Intrasellar Chordoma Mimicking Pituitary Macroadenoma with Hyperprolactinemia and Hypopituitarism: Clinical Images with a Surgical Video

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Intrasellar chordoma mimicking pituitary macroadenoma with hyperprolactinemia and hypopituitarism: Clinical images with a surgical video

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Short title: Chordoma mimicking pituitary adenoma

Intrasellar chordoma mimicking pituitary macroadenoma with hyperprolactinemia and hypopituitarism: Clinical images with a surgical video

Abstract

A purely intrasellar chordoma is rare among skull base chordomas and is recognized as originating from ectopic embryological notochord located in the sella turcica. In view of its rarity and non-specific symptoms, clinicians may misdiagnose intrasellar chordoma as pituitary adenoma based on preoperative radiographic images. In this report, we present an intrasellar chordoma that clinically mimicked pituitary macroadenoma with hyperprolactinemia and hypopituitarism and was successfully resected by endoscopic endonasal transsphenoidal surgery. This case demonstrated radiographic features that chordoma should be suspected in sellar lesions. The enlarged sellar with thinned remodeled bone without clival destruction was firstly reminiscent of pituitary adenoma, whereas the very high signal on T2-weighted images and heterogeneous enhancement characteristically suggested chordoma. This rare diagnosis must be considered in the preoperative evaluation of sellar lesions because it can affect how the neurosurgeon prepares for surgery and the surgical goals.

Introduction

Skull base chordomas are centrally located and most commonly in the clivus directly below the sella turcica.[1,2] These tumors that form in the intrasellar region rather than as an upward extension of the clival region are extremely rare.[3,4] Purely intrasellar chordomas are recognized as originating from ectopic notochordal tissue located in the sella turcica.[5] Clinicians may make a misdiagnosis of pituitary adenoma based on preoperative radiographic images due to the rarity of intrasellar chordoma and its non-specific symptoms.[6–14]

This report describes an intrasellar chordoma that mimicked pituitary macroadenoma with hyperprolactinemia and was resected by an endoscopic endonasal transsphenoidal approach. We hope that this report and its accompanying surgical video will be helpful for neurosurgeons who encounter similar cases.

Case presentation

A 44-year-old woman presented with progressive visual impairment. Neuro-ophthalmic examination revealed bitemporal hemianopia and left visual impairment. Computed tomography imaging showed an isodense sellar lesion without clival destruction (Figure 1). Magnetic resonance imaging showed a heterogeneously contrast-enhanced solid tumor (28 × 24 × 31 mm) with very high-intensity on T2-weighted images (Figure 2). Endocrine examination showed hyperprolactinemia (128.6 ng/ml), hypothyroidism, adrenal insufficiency, and diabetes insipidus. There was no evidence of inflammatory disease or malignancy in blood examinations.

The patient underwent endoscopic endonasal transsphenoidal surgery for resection of the lesion. On opening the dura of the sellar floor, we identified an encapsulated, soft, pinkish tumor that was distinct from normal pituitary gland tissue (Video 1). The tumor was successfully resected by debulking followed by microdissection (Figure 3). Histopathological

examination was positive for brachyury, which is typically associated with chordoma (Figure 4). Postoperatively, the patient's visual impairment resolved completely, although hormone replacement for hypopituitarism was needed. Follow-up is ongoing with a strategy of waiting for reoperation/radiotherapy until recurrence because of the achievement of total resection and low Ki-67 index (3%).

Discussion

Intrasellar chordoma is a rare diagnosis that must be considered in the evaluation of sellar lesions because it can affect the neurosurgeon's preparation, surgical goals, and postoperative management.[15] If neurosurgeons encounter an intrasellar chordoma during surgery, an appropriate response with standard techniques for maximal resection is required.

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Declarations of interest

None.

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112 **Figure legends**

113 **Figure. 1** Preoperative computed tomography images. Sagittal brain window image (A)
114 shows an isodense sellar lesion. Sagittal (B) and axial (C) bone window images show
115 ballooning of the sella turcica and thinning of the dorsum sellae without intratumoral
116 calcification or clival invasion.

117

118 **Figure. 2** Preoperative sagittal (A–C), coronal (E, F), and axial (G, H) magnetic resonance
119 images with position of coronal and axial slice (H). A solid sellar tumor with the right
120 cavernous sinus invasion shows iso-intensity on T1-weighted image (A), very high-intensity
121 on T2-weighted images (C, E, G), and well-enhanced margin with heterogeneous internal
122 enhancement on contrast-enhanced T1-weighted images (B, D, F).

123

124 **Figure. 3** Postoperative sagittal (A), coronal (B), and axial (C) contrast-enhanced T1-
125 weighted images confirm total resection of the tumor with anatomic preservation of the
126 normal pituitary gland tissue (white arrow in B).

