellar regions [4]. It is difficult to differentiate pituicytoma from other pituitary tumors, because of the lack of specific radiological findings and the low incidence [1–4]. However, this tumor is reported to be much more vascular and attached to vital anatomical structures than other suprasellar tumors, including pituitary macroadenoma, meningioma and so on. The property of pituicytoma makes it difficult to achieve safety and complete resection of this tumor [1–3]. Therefore, it is very important to recognize the characteristic features of pituicytoma including neuroimaging, pathological findings and the risks of surgical procedures. Here, we report a rare case of pituicytoma occurring in the sellar region resected by endoscopic transsphenoidal surgery (ETSS). We suggest useful indicators for the accurate diagnosis and pitfalls for the surgical procedure.

2. Case report

A 38-year-old man presented to our department with a 3-month history of polyuria, erectile dysfunction and visual deterioration. Intracranial computed tomography (CT) revealed an iso-dense mass without calcification in the sellar region, accompanied by expansion of the sellar turcica (Fig. 1A). Magnetic resonance imaging (MRI) demonstrated that the tumor was isointense on T1-weighted imaging (T1WI) and T2WI, and homogeneously enhanced to a high degree with gadolinium (Gd). The tumor was extending into the suprasellar region, compressing the optic chiasm. The Gd enhanced anterior pituitary gland was displaced anteriorly, and high intense on T1WI which suggested posterior pituitary was completely disappeared (Fig. 1B, C). Examination for hormones related to the anterior pituitary showed hypogonadism (testosterone: 0.103 ng/ml). In terms of posterior pituitary function, arginine vasopressin level was severely disturbed (0.6 pg/

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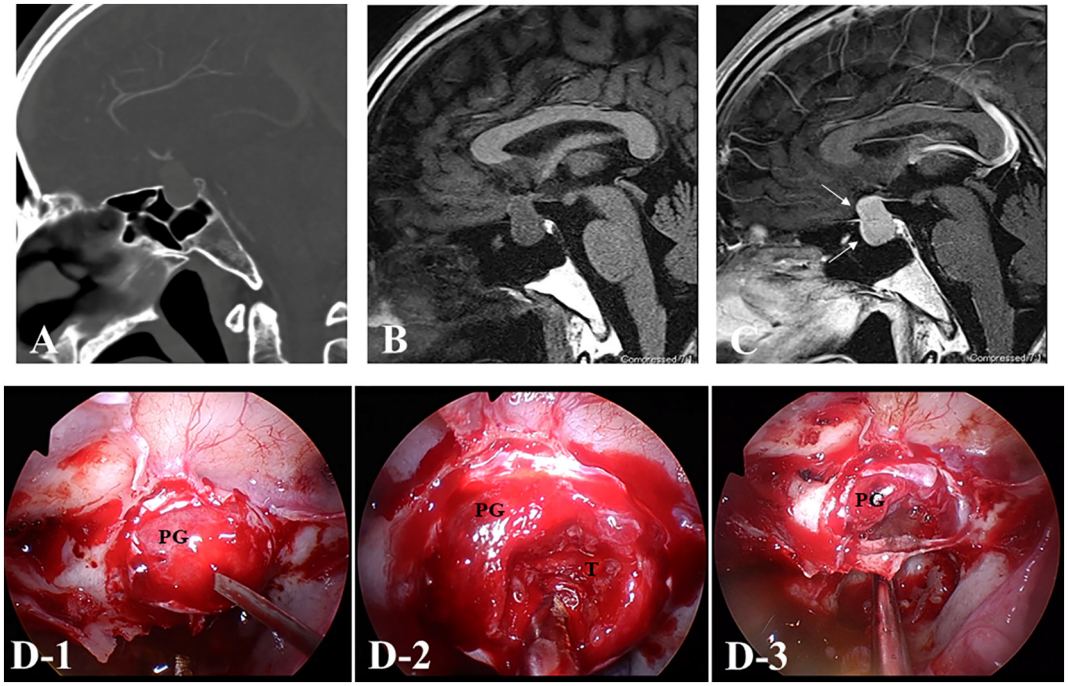

Fig. 1. Preoperative sagittal (A) computed tomography (CT) shows an iso-dense mass in the sellar region accompanied by expansion of the sellar turcica. T1-weighted (B: sagittal view) and gadolinium (Gd)-enhanced T1-weighted (C: sagittal view) magnetic resonance imaging (MRI) reveals a tumor mass in the sellar region extending into the suprasellar region. The tumor is homogeneously enhanced to a high degree with Gd. Enhanced anterior pituitary gland is displaced anteriorly (white arrows), and hyperintense on T1WI which suggested posterior pituitary is disappeared. Intraoperative findings from endoscopic trans-sphenoidal surgery (PG: pituitary anterior gland; T: tumor). D-1) Endoscopic view during operation showing the normal anterior pituitary gland located immediately under the bottom of the sella turcica. D-2) The tumor placed below the anterior pituitary gland. Macroscopic examination showed a solid, rubbery-firm consistency and hypervascularity. D-3) The tumor originated from the posterior pituitary gland and had a clear margin between the anterior pituitary. We achieved gross total resection of the tumor.

