

# Incidentally detected sellar spine in a patient with Cushing's syndrome: a case report

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## Abstract

Sellar spine, a bony spur extending anteriorly from the dorsum sellae, is a very rare anatomical variant. Several hypotheses regarding its etiology have been proposed, including the strongly supported theory of a cephalic ossified notochordal remnant. Sellar spine is usually detected incidentally in patients who have no definite symptoms, but several cases have reportedly accompanied endocrinopathies such as precocious puberty, hypopituitarism, or galactorrhea/oligomenorrhea. However, no published reports have described sellar spine in a patient with Cushing's syndrome. We herein report a case of sellar spine detected during the evaluation of Cushing's disease in a 29-year-old woman who underwent inferior petrosal sinus sampling, computed tomography, magnetic resonance imaging, and exploratory surgery. There was no evidence of a pituitary microadenoma, but a sellar spine was present in the operative field. Thus, the sellar spine might have caused Cushing's syndrome in this case, although the exact mechanism is unknown.

## Keywords

Sellar spine, Cushing's syndrome, inferior petrosal sinus sampling, pituitary adenoma, exploratory surgery, sellar magnetic resonance imaging

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## Introduction

Cushing's syndrome is a disorder caused by high cortisol levels. Its causes may be classified into adrenocorticotropic hormone (ACTH)-dependent, ACTH-independent, or iatrogenic. The most common cause of ACTH-dependent Cushing's syndrome is pituitary oversecretion of ACTH (Cushing's disease). Almost all patients with Cushing's disease have a pituitary adenoma, which is more likely to be a microadenoma than a macroadenoma, and the tumor is not always visible on imaging because of its small size and similar signal and enhancement pattern to that of the normal gland.<sup>1</sup> When the origin of hypercortisolism is uncertain, bilateral inferior petrosal sinus sampling (BIPSS) is performed. This is a highly accurate, minimally invasive procedure used to diagnose the pituitary source of ACTH oversecretion.<sup>2,3</sup>

Sellar spine, a rare anatomical variant, is a bony spur extending anteriorly from the dorsum sellae. One strongly supported theory is that it is a cephalic ossified notochordal remnant; however, its etiology remains unclear. Most cases of sellar spine are incidentally detected based on radiologic findings, but several studies have described sellar spine and concurrent endocrinopathies or neuro-ophthalmological

manifestations such as hypopituitarism or precocious puberty.<sup>4-11</sup> To the best of our knowledge, however, no reports have described sellar spine in a patient with Cushing's syndrome. We herein present a case of incidentally detected sellar spine mimicking a pituitary adenoma on radiologic examinations during the evaluation of a patient with Cushing's syndrome.

## Case report

A 29-year-old woman was admitted to our hospital for the evaluation of longstanding general weakness and recent body weight gain. She had a 2-year history of progressive hirsutism, a buffalo hump, and irregular menstruation, and Cushing's syndrome was suspected. The results of her blood and urinary tests were compatible with ACTH-dependent Cushing's syndrome (Table 1). Gadolinium-enhanced sellar dynamic magnetic resonance imaging (MRI), contrast-enhanced abdominal computed tomography (CT), and a high-dose dexamethasone suppression test were performed to determine the origin of ACTH oversecretion. Plasma cortisol was not suppressed after the high-dose dexamethasone suppression test, and there was no evidence of adrenal tumors or hyperplasia on abdominal CT. Pre-contrast sagittal and coronal

