

A Case of Tuberculum Sellae Chordoid Meningioma Treated via Extended Endoscopic Endonasal Trans-sphenoidal Surgery

Chihiro Yagi,¹ Shun Yamamuro,¹ Yoshinari Ozawa,¹ Sodai Yoshimura,¹ Koichiro Sumi,¹ and Atsuo Yoshino¹

A 68-year-old female was admitted to our hospital with right-sided hemianopsia. Magnetic resonance imaging (MRI) demonstrated a well-enhanced tuberculum sellae region tumor. The patient underwent surgical tumor resection via an extended endoscopic endonasal trans-sphenoidal approach and the tumor was totally removed. The mass was extremely soft and there was no clear attachment between it and the dura mater. Furthermore, the histopathological findings obtained for the tumor during intra-operative rapid diagnosis were divergent from typical meningioma. We therefore diagnosed the tumor intra-operatively as a pituitary adenoma. However, the post-operative pathological diagnosis for the tumor was chordoid meningioma (CM). CM is a rare subtype of meningioma, and most of such tumors arise in the convexity. In the preoperative MRI in the present case, meningioma was suspected; however, since we did not consider CM for differential diagnosis, we failed to reach an accurate diagnosis during the operation. Tuberculum sellae CM is very rare, and only a few cases have been reported previously. The surgical strategy will differ greatly depending on whether the tumor is a meningioma or a pituitary adenoma, especially when treatment involves the dura mater. The pre and/or intra-operative diagnosis is thus very important for developing an accurate treatment strategy. We report here the details of our rare case and describe the intra-operative features of tuberculum sellae CM.

Keywords: tuberculum sellae meningioma, chordoid meningioma, trans-sphenoidal surgery, neuroendoscopy, endonasal skull base surgery

Introduction

Chordoid meningioma (CM) is a rare subtype of meningioma, categorized as WHO grade II due to its high recurrence rate and proliferative ability. Such tumors are typically found in the convexity, and rarely appear in the tuberculum sellae region.^{1,2} To date, only a few cases of CM presenting in the tuberculum sellae region have been reported.^{3–7}

¹Department of Neurological Surgery, Nihon University School of Medicine, Tokyo, Japan

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We describe here a very rare case of tuberculum sellae CM operated on by endoscopic trans-sphenoidal surgery, and discuss the treatment strategy for this rare tumor.

Case Report

A 68-year-old female visited an ophthalmic clinic complaining of visual disturbance. Since right-sided hemianopsia and a supra-sellar mass were found, she was referred to our institution. She had no significant medical history other than mild diabetes. Her consciousness level was clear and neurological examinations revealed no abnormalities except for the right-sides hemianopsia. Laboratory investigations including for pituitary hormones did not yield any abnormal values. Magnetic resonance imaging (MRI) disclosed a homogeneous well-enhanced 1.7 cm mass, iso-intense on T1-weighted and high-intense on T2-weighted imaging, in the supra-sellar region. The mass was attached to the tuberculum sellae and extended to the frontal skull base (Figs. 1a–1d). A computed tomography scan revealed no calcification of the tumor (Fig. 1e). An angiogram showed no tumor stain and/or feeding arteries (Fig. 1f). The mass was diagnosed pre-operatively as a meningioma. The patient underwent surgical tumor resection via an extended endoscopic endonasal trans-sphenoidal approach. The tumor was very soft and easily suctioned (Figs. 2a–2d). There was no attachment between it and the dura mater. Since the tumor hardness and intra-operative histopathological findings were divergent from a typical meningioma (Figs. 3a and 3b), we mis-diagnosed the tumor as a pituitary adenoma. In accordance with the intra-operative diagnosis, we preserved the dura mater as far as possible to prevent cerebrospinal fluid leakage. The skull base was repaired by using subcutaneous fat tissue and a nasal septum mucosal flap.

