



Case report

Pituicytoma: A rare case report of sellar and suprasellar tumor

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ABSTRACT

Introduction: Pituicytomas are low-grade glial tumors in the sellar and suprasellar region. They may be easily confused with pituitary lesions. We review the literature in order to better understand and categorize the natural history, clinical presentations, and treatments.

Presentation of case: A 45-year-old female patient who complained of left eye blurred vision for 2 months. The imaging study revealed a solid sellar tumor with marked homogeneous enhancement following intravenous administration of gadolinium, and compression of the optic chiasm. Thus, under the preoperative diagnosis of pituitary macroadenoma, the patient underwent endoscope-assisted surgery via the transsphenoidal approach. The patient recovered well after surgery. The histopathological diagnosis was pituicytoma, WHO grade I.

Clinical discussion: Pituicytomas are defined as a circumscribed low-grade glial tumor arising from the neurohypophysis or infundibulum with bipolar spindle cells arranged in a fascicular or storiform pattern (a cartwheel). The clinical symptoms are variable depending on the tumor size and location. They usually present due to mass effect. The radiographic characteristics are not nonspecific. The diagnosis of pituicytoma is based on histopathological evidence. Pituicytomas consist of a solid proliferation of elongated spindle cells arranged in interlacing fascicles and/or in a “storiform” pattern. In immunohistochemical studies, pituicytomas was strongly expressed in TTF-1.

Conclusion: Pituicytomas are benign, slow-growing glial tumors. It is difficult to diagnosed before operation as its clinical presentations and imaging studies resemble those of non-functional pituitary adenomas. The best chance of successful treatment is gross total resection by the endoscopic approach or transcranial approach.

1. Introduction and importance

Pituicytomas are rare circumscribed low-grade glial tumors arising from the neurohypophysis or infundibulum. The tumor is slow growing and benign, and histologically corresponds to World Health Organization (WHO) grade I [1,2]. They may be easily confused with pituitary lesions, including pituitary adenoma, craniopharyngiomas, and so on, due to their location in the sellar and suprasellar region. In this study, we review the literature in order to better understand and categorize the natural history, clinical presentation, and treatments. This case report has been reported in line with the SCARE 2020 criteria [3].

2. Case presentation

A 45-year-old female patient who complained of left eye blurred vision over the left side field for 2 months. Progressive left eye blurred vision with intermittent headache and photophobia was noted over 2

weeks prior to admission.

No visual field defect was observed by neurological examination. Visual evoked potential (VEP) was normal. Endocrine studies for the pituitary gland were normal except for a mildly elevated level of serum prolactin (PRL) (61.29 ng/mL). The Brain Magnetic Resonance Imaging (MRI) study disclosed a solid sellar tumor with marked homogeneous enhancement following intravenous administration of gadolinium, and compression of the optic chiasm was noted. The tumor was isointense on T1-weighted images and hyperintense on T2-weighted images [Fig. 1]. Thus, under the preoperative diagnosis of pituitary macroadenoma with optic neuropathy, the patient underwent endoscope-assisted surgery via the transsphenoidal approach.

Gross total resection of the mass was performed by our surgeon. It bled easily during the operation, but hemostasis was successfully achieved using FIOSEAL hemostatic matrix and bipolar coagulator. An intraoperative view revealed a yellowish soft tumor with slight sella floor erosion.

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The patient recovered well after surgery. The symptom of blurred vision was much improved. In addition, no CSF leakage or transient diabetes insipidus were found. Hormone levels were within the normal ranges after operation. Postoperative visual field was examined and it was almost normal [Fig. 2].

The follow-up MRI at 1 year later after operation showed grossly

total resection without evidence of residual tumor or tumor recurrence [Fig. 1].

