

Case Report

A craniopharyngioma with spontaneous involution of a gadolinium-enhanced region on magnetic resonance imaging

Yasuyuki Kinoshita, Atsushi Tominaga, Satoshi Usui, Kaoru Kurisu

Department of Neurosurgery, Graduate School of Biomedical and Health Sciences, Hiroshima University, Hiroshima 734-8551, Japan

E-mail: *Yasuyuki Kinoshita - y-kinoshita@hiroshima-u.ac.jp; Atsushi Tominaga - atom@hiroshima-u.ac.jp; Satoshi Usui - usui@hiroshima-u.ac.jp;
Kaoru Kurisu - kuka422@hiroshima-u.ac.jp

*Correspondence author

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Abstract

Background: It has not been reported previously that the solid enhancing portion of a craniopharyngioma has involuted without a change in cyst size.**Case Description:** We herein report a case of a craniopharyngioma with spontaneous involution of a solid gadolinium (Gd)-enhanced region on magnetic resonance imaging (MRI). A 44-year-old female was referred to our department with a mass on MRI associated with headaches and polyuria. The images showed a suprasellar cystic lesion progressing from the posterior pituitary lobe to the right hypothalamus along the pituitary stalk. Examinations of the cerebrospinal fluid showed aseptic meningitis and a positive titer of beta-human chorionic gonadotropin (HCG- β) preoperatively. The hypothalamic lesion became enlarged over the following 3 weeks, and a biopsy of the posterior pituitary lobe was performed via the endonasal transsphenoidal approach under a preoperative diagnosis of a germ cell tumor (GCT). The histological diagnosis was a craniopharyngioma, and the patient's postoperative findings on MRI were atypical: The solid Gd-enhanced region in the hypothalamus had spontaneously decreased in size and the peritumoral edema had improved, although the biopsy site in the posterior pituitary lobe was distant from the area of shrinkage. We speculated that the involutinal portion on MRI mimicking a tumor was actually the normal hypothalamus, which was abnormally enhanced due to a disruption of the blood-brain barrier caused by the craniopharyngioma.**Conclusion:** Gd-enhanced regions of parenchyma neighboring the cysts of craniopharyngioma should be carefully managed, taking into consideration the possibility of the enhancement of normal tissue.**Key Words:** Craniopharyngioma, HCG- β , involution, inflammation, meningitis**Access this article
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INTRODUCTION

Craniopharyngiomas can spontaneously decrease in volume due to a rupture of the cyst.^[5,8,9,11] In contrast, no cases involving shrinkage of the solid enhanced

region within a short period have been reported. We herein report an atypical case of a craniopharyngioma associated with the spontaneous involution of a gadolinium (Gd)-enhanced region on magnetic resonance imaging (MRI) after a biopsy.

CASE REPORT

A 44-year-old female was referred to our department with a suprasellar mass on MRI associated with headaches and polyuria. A neurological examination showed bitemporal lower quadrantanopsia. Endocrinological tests revealed disturbances in the secretion of ACTH, TSH, LH, and FSH and diabetes insipidus (DI). MRI demonstrated an intra- and suprasellar cystic lesion appearing as an iso-/low-intensity region on a T1-weighted image (WI) and an iso-/high-intensity region on a T2-WI [Figure 1a, b]. The edematous area in the right hypothalamus was evaluated with T2-WI. Gd-DTPA coronal and sagittal T1-WI showed the presence of an enhanced irregular area progressing from the posterior pituitary lobe to the right hypothalamus along the pituitary stalk [Figure 1c, d]. Computed tomography (CT) disclosed an isodense lesion with a small area of calcification [Figure 2a, b]. The serum titers of beta-human chorionic gonadotropin (HCG- β) and alpha-fetoprotein (AFP) were undetectable; however, the titer of HCG- β (1.4 mIU/ml) was positive in the cerebrospinal fluid (CSF). Examinations of the CSF also revealed findings of aseptic meningitis, including a xanthochromic appearance, a high monocyte count (total cells: 46/3 μ L, polycytes: 0/3 μ L, and monocytes: 46/3 μ L), a normal glucose level (44 mg/dL), and elevation of proteins (253 mg/dL) in CSF.