Pathological examinations demonstrated that the epithelial-like eosinophilic tumor cells were arranged in cords or trabeculae running through an abundant mucoid matrix. The chordoid areas of the tumor specimen amounted to more than 50% and were interspersed with microcystic and metaplastic type cells (Figs. 4a and 4b). The tumor cells were positive for periodic acid schiff (PAS) and mucicarmine staining (Figs. 4c and 4d). Immunohistochemically, they exhibited positive expression for epithelial membrane antigen (Fig. 4e), whereas there were negative expressions for S-100, cytokeratin, glial fibrillary acid protein, synaptophysin and chromogranin A. These pathological findings were consistent with CM. The MIB-1 labeling index was <3% (Fig. 4f). The patient's post-operative course was uneventful, and MRI revealed no residual tumor (Figs. 5a and 5b).

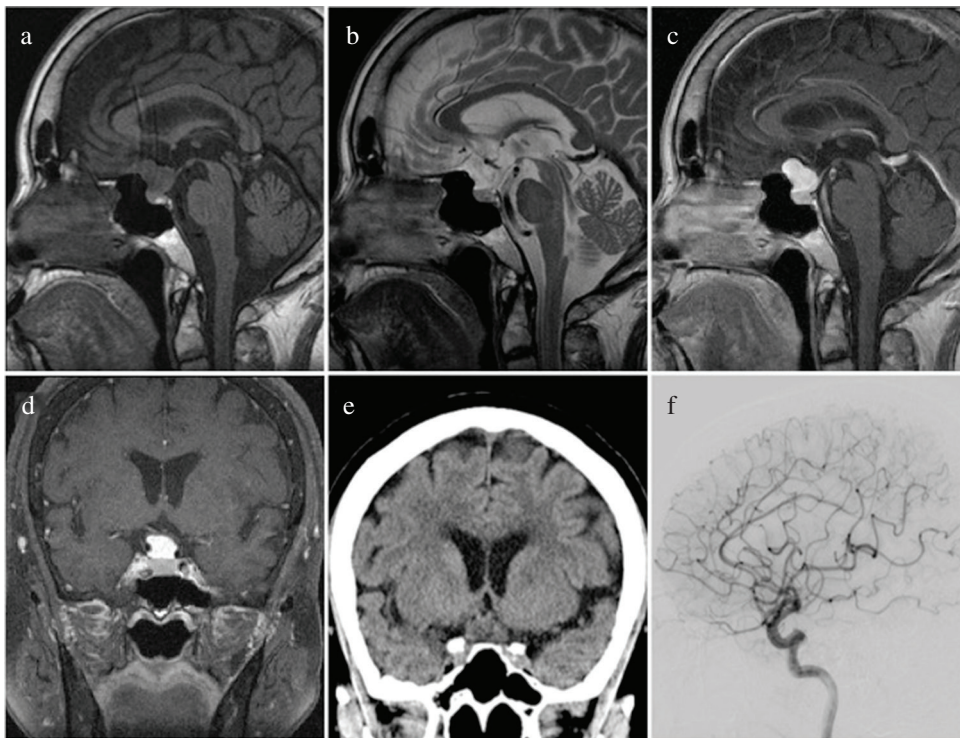


Fig. 1 (a–d) Pre-operative magnetic resonance imaging. The mass showed iso-intensity on T1-weighted imaging (a), and high-intensity on T2-weighted imaging (b). The mass was strongly and uniformly contrasted on T1-weighted imaging with gadolinium-enhancement (c and d). Neither peritumoral edema nor dura mater thickness was observed. (e) Pre-operative computed tomography scan. The mass displayed no calcification. (f) Pre-operative angiogram. There was no tumor stain and/or feeding arteries.