Pathological examination revealed fragmented of normal pituitary gland and tumor composed of mixed round to spindle, amphophilic cells, and round, eosinophilic granular cells. The tumor was close to the respiratory tract epithelium with entrapped submucosal glands. Marked

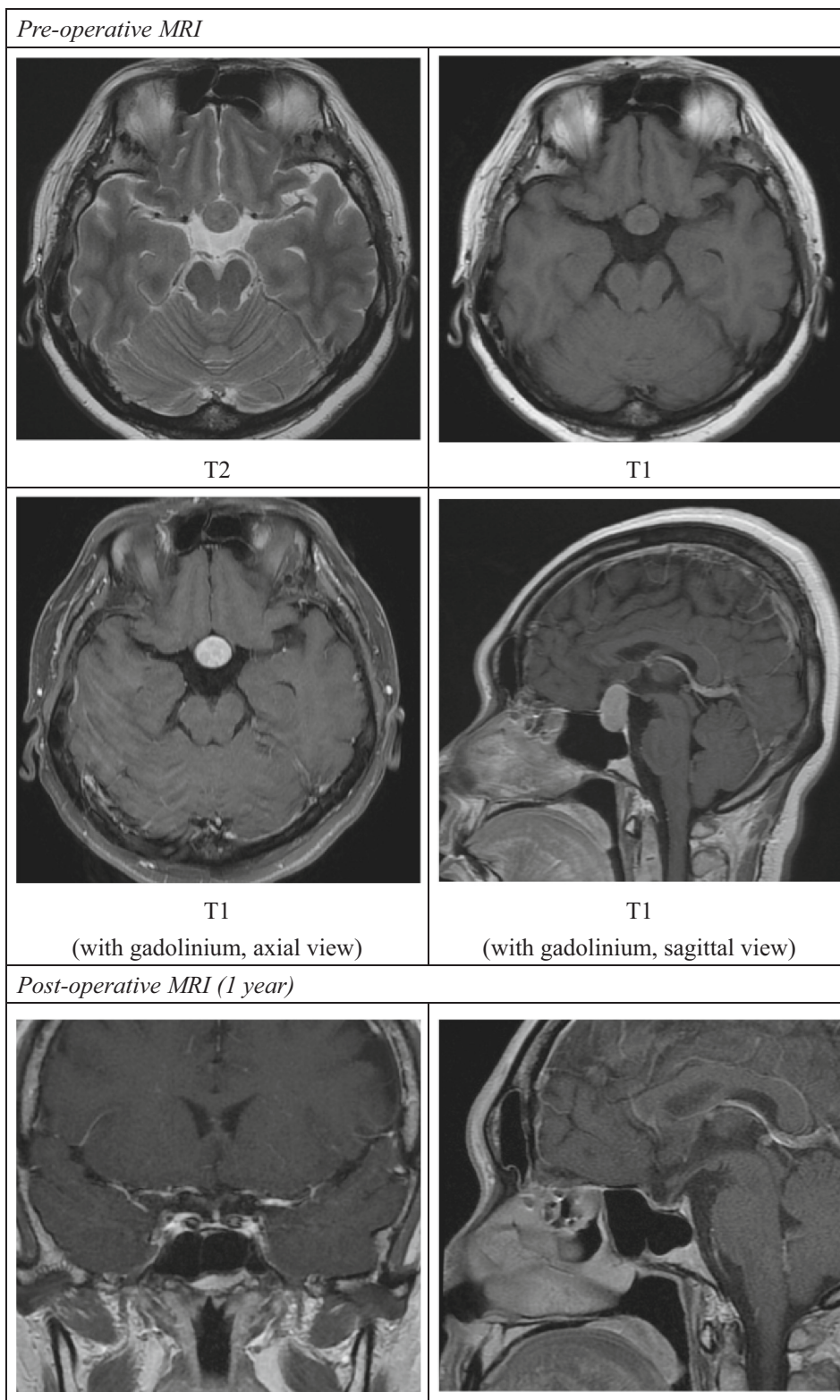


Fig. 1. Brain MRI.