127

128 **Figure. 4** Photomicrographs show tissue corresponding to histopathological diagnosis of
129 chordoma (A, B).

Fig. 1

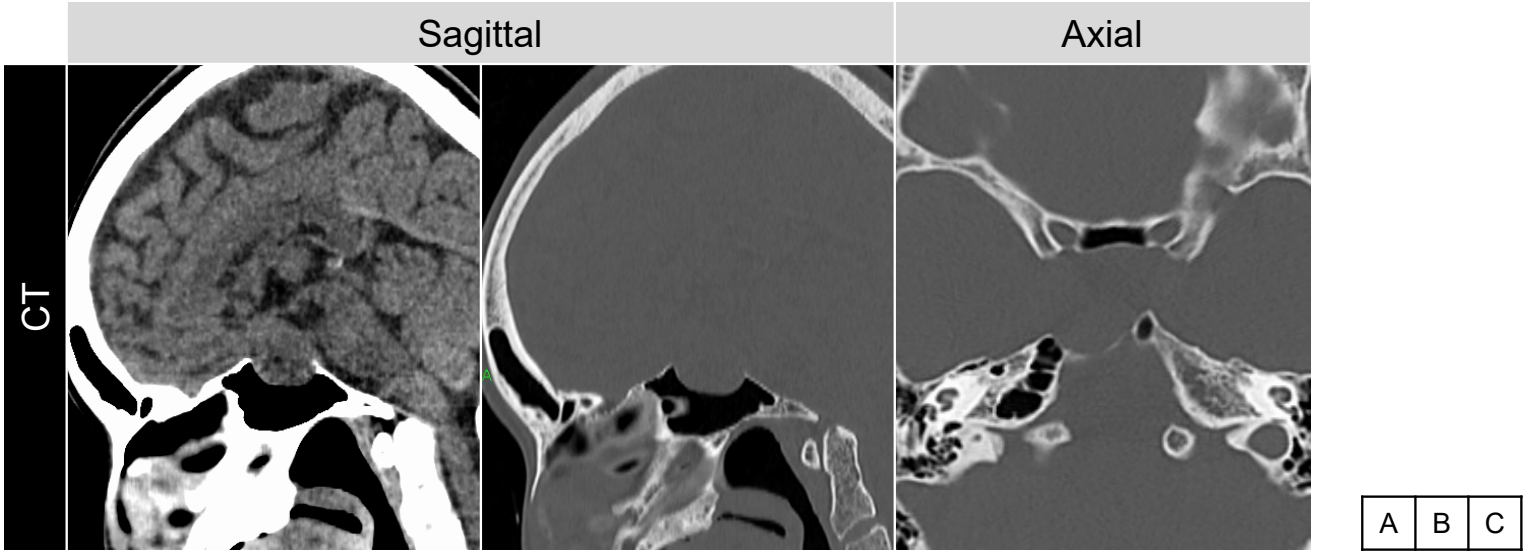
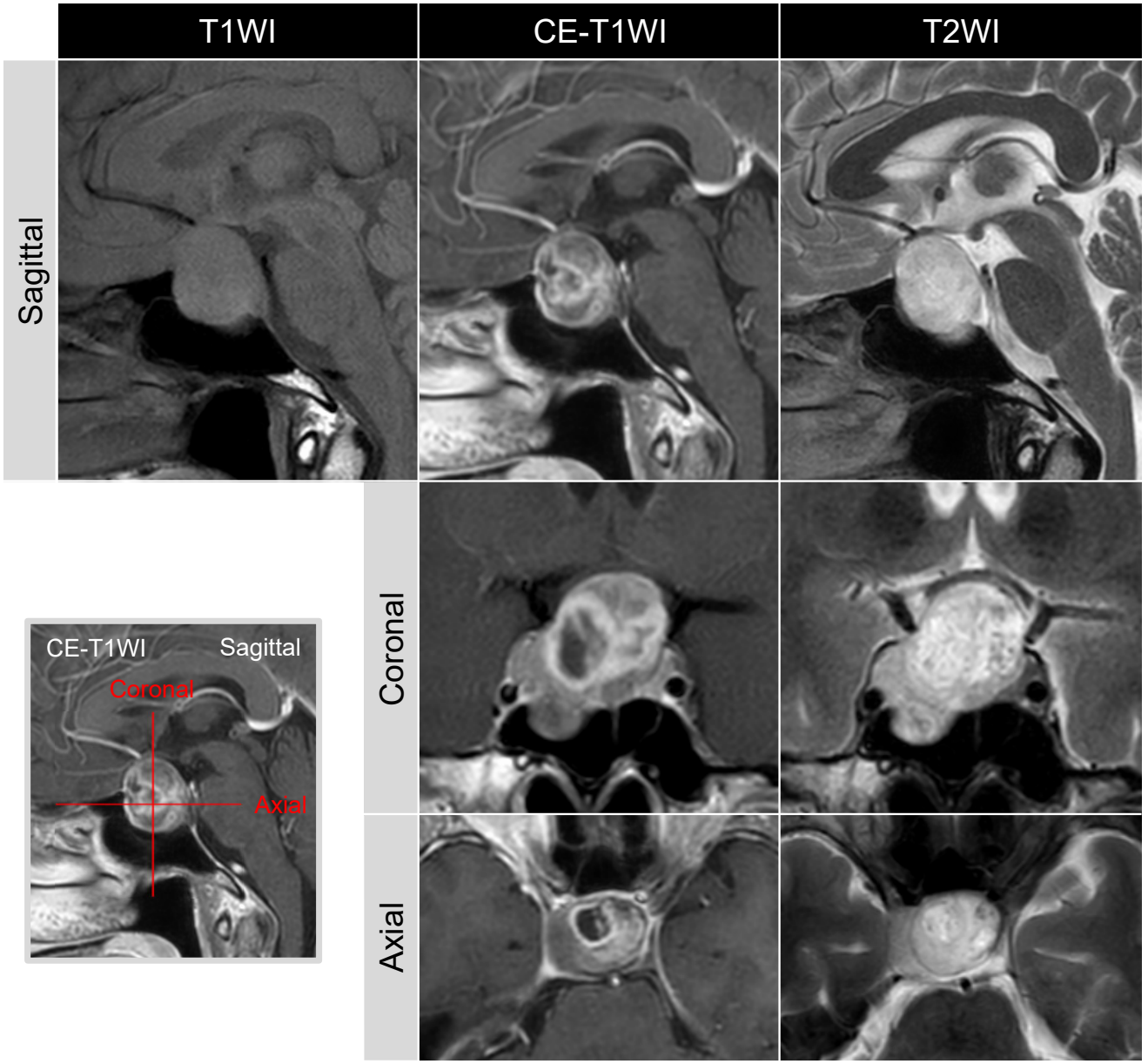