The lesion had become enlarged, infiltrating the hypothalamus by 3 weeks after the initial MRI study [Figure 1e, f], and a biopsy was performed via the transsphenoidal approach through the right nostril under a preoperative diagnosis of a germ cell tumor (GCT). Hydrocortisone sodium succinate (300 mg) was additionally injected to prevent acute adrenocortical insufficiency on the day of the procedure. Therapy with hydrocortisone (20 mg/day), thyroid hormone (50 μ g/day) and DDAVP (5.0 μ g/day) was administered after the biopsy. Biopsy specimens were obtained from the posterior pituitary lobe to generate permanent sections for a histological analysis. The histological examination showed multistratified epithelial tissue, identifying the tumor to be a papillary craniopharyngioma [Figure 3].

The tumor exhibited an atypical postoperative course in that [Figure 4a-c]. To expect furthermore the shrinkage of the tumor, MRI was performed 2 months after the biopsy and showed additional involution of the enhanced area with an improvement in the edema in the hypothalamus [Figure 4d-f]. Transcranial surgery was performed to remove the residual tumor via the subfrontal-pterional approach with orbitotomy 2 months after the biopsy. The tumor was sub-totally removed, and the content of the cyst was found to consist of yellowish mucus similar to motor oil. The second histological examination also identified the tumor to be a papillary craniopharyngioma.

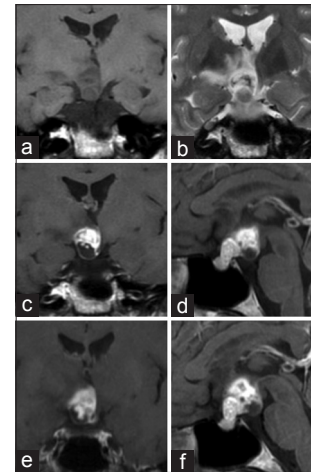


Figure 1: Initial coronal (a-c) and sagittal (d) magnetic resonance imaging results showing a cystic tumor in the right hypothalamus, pituitary stalk and posterior pituitary lobe as a low-/iso-intensity area on a T1-weighted image (a) and an iso-/high-intensity area on a T2-weighted image (b), with enhancement on T1-weighted images following gadolinium injection (c and d). An edematous area was observed in the right hypothalamus on the T2-weighted image (b). Preoperative Gd-enhanced coronal (e) and sagittal (f) T1-weighted images obtained 3 weeks after the initial examination showing an enlarged lesion infiltrating into the right hypothalamus

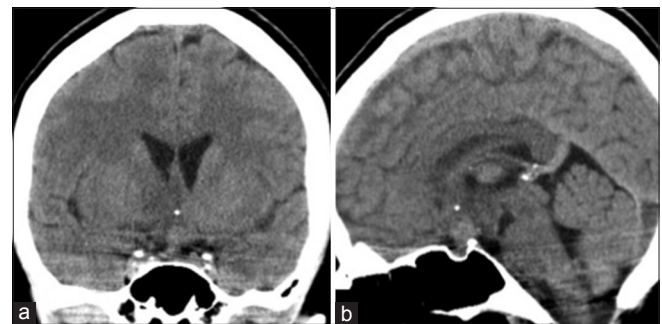


Figure 2: Initial coronal (a) and sagittal (b) images obtained using computed tomography showing an isodense tumor in the suprasellar region with a small area of calcification

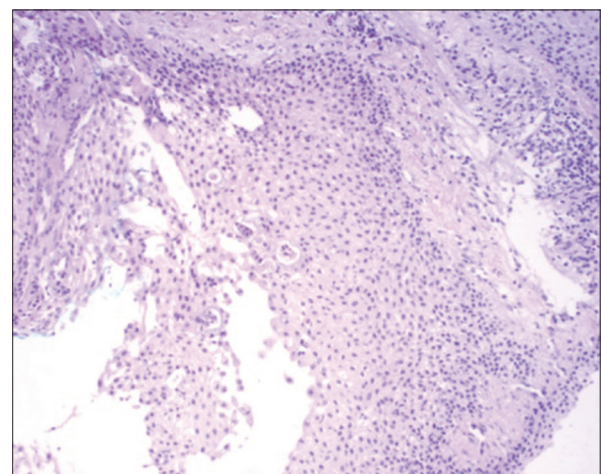


Figure 3: Photomicrographs of the tumor. Original magnification $\times 100$. Hematoxylin and eosin staining revealed a fragment showing multistratified epithelial tissue with focal palisading of nuclei and myxoid fibrous tissue