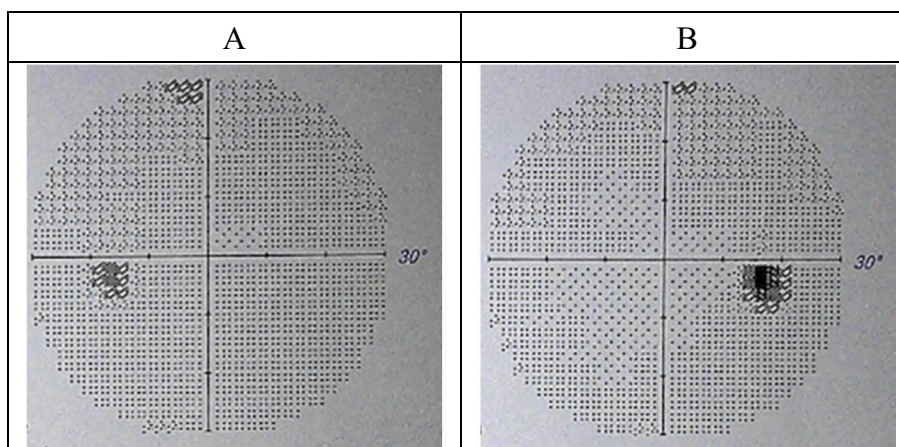


Fig. 2. Postoperative visual field. A: Oculus Sinister (O.S), B: Oculus Dexter (O.D).

lymphoplasmacytic infiltration was observed. The vasculature was also easily seen, with focal staghorn-like appearance. No tumor necrosis was found. Immunohistochemical (IHC) studies were positive for epithelial membrane antigen (EMA), S-100 protein; and diffuse strong positive staining for thyroid transcription factor 1 (TTF-1) and focal weak positive staining for CD99 were found. However, results were negative for cytokeratin (CK) AE1/AE3, CAM5.2, glial fibrillary acidic protein (GFAP), CD34, and signal transducer and activator of transcription 6 (STAT6). The histopathological diagnosis was pituitaryoma, WHO grade I. Histopathological features are shown in Fig. 3.

3. Clinical discussion

Pituitaryomas are defined as a circumscribed low-grade glial tumor arising from the neurohypophysis or infundibulum with bipolar spindle cells arranged in a fascicular or storiform pattern (a cartwheel). The tumor is slow growing and benign, and histologically corresponds to World Health Organization (WHO) grade I [2,4]. It occurs in adults with a slight predominance in males.

3.1. Clinical presentation

The clinical symptoms are variable depending on the tumor size and location. They usually present due to mass effect, such as visual acuity or visual field defects, which are caused by compression of the optic nerves and/or optic chiasm. Pituitary stalk compression results in pan-hypopituitarism or hyperprolactinemia. The patients may also complain of headache, dizziness, and fatigue. Rarely, the tumor may present with sudden onset of symptoms due to spontaneous hemorrhage [5].

3.2. Imaging findings

Pituitaryomas are solid intra- or suprasellar tumors that tend to be well circumscribed with homogeneous appearance on computed tomography (CT) or magnetic resonance imaging (MRI) scans. Sometimes, they can exhibit sellar enlargement and bony remodeling. These findings are similar to those of pituitary adenoma.

On the MRI scans, pituitaryomas commonly appeared hypointense to isointense on T1-weighted images and hyperintense on T2-weighted images with marked homogenous enhancement after administration of gadolinium [1,6–9]. This presentation demonstrates a highly vascular tumor. Therefore, the radiological differential diagnosis should include other sellar or suprasellar tumors including meningioma, craniopharyngioma, hemangiopericytoma, granular cell tumor, and pilocytic astrocytoma. The rapidity of enhancement correlates with increased

vascularity of these lesions, according to Gibbs et al. [10]. They reported the presence of a significant vascular blush of these tumors on selected internal carotid artery angiograms. Thus, we can speculate that for pituitaryoma, there may be a slightly delayed enhancement on dynamic contrast-enhanced studies, unlike pituitary adenoma, which is apparently enhanced during the earlier arterial phase. In addition, the angiogram revealed prominent arterial feeding from the superior hypophyseal arteries, which supply both the diaphragma sellae and the pituitary stalk, and the appearance of a thickened stalk suggests the tumor is of infundibular origin. Distinguishing this vascular pattern from meningioma can be problematic, but the absence of external carotid artery dural feeders favors pituitaryoma [10,11].

The radiographic preoperative characteristics of pituitaryomas are not nonspecific, and it is often difficult to identify these tumors before operation, especially for nonfunctioning pituitary adenoma in our case. The distinctive diagnosis of pituitaryoma is depended on histologic appearance and immunohistochemical findings.