**Table 1.** Results of preoperative laboratory tests.

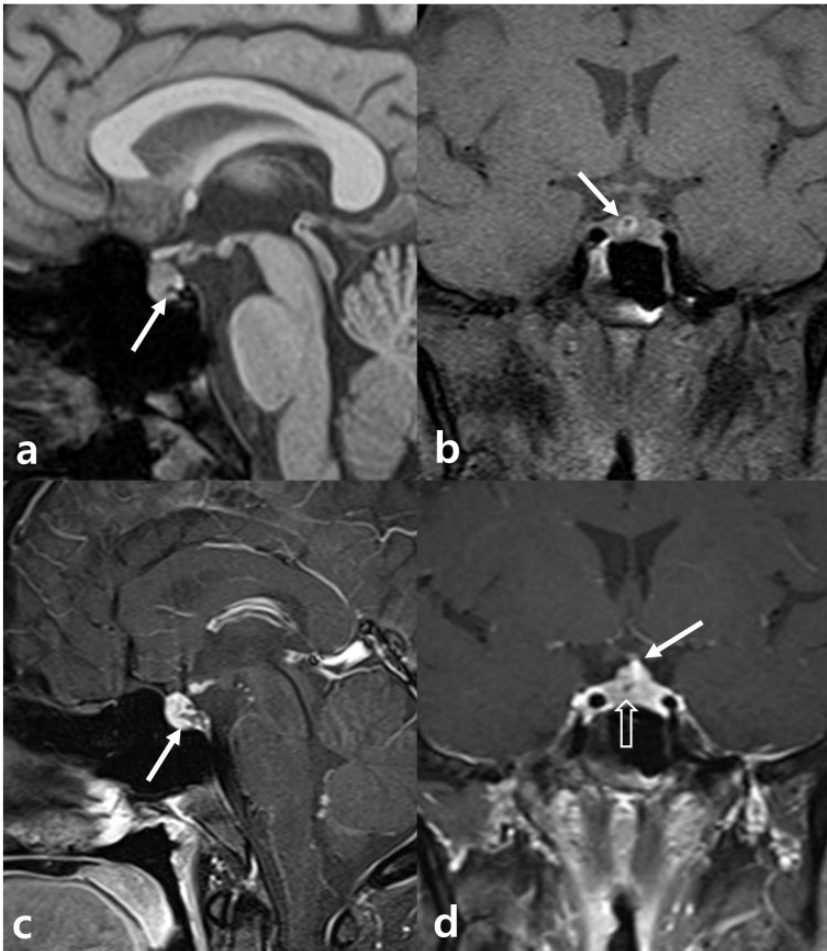
	Value	Reference range
Plasma ACTH (pg/mL)	26.4	0–60
Plasma cortisol (µg/dL)	15.7	3–23
24-hour urinary free cortisol (µg/dL)	1304	4.3–176
Overnight 1-mg dexamethasone suppression test		
Plasma cortisol (µg/dL)	10.9	(normal, <1.8)
Low-dose 0.5-mg dexamethasone suppression test		
Plasma cortisol (µg/dL)	21.2	(normal, <1.8)
High-dose 8-mg dexamethasone suppression test*		
Plasma cortisol (µg/dL)	18.9	

ACTH, adrenocorticotropic hormone.

\*Reduction in plasma cortisol by >50% overnight is consistent with Cushing's disease.

T1-weighted images showed a rod-like hypointense lesion anteriorly protruding from the dorsum sellae within the right posterior pituitary bright spot of the pituitary gland. The pituitary stalk was displaced to the left, and the pituitary gland was deformed by this lesion (Figure 1).

Although no apparent contrast enhancement was observed in the T1 hypointense lesion on sellar dynamic gadolinium-enhanced MRI, the neurosurgeons and radiologists initially determined that the lesion was a possible pituitary adenoma. The pituitary gland was 10 mm tall, which



**Figure 1.** (a) Sagittal non-enhanced fat-suppressed T1-weighted image (TIWI) showing the sellar spine passing through and disrupting the posterior lobe of the pituitary gland, manifesting as a bright signal of the posterior portion of the pituitary gland. (b) Coronal non-enhanced fat-suppressed TIWI showing the sellar spine, which appears as a black dot, within a high-signal rim caused by the posterior pituitary bright spot. (c) Sagittal contrast-enhanced fat-suppressed TIWI showing an osseous spine (arrow), which is isointense to the dorsum sellae, protruding into the pituitary fossa, and causing upward displacement and globular deformation of the pituitary gland (height, 10 mm). (d) Coronal contrast-enhanced fat-suppressed TIWI showing slight left-sided deviation of the pituitary stalk (arrow) by the sellar spine (arrowhead).

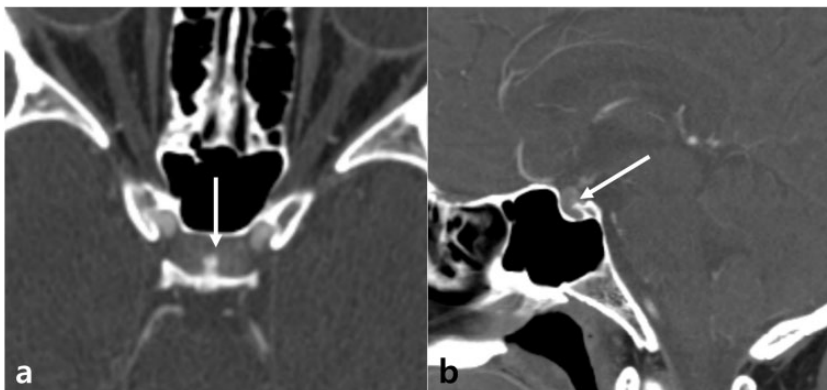
was within the normal range for the patient's age. BIPSS was performed because the dexamethasone suppression tests and sellar MRI showed equivocal results for confirming the etiology of Cushing's syndrome. The bilateral inferior petrosal sinuses and peripheral ACTH concentrations were assessed before and 3, 5, and 10 minutes after intravenous bolus injection of desmopressin, and the result of BIPSS indicated the possible presence of an ACTH-secreting adenoma in the right side of the pituitary gland (Table 2). Based on the results of MRI and BIPSS, we decided to perform surgery for treatment. A preoperative CT scan revealed that the lesion in