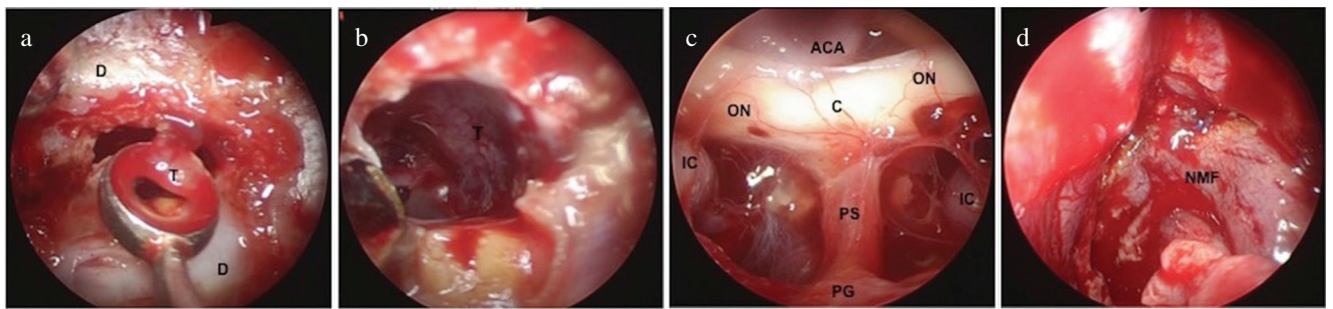


Fig. 2 Intra-operative findings. (a and b) Images obtained during tumor removal. The tumor was very soft and could be easily removed. (c) Image obtained after tumor removal. The tumor was totally removed. (d) Image obtained during skull base plasty. A nasal septum mucosal flap was used to prevent cerebrospinal fluid leakage. D: dura mater, T: tumor, ACA: anterior cerebral artery, C: chiasma, IC: internal carotid artery, ON: optic nerve, PS: pituitary stalk, PG: pituitary gland, NMF: nasal mucosal flap.

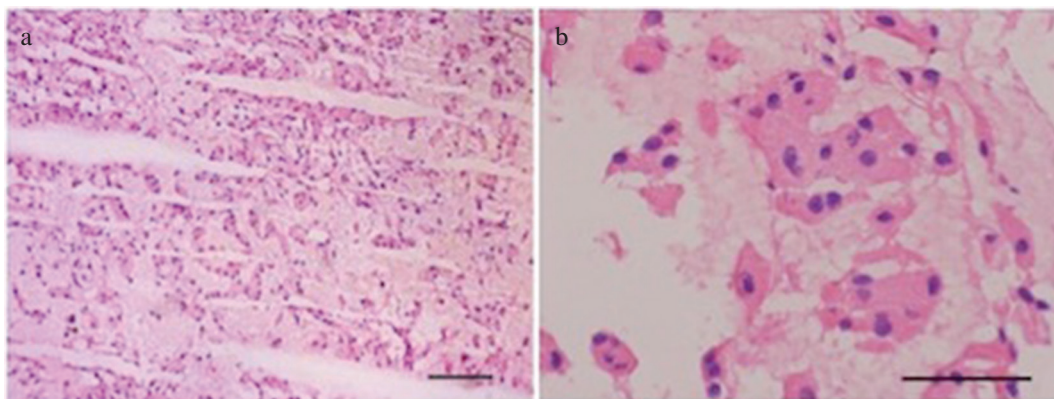


Fig. 3 Findings of intra-operative rapid pathological diagnosis. There was divergence from a typical meningioma. (a) Bar = 100 μ m. (b) Bar = 50 μ m.