3.3. Histopathology

The diagnosis of pituitaryoma is based on histopathological evidence. In the 2016 WHO classification of CNS tumors, pituitaryoma was defined as a low-grade glial tumors found in the posterior pituitary and infundibulum, presumably arising from pituitary cells. Pituitaryomas consist of a solid proliferation of elongated spindle cells arranged in interlacing fascicles and/or in a “storiform” pattern [1].

The tumor cells have an abundant eosinophilic cytoplasm and a rich capillary network. Tumor cell nuclei are round to oval, with rare to absent evidence of atypia or mitotic figures. There are no Rosenthal fibers or eosinophilic granular bodies, which usually helps to distinguish between pituitaryomas and pilocytic astrocytomas [12]. Also, there is a lack of Antoni A and B pattern, psammoma bodies, which could help to distinguish pituitaryomas from schwannomas and meningiomas.

In immunohistochemical studies, pituitaryomas was strongly expressed in TTF-1 and show negative or low-moderately positive staining for GFAP. Pituitary cells are considered to be modified neuroglial cells and show positive immunohistochemical staining for GFAP and TTF-1. TTF-1 is strongly expressed in fetal and adult human pituitary cells. It is specifically expressed in pituitaryomas, granular cell tumors, and spindle cell oncocytomas. Thus, it is useful for distinguishing pituitaryomas from other sellar tumors [13]. In addition, pituitaryomas demonstrated positive immunofluorescence staining for S-100, vimentin protein. Other markers, including EMA, SSTR2A, and synaptophysin, revealed variable staining in the literature [Table 1] [1,7,14,15].