the posterior pituitary fossa was a 5-mm-long osseous spur arising from the dorsum sellae, which was indicative of a sellar spine, not a pituitary adenoma (Figure 2). However, the patient underwent neuronavigation-guided endoscopic endonasal transsphenoidal adenectomy for exploration of a radiologically imperceptible microadenoma. No distinct pituitary adenoma was noted; the only abnormality in the operative field was the sellar spine. Therefore, the surgeon resected the most tumorous part of the pituitary gland with the surrounding normal adeno-hypophyseal tissue. Pathologic examination of the biopsied tissue showed a tiny Rathke's cleft cyst

**Table 2.** Results of bilateral inferior petrosal sinus sampling.

	Basal	3 minutes	5 minutes	10 minutes
Right IPS (pg/mL)	43.6	93.4	85.9	50.2
Left IPS (pg/mL)	25.2	22.3	27.3	22.9
Periphery (pg/mL)	20.5	19.8	30.9	30.7
Central-to-peripheral ratio of ACTH concentration (IPS/periphery $\geq 2$ )				
Right	2.1			
Left	1.2			
Lateralization of pituitary microadenoma (gradient $\geq 1.4$ )				
Right IPS/periphery (3 minutes)	4.7			

ACTH, adrenocorticotrophic hormone; IPS, inferior petrosal sinus.



**Figure 2.** (a) and (b) Axial and sagittal contrast-enhanced computed tomography images showing an osseous spine (arrow) arising in the midline of the dorsum sellae and protruding into the sella.

with mild disruption of the acinar structure and reticulin network but no evidence of an ACTH-secreting adenoma. Immunohistochemical staining for ACTH of the specimen was negative. One month after the surgery, the patient's morning serum cortisol level measured 21.7  $\mu\text{g}/\text{dL}$ ; however, the follow-up laboratory test did not show complete remission of Cushing's syndrome (Table 3). Nevertheless, the patient's symptoms associated with Cushing's syndrome became less severe, and she was therefore planned to undergo regular follow-up.

Institutional review board approval was not obtained for this study because our institution does not require such approval for a single case report. Written informed consent was obtained from the patient for publication of these data.

## Discussion

The incidence of sellar spine is very low, occurring in approximately 1 in every 5000 to 8000 people.<sup>11,12</sup> The etiology of sellar spine remains unclear; hypotheses include an ossified cephalic notochordal remnant, ossified vascular channel, and dural fold.<sup>13</sup> During normal embryonic development, the cephalic tip of the notochordal segment regresses and finally separates from the posterior lobe of the pituitary gland. In cases of sellar spine, however, the cephalic notochordal segment remains and subsequently ossifies. This is one of the most reliable hypotheses of the etiology of sellar spine.

Although sellar spine is considered an incidental finding, several published studies have focused on the association of this anomaly with concurrent endocrinopathies (Table 4). Eguchi et al.<sup>10</sup> described a patient with simultaneous amenorrhea, sellar spine, and prolactinoma but offered no clear explanation about the relationship between the tumor and sellar spine. Cases of sellar spine accompanied by hypopituitarism have been reported more frequently.<sup>4,8,9</sup> One report explained the possible mechanism of hypopituitarism based on the sellar spine passing through and destroying the pituitary gland.<sup>4</sup> In contrast, few cases associated with pituitary hyperfunction, as in the present case, have been reported. Hosokawa et al.<sup>5</sup> reported a case of precocious puberty in an 8-year-old girl with an enlarged pituitary gland and showed a subsequent pressure effect that could have led to detection of the ossified bony spur. Similarly, Matsumoto et al.<sup>7</sup> reported sellar spine in a 15-year-old boy who showed elevated serum follicle-stimulating hormone and luteinizing hormone levels. It was difficult to determine the exact etiology of the patients' symptoms in these reports, but the authors proposed the hypothesis that glandular deformation or stalk deviation caused by sellar spine might have been associated with the elevated pituitary hormones.