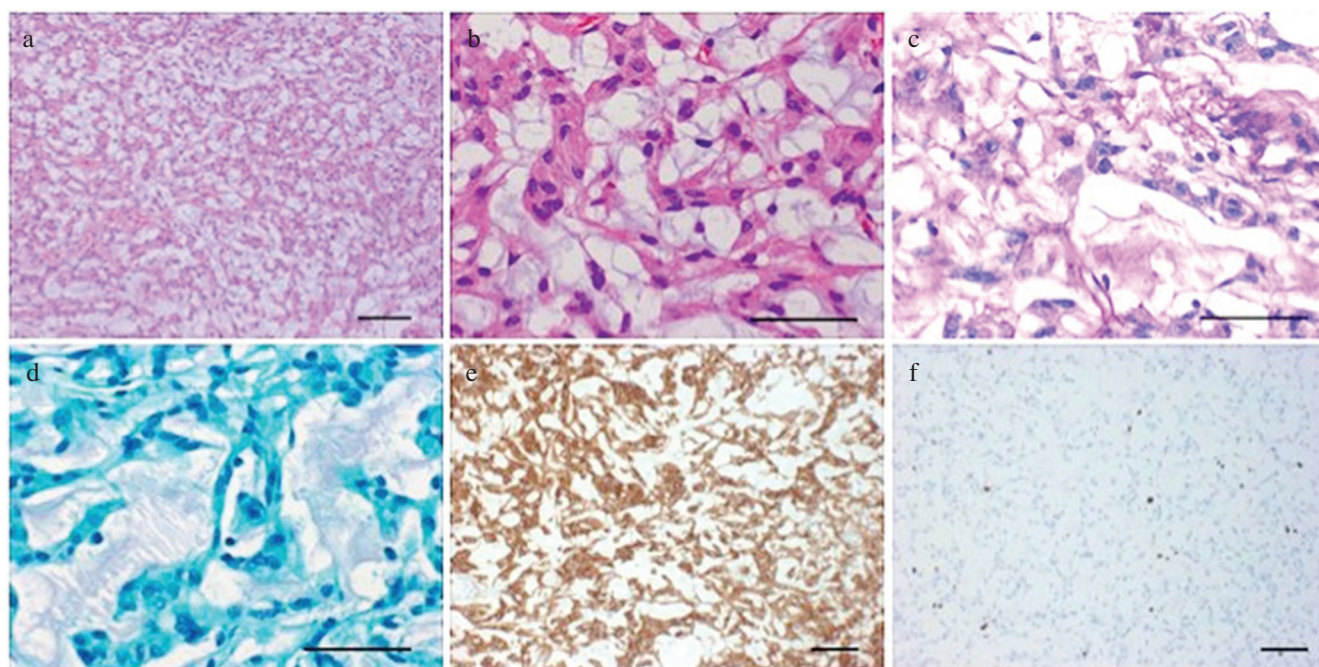


Fig. 4 Pathological findings. (a and b) Eosinophilic epithelioid or spindle tumor cells were arranged in cords or trabeculae within a pale basophilic mucoid matrix. (c and d) A mucous-rich matrix was also observed on PAS (c) and mucicarmine (d) staining. (e) The tumor cells were positive for epithelial membrane antigen. (f) Ki-67 positive cells amounted to <3%. (a, e, f) Bar = 100 μ m. (b, c, d) Bar = 50 μ m.

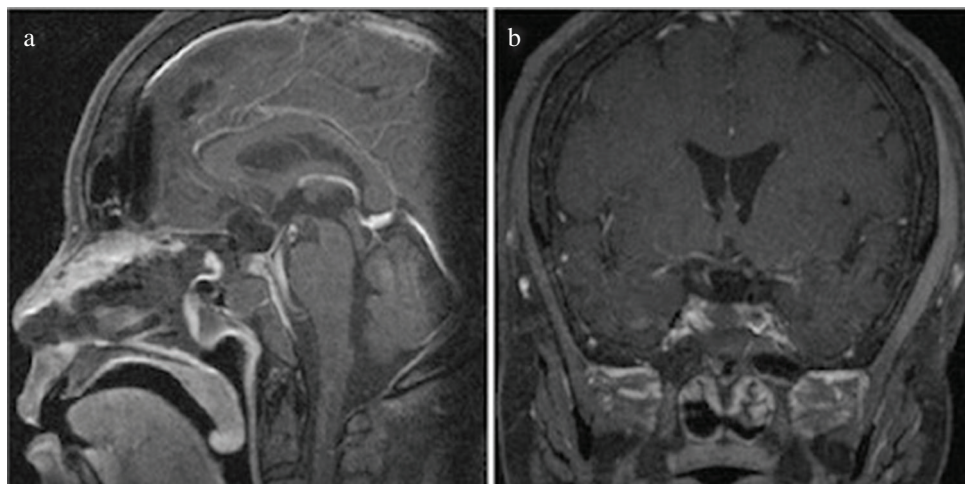


Fig. 5 Post-operative magnetic resonance imaging with gadolinium-enhancement performed on the day after operation (a: sagittal view, b: coronal view). There was no enhanced lesion.

Discussion

Chordoid meningioma is a rare subtype of meningioma (0.3–1.0%).²⁾ Indeed, only two cases of CM have been diagnosed among 311 meningiomas (0.64%) in the last 8 years at our institution (Nihon University Itabashi Hospital). This tumor was slightly more prevalent in females, and the mean age was the mid-30–40s.^{1,8,9)} Choy et al.¹⁾ noted that recurrence was encountered in 51 of 221 patients (23.1%), and the median time of progression free survival was 136 months. According to past reviews, the tumors typically developed from the supratentorial region and the most common site was the convexity.^{1,9)} CM that develops from the tuberculum sellae region is very rare. To date, only 10 cases of tuberculum sellae CM have been reported in the English

literature.^{3–7)} We describe here a very rare case of tuberculum sellae CM, which was surgically treated via an endonasal approach using a neuro-endoscope.