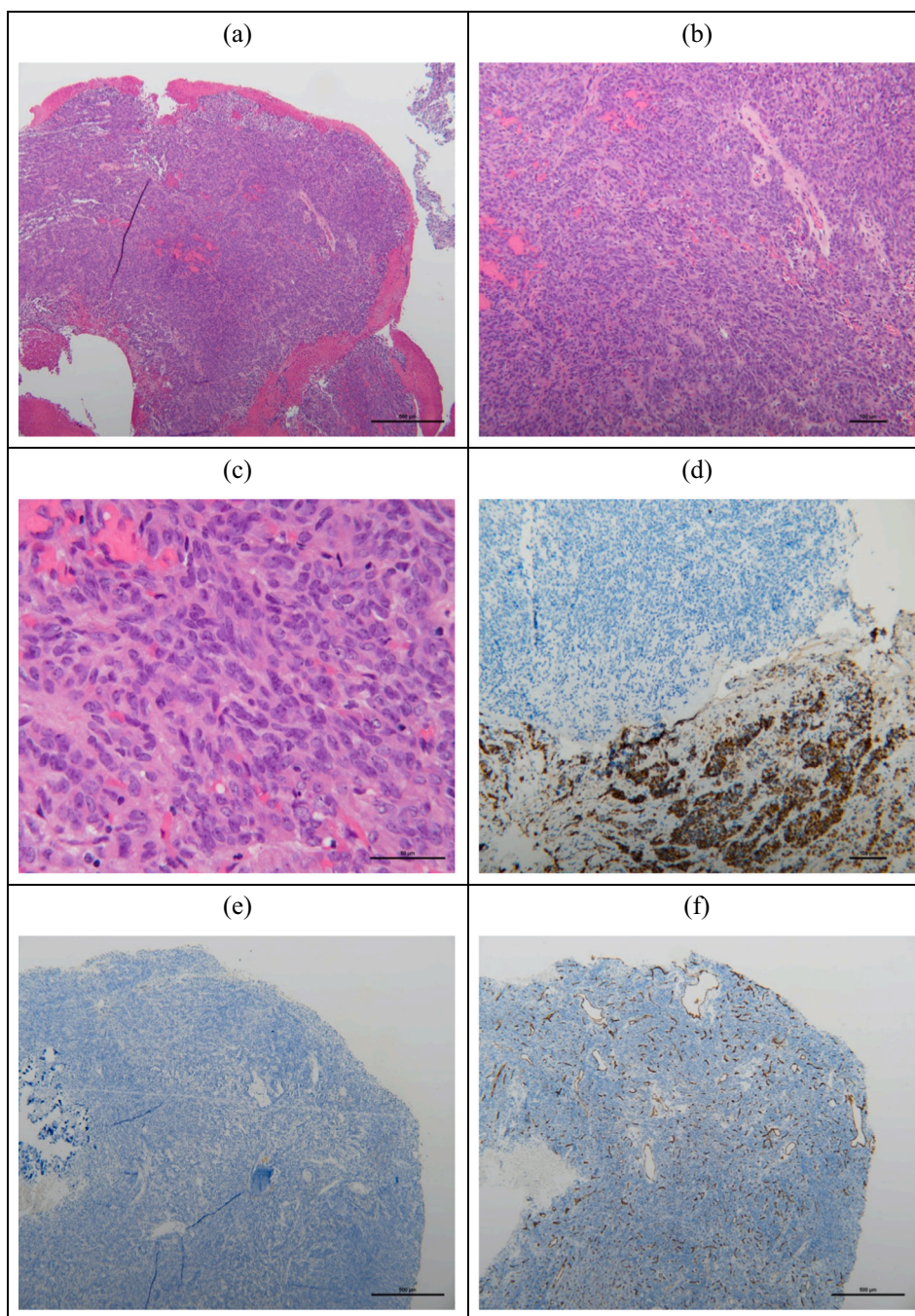


Fig. 3. Histopathologic features of pituicytoma. (a, b, c) Tumors were composed of mixed round to spindle, amphophilic cells and round, eosinophilic granular cells. (a) 500 μm (b) 100 μm (c) 50 μm . Immunohistochemical (IHC) studies: Negative for (d) Cytokeratin (CK), (e) Glial fibrillary acidic protein (GFAP), (f) CD34, (g) Signal transducer and activator of transcription 6 (STAT6), (h) CAM5.2.

Positive for (i, j) Thyroid transcription factor 1 (TTF-1): diffuse strong, (k) S-100 protein (l) epithelial membrane antigen (EMA), (m) CD99, focal weak.

3.4. Treatments

Complete resection of the tumor is the main treatment for pituicytoma. The role of adjuvant radiotherapy and chemotherapy is unclear now and needs further study for it. The local recurrence rate is usually low and no malignant transformation or cerebrospinal dissemination has been reported. The choice of surgical route used may depend on the tumor size, whether invasion to other structures has occurred, and the technique of the surgeon. The craniotomy transcranial approach, endoscope-assisted transsphenoidal approach, or even extended

endoscope-assisted transsphenoidal approach with skull base reconstruction showed difference in the rate of tumor recurrence and postoperative complications such as infection, vessels or nerve injury. Thus, the surgical resection should be also considered about functional preservation. Postoperative radiotherapy should be recommended in patients where gross total resection is not feasible. And an MRI for closely following up after operation is essential, even for those patients undergoing complete tumor resection.

Given the benign and slow-growing characteristics of pituicytomas, asymptomatic patients may choose a wait-and watch approach. When