Fig. 2



A	B	C
H	D	E
	F	G

Fig. 3

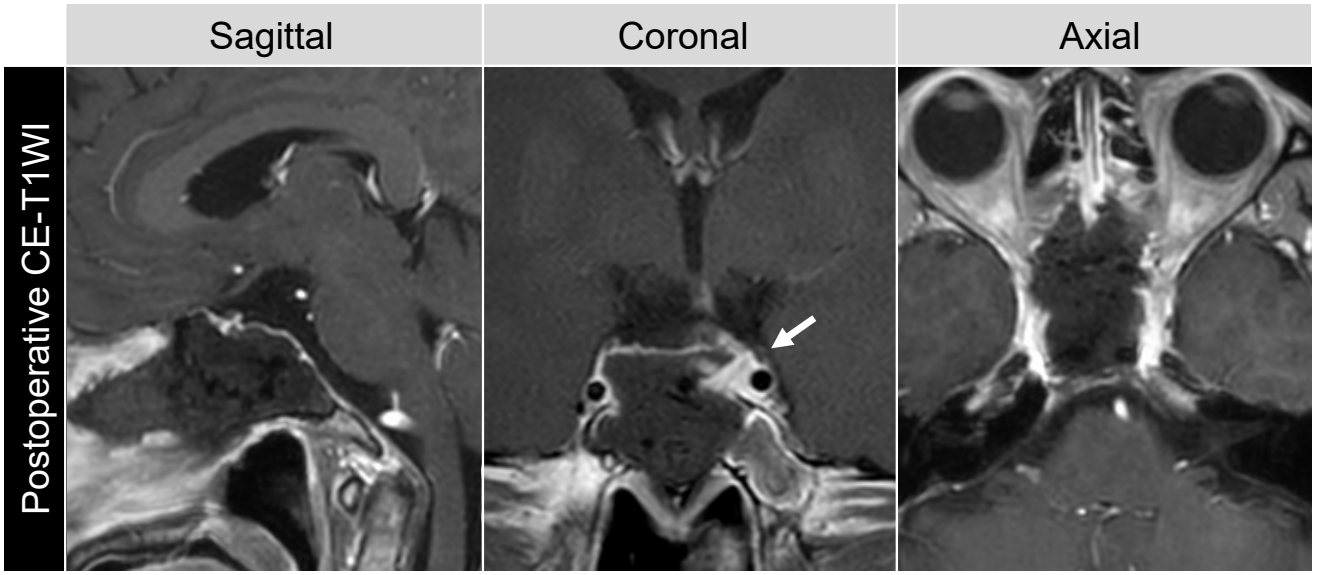


Fig. 4