With the recent progress in neuro-endoscopic surgery, opportunities to remove frontal skull base tumors via an endonasal approach are increasing. Since the frequency of post-operative cerebrospinal fluid leakage is higher in the endonasal skull base surgery, secure skull base plasty, including manipulation of the dura mater, is important. On the other hand, when operating on meningiomas, the dura mater requires reliable processing to prevent recurrence. In this context, the operation strategy will be greatly different depending on whether the tumor is a meningioma or not. The pre and/or intra-operative diagnosis thus becomes

important in deciding the treatment strategy. However, rare subtypes of meningiomas are sometimes difficult to diagnose. Since the intra-operative findings in the present case were divergent from typical meningiomas, it proved difficult to make an accurate diagnosis. During the operation, we thought that the tumor might be a pituitary adenoma, based on the following three enigmatic points. First, the tumor was extremely soft and easily suctioned. Second, there was no attachment between the tumor and tuberculum sellae dura mater. Third, we could not identify the pathological features of typical meningiomas during the intra-operative rapid pathological diagnosis procedure. Since we diagnosed the tumor as a pituitary adenoma, we preserved the dura mater to prevent cerebrospinal fluid leakage. This might have increased the chances of recurrence of meningioma. Ultimately, the main cause of the intra-operative misdiagnosis in the present case was a lack of knowledge and experience regarding CM. It is important therefore to keep CM in mind in the differential diagnosis of tumor of this region.

We strongly regarded the tumor to be a meningioma before surgery, because the MRI in the present case was very typical of meningioma. However, we did not consider preoperatively the rare type of meningioma, CM, existing in the tuberculum sellae region. Baal et al.¹⁰ reported that diffusion weighted MRI (DWI) and apparent diffusion coefficient (ADC) values were useful in the preoperative prediction of CM, since the ADC values of CM tended to be significantly higher than those in other meningioma subtypes. It could have been helpful if we had undertaken DWI preoperatively, but we did not. Moreover, in retrospect, intra-operative confirmation of tumor attachment with the dura mater would not have been sufficient, because the skull base bone in the tuberculum sellae region was inadequately opened. Since we mis-diagnosed the tumor as a pituitary adenoma, the tumor origin was considered to be the pituitary gland during the operation, so that skull base opening might be inadequate. If attachment could have been verified at operation, mis-diagnosis might not have occurred.

A therapeutic consensus for CM has not yet been established. There have been several reports recommending postoperative radiation therapy because of the tumor's high recurrence rate.^{9,11} On the other hand, Wang et al.⁴ indicated that there was no statistical difference in tumor recurrence regardless of post-operative radiation therapy, when gross total resection was achieved. The rate of tumor resection by surgery is considered as the factor most related to tumor

recurrence.^{1,4,6,11} On the other hand, Choy et al.¹ reported that tumor recurrence was confirmed in 8.7% of cases within 2 years even if the mass was totally removed. Since there was no residual tumor in our case, postoperative radiation therapy has not been performed. Furthermore, the tuberculum sellae dura mater, which might be considered as the site of tumor genesis, still remains. For these reasons, close follow-up is necessary in the present patient since she has a risk of recurrence.

In conclusion, CM should be considered in the differential diagnosis of tumors of tuberculum sellae region. Further basic and clinical research on additional cases is also needed, in order to establish a therapeutic consensus for tuberculum sellae CM in the future.

Conflicts of Interest

The authors declare that they have no financial or other conflicts of interest in relation to the present research and its publication.

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Corresponding author:

Shun Yamamuro, MD, PhD, Department of Neurological Surgery, Nihon University School of Medicine, 30-1 Oyaguchi-kamicho, Itabashi-ku, Tokyo 173-8610, Japan.

✉ yamamuro.shun@nihon-u.ac.jp