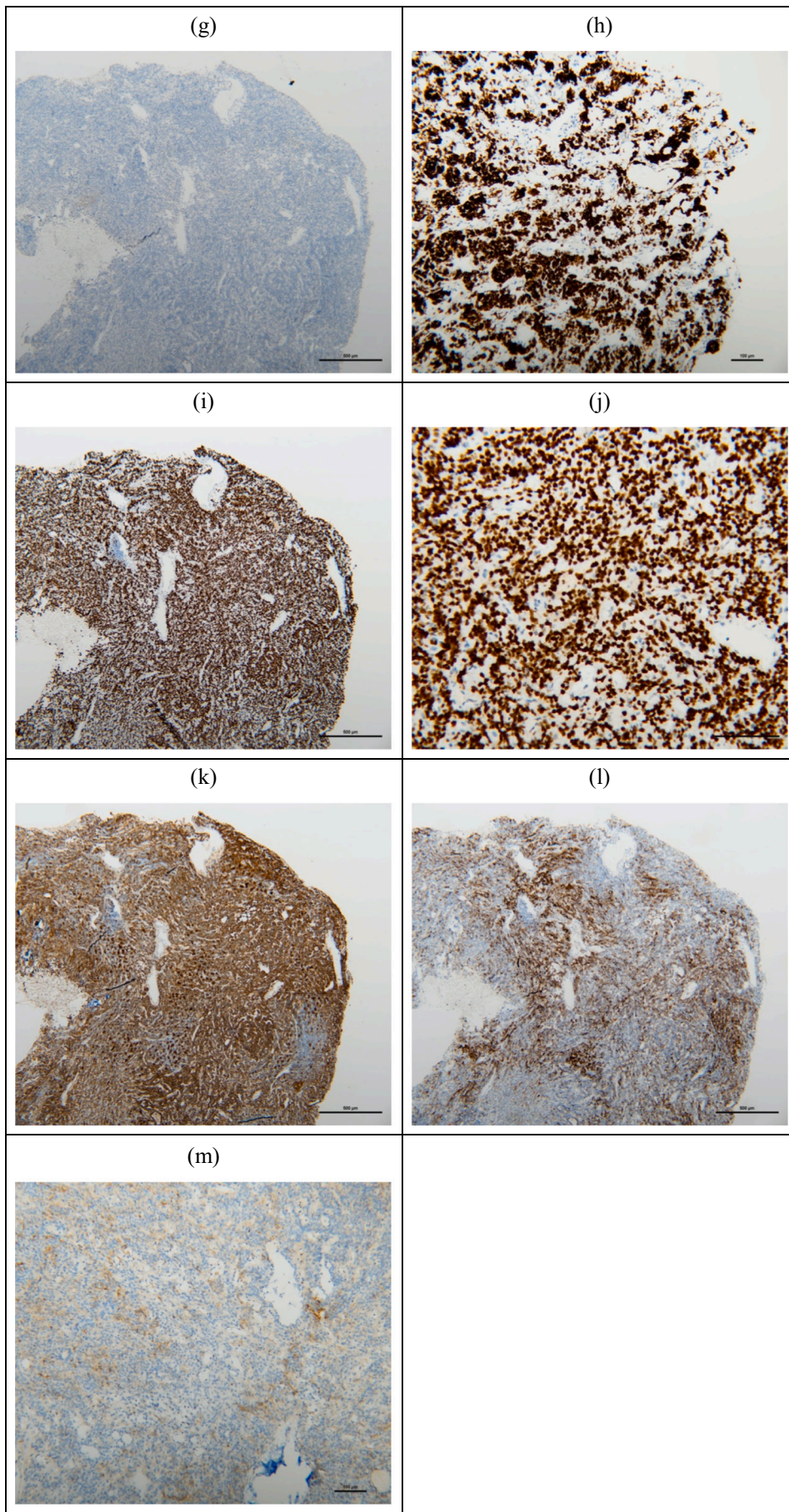


Fig. 3. (continued).

Table 1
Immunohistochemistry of Pituicytoma.

	TTF-1	GFAP	PAS	S-100	Vimentin
Pituicytoma	++	+/-	-	+/-	+/-
	Synaptophysin	EMA	α1-antitrypsin	SSTR2A	
Pituicytoma	+/-	+/-	+/-	+/-	

TTF-1, Thyroid transcription factor 1; GFAP, Glial fibrillary acidic protein; PAS, Periodic acid-Schiff.

EMA, Epithelial membrane antigen; SSTR2A, Somatostatin Receptor 2.

the tumor recurs or progresses, it is recommended to re-operate and remove the lesion completely as far as possible [16,17].

4. Conclusion

Pituicytomas are benign, slow-growing glial tumors that arise from the neurohypophysis or infundibulum. It is difficult to diagnosed before operation as its clinical presentations and imaging studies resemble those of non-functional pituitary adenomas. The best chance of successful treatment is gross total resection by the endoscopic approach or transcranial approach.

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Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

CF Shen, resident, first author, writing the paper, literature review.
SY Liu, surgeon, background research, edit the manuscript.
CH Lee, surgeon, background research, edit the manuscript.
SY Pan, background research, literature review.
CC Shen, supervision.

Research registration

Not applicable.

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Declaration of competing interest

All authors declare that they have no competing interests.

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