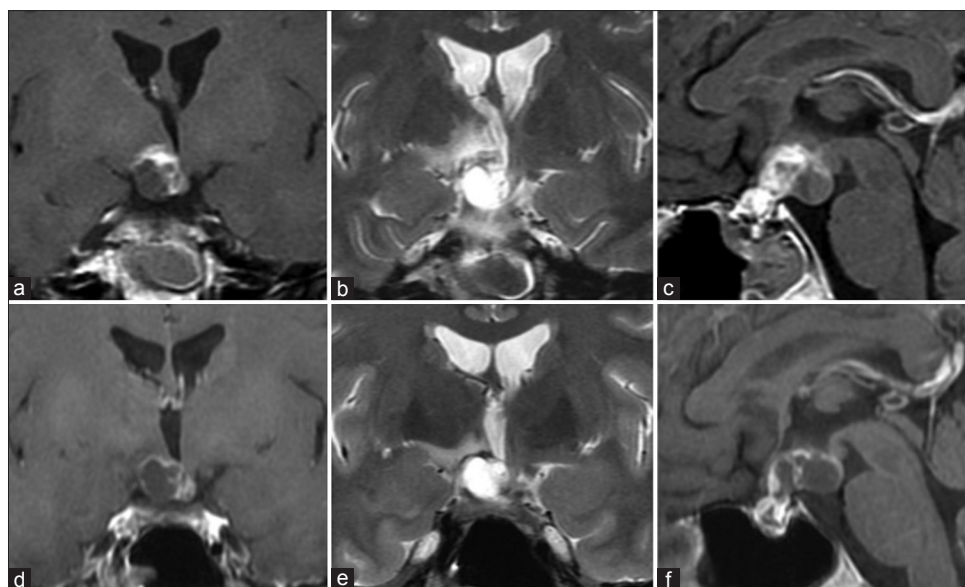


Figure 4: Gd-enhanced coronal (a) and sagittal (c) T1-weighted images obtained 6 days after the biopsy showing involution of the Gd-enhanced lesion in the right hypothalamus. The edema in the right hypothalamus persisted on a coronal T2-weighted image (b). Two months after the biopsy, Gd-enhanced coronal (d) and sagittal (f) T1-weighted images showing further involution of the Gd-enhanced lesion in the right hypothalamus with an improvement in the edema on a coronal T2-weighted image (e)

DISCUSSION

In the present case, the titer of HCG- β was positive in the CSF and the tumor seemed to infiltrate into the intraaxial hypothalamus within a short period on preoperative MRI. Moreover, the Gd-enhanced region in the hypothalamus exhibited spontaneous involution with an improvement in the perifocal brain edema after the biopsy of the posterior pituitary lobe. These findings were confusing with respect to our management of the tumor based on the histological results of the biopsy.

HCG- β is a well-known useful tumor marker for GCT.^[1] However, the immunohistochemical expression of HCG- β had been detected in various tumors other than GCTs, including cystic pituitary adenomas, Rathke's cleft cysts, and craniopharyngiomas.^[2,3,10] In this case, we could not distinguish a craniopharyngioma from a GCT by the detection of HCG- β in the CSF.

The spontaneous involution of craniopharyngiomas has been reported in literature.^[5,8,9,11] The reduction in size, however, was caused by the spontaneous rupture of the cyst. It was difficult to understand why the solid Gd-enhanced region spontaneously shrank, because we interpreted the Gd-enhanced region as a part of the tumor. We retrospectively speculate that we may have observed a normalization of the blood-brain barrier permeability in peritumoral normal brain. The leakage of cyst content can induce chemical aseptic meningitis,^[5,9] while the cysts of craniopharyngioma lesions can stimulate inflammation in peritumoral structures.^[4,6,7] In the present case, findings of aseptic meningitis were also observed, and it is reasonable to consider that the cyst contents

of the craniopharyngioma-induced inflammation and disrupted the blood-brain barrier in the hypothalamus, mimicking a Gd-enhanced tumor on MRI. Although the precise pathophysiology underlying the improvement in the enhanced hypothalamus cannot be explained, the administration of steroids or improvements in meningitis may relieve inflammation in the hypothalamus. The present case indicates the possibility of a disrupted blood-brain barrier in a patient with a craniopharyngioma that presented as a Gd-enhanced area of normal brain tissue, mimicking a tumor.

CONCLUSIONS

We herein reported an atypical case of a craniopharyngioma showing the spontaneous involution of a Gd-enhanced region in the hypothalamus on MRI. We speculate that the features observed in this case were not due to shrinkage of the craniopharyngioma itself, but rather improvements in abnormally enhanced hypothalamic tissue neighboring the craniopharyngioma. Gd-enhanced regions of parenchyma neighboring the cysts of craniopharyngioma should be carefully managed, taking into consideration the possibility of the enhancement of normal tissue.

Disclosure statement

The content of this manuscript has not been published in full elsewhere in any form. The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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