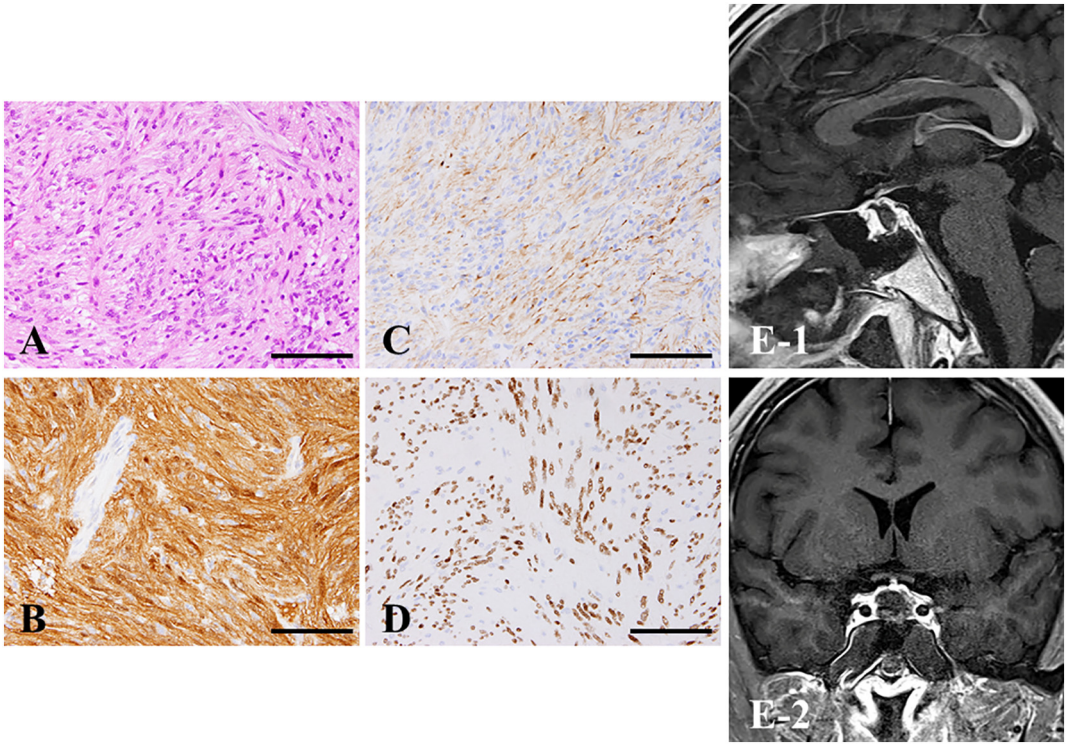

Fig. 2. Histopathology of the resected tumor shows round to spindle-shaped cells with fascicular or storiform growth patterns. Nuclei are round to oval in appearance without evidence of cellular atypia or mitotic figures (hematoxylin and eosin (HE) staining) (A). Most tumor cells are immunoreactive for S-100 protein (B) and GFAP (C). Almost all tumor cells are strongly positive for TTF-1 (D). Magnification, ×400. Scale bar, 100 μm. Postoperative (E-1) sagittal and (E-2) coronal images of Gd-enhanced MRI at 10 months after surgical resection shows no residual tumor in the sellar region.

ml; normal), reflecting diabetes insipidus. Preoperative differential diagnoses included pituitary adenoma, germ cell tumor, sellar meningioma and glioma arising from the posterior pituitary. To confirm the histological diagnosis, the tumor was resected by ETSS. Intraoperative findings showed the normal anterior pituitary gland was displaced anteriorly within the solid tumor. This tumor was firm, rubbery and highly vascular with massive bleeding. The tumor originated from the posterior pituitary gland and had a clear margin between the anterior pituitary. We achieved gross total resection to reduce compression of the optic nerve (Fig. 1D). Histopathology obtained from hematoxylin and eosin staining showed solid sheets of elongated, bipolar, spindle-shaped cells arranged in storiform pattern with a vague perivascular architecture. Most nuclei were round to oval with no evidence of cellular atypia or mitotic figures (Fig. 2A). Immunohistochemical studies were performed using antibodies for glial fibrillary acidic protein (GFAP), S-100, Ki-67 and thyroid transcription factor (TTF)-1. Tumor cells were immunoreactive for S-100 protein, GFAP (Fig. 2B, C) and the Ki-67 (MIB-1) proliferation-related labeling index was low, at 2.0%. In addition, almost all tumor cells appeared strongly positive for TTF-1 (Fig. 2D). Taking all these results into account, the final diagnosis was pituicytoma, belonging to tumors of the posterior pituitary in accordance with WHO classification of CNS tumors (2016) [3]. The postoperative course was uneventful, other than diabetes insipidus. MRI at 10 months after the initial treatment did not show any residual tumor (Fig. 2E) and neurological findings and hormonal dysfunction returned to near-normal.

3. Discussion

Pituicytomas are rare primary tumors originating from so-called pituicytes in the neurohypophysis and pituitary stalk. This pathology was considered a low-grade, spindle cell astrocytic tumor originating in the posterior pituitary or its stalk according to the 2007 WHO classification of CNS tumors. However, in the current 2016 WHO classification of CNS tumors, the term pituicytoma is restricted to a distinct group of low-grade glial tumors found in the posterior pituitary and infundibulum, presumably arising from pituicytes [4]. In the previous reports, pituicytomas occurred predominantly in adults, with a mean age of 46.9 years and a slight male predominance. The most common symptoms of pituicytoma are visual disturbance and headaches. Sometimes found on sexual dysfunction and diabetes insipidus. MRI generally depicts pituicytoma as isointense on T1WI, and iso or slightly hyperintense on T2WI. Strong enhancement has been seen after administration of contrast agents [1–3]. Another report described the enhanced anterior pituitary gland displaced anteriorly from the tumor mass, potentially reflecting the derivation of pituicytoma from the posterior pituitary lobe [4]. This forward deviation of anterior pituitary lobe is an extremely interesting and important finding and seems to be the key point to preoperative diagnosis of pituicytoma. In fact, our case presented with bitemporal hemianopsia similar to other suprasellar tumors. However, the patient showed biochemical evidence of endocrinopathy in the form of diabetes insipidus. This clinical symptom is associated with posterior pituitary dysfunction. In addition, MRI demonstrated homogeneous strong enhancement with Gd and the anterior pituitary gland was recognized in front of the tumor mass. Such clinical signs and imaging findings are very important to diagnose pituicytoma accurately before surgery, and we think that this recognition leads to the safety and effective operation for pituicytoma.

In the revised WHO classification of CNS tumors (2016), the definition of pituicytoma is as follows: “A circumscribed and generally solid low-grade glial neoplasm that originates in the neurohypophysis or infundibulum”. Microscopically, pituicytoma mainly consists of a solid, compact architecture almost entirely comprising elongated, bipolar, spindle-shaped cells arranged in a fascicular or storiform pattern. Cell borders are readily apparent and an infiltrative pattern is generally not seen. The nuclei are moderately sized and oval to elongate, with only mild irregularity of nuclear borders, and mitotic figures are rare [4]. With regards to immunohistochemical characteristics, pituicytomas do not demonstrate immunostaining for pituitary hormones or neuroendocrine markers. In general, pituicytes are glial cells that support the large axons of vasopressin- and oxytocin-producing hypothalamic neurons, and the cells are considered to be modified neuroglial cells with positive immunohistochemical staining for GFAP. Moreover, pituicytomas were found to strongly express the nuclear transcription factor TTF-1, and this factor is strongly expressed in fetal and adult human pituicytes [3,4]. Lee et al. reported that TTF-1 is specifically expressed in pituicytomas and provides a useful marker for distinguishing pituicytoma from other sellar tumors [5]. In our case, the tumor was composed of solid sheets of elongated, bipolar, spindle-shaped cells that were typically arranged in short fascicles and a storiform pattern microscopically. This structure was immunopositive for GFAP and S-100, and almost all tumor cells were strongly immunopositive for TTF-1. These findings are consistent with pituicytoma, in consideration of morphological studies and immunohistochemical analysis based on the WHO classification in 2016. We think that TTF-1 is the most useful marker to diagnose pituicytoma accurately.

Regarding treatment of pituicytoma, surgical resection is the most important part of treatment. Recently, endoscopic trans-sphenoidal surgery is becoming a common procedure from viewpoint of an effective and less-invasiveness for pituicytoma located in the sellar region [1–3]. On the other hand, these tumors are reported to be much more vascular and attached to normal anatomical structures such as the infundibulum or posterior pituitary lobe than other suprasellar tumors, including pituitary adenoma and meningioma. The massive bleeding resulting from tumor hypervascularity and strict attachment to normal structures are probably the main reason for the difficulty of achieving complete resection [1–3]. In our case, we were able to achieve gross total resection without being surprised at the strict attachment and massive bleeding, resulting in excellent tumor control after 10 months of follow-up without neurological complications. We are convinced that taking into account the possibility of pituicytoma before surgery resulted in safety and total removal. Therefore, it is very important to recognize the detailed character of pituicytoma. Further experience and longer patient follow-up is required.

4. Conclusion

We suggest that pituicytoma should be included as a differential diagnosis for pituitary tumors because of the potential for significant intraoperative bleeding and strict attachment to normal structures, unlike common pituitary neoplasms. So, careful identification of clinical signs, MRI findings and detailed evaluation of immunohistochemical studies are necessary for accurate diagnosis and appropriate treatment selection for pituicytoma.

Conflicts of interest

None of the authors have any commercial or financial involvement in connection with this study that represents or appears to represent any conflicts of interest.

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Ethical approval

The clinical study of the above-mentioned case report was approved by the Ethics Committee for Clinical Research of Ehime University Hospital, and informed consent was obtained from the patient prior to

initiating the study.

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