In our case, sellar spine was detected in a patient with Cushing's syndrome; such a case has not been previously reported. There is always a possibility of false ACTH results depending on various

**Table 3.** Results of postoperative laboratory tests.

	Postop 1 month	Postop 3 months	Postop 4 months
Plasma ACTH (0–60 $\text{pg}/\text{mL}$ )	47	48	74
Plasma cortisol (3–23 $\mu\text{g}/\text{dL}$ )	21.7	13.2	25.6
24-hour urinary free cortisol (4.3–176 $\mu\text{g}/\text{dL}$ )	160.0	1457.5	124.2

Postop, postoperative; ACTH, adrenocorticotropic hormone.

**Table 4.** Summary of published cases of sellar spine with endocrine or neuro-ophthalmologic symptoms.

Reference	Age, years	Sex	Symptoms	Hormone study	Size of sellar spine	Neoplastic sellar mass	Modality
LaMasters et al. <sup>11</sup>	24	F	Amenorrhea	Normal	5 mm	Not detected	CT
Jacinto et al. <sup>8</sup>	40	M	Weight gain	Partial hypopituitarism	Not measured	Not detected	CT
Eguchi et al. <sup>10</sup>	21	F	Galactorrhea, oligomenorrhea	Hyperprolactinemia	Not measured	Pituitary adenoma	CT, MRI
Abs et al. <sup>4</sup>	39	M	Pubertal delay	Hypopituitarism	9 mm	Not detected	CT, MRI
Matsumoto et al. <sup>7</sup>	15	M	Excessive height	Elevated serum LH and FSH	4 mm	Not detected	CT, MRI
Zucchini et al. <sup>9</sup>	5.4	F	Short stature	Isolated GH deficiency	Not measured	Not detected	MRI
Chivukula et al. <sup>6</sup>	6.7	F	Short stature	Isolated GH deficiency	Not measured	Not detected	MRI
	19	F	Bitemporal visual defects, menstrual irregularity	Normal	Not measured	Not detected	CT, MRI
Hosokawa et al. <sup>5</sup>	8	F	Precocious puberty	Elevated serum LH in LH-releasing hormone loading test	3.8 mm	Not detected	CT, MRI
Present case	29	F	Weight gain	Elevated ACTH and cortisol	5 mm	Not detected	CT, MRI

F, female; M, male; ACTH, adrenocorticotropic hormone; CT, computed tomography; FSH, follicle-stimulating hormone; GH, growth hormone; LH, luteinizing hormone; MRI, magnetic resonance imaging.



techniques and equipment types,<sup>14</sup> and we should also be aware of the possibility of false-positive results of BIPSS. We did not perform a fine-cut multi-slice CT scan to scrutinize the presence of a microadenoma before surgery, which was one limitation in this case; however, there was no evidence of a pituitary microadenoma on high-quality dynamic MRI. The only abnormality in the operative field was the sellar spine. The neurosurgeon did not resect the sellar spine because of the limited approach via endoscopic endonasal transsphenoidal adenectomy and lack of evidence with regard to concurrent sellar spine and endocrinopathy or the surgical management of it. The patient's postoperative laboratory test results showed no improvement. Therefore, we hypothesized that the Cushing's syndrome in the present case might have been associated with the unresected sellar spine, although the exact mechanism is unknown. Deviation of the pituitary stalk and deformation of the pituitary gland by the sellar spine demonstrated on MRI were similar to the findings in previously reported cases of pituitary hyperfunction. In experimental models, resultant stimulation of the hypothalamic-pituitary-adrenal axis within the pituitary stalk by trauma or an enlarged sella turcica and a subsequent increase in the plasma ACTH level have been proposed.<sup>15</sup> Likewise, the patient's plasma ACTH level did not show a normal circadian rhythm, while the levels of other hormones within the pituitary axis (human growth hormone, follicle-stimulating hormone, luteinizing hormone, thyroid-stimulating hormone, prolactin, and cortisol) were in the normal range in this case. Further research efforts are needed to determine the exact pathomechanism underlying the association between sellar spine and possible pituitary hyperfunction.

A few published reports described MRI findings of sellar spine.<sup>5,7,16–18</sup> On T1- and T2-weighted MRI, most sellar spines were

clearly shown as a hypointense structure protruding into the pituitary posterior lobe. On coronal T1-weighted images, sellar spine typically appears as a black dot surrounded by a hyperintense rim caused by the posterior pituitary bright spot. In addition to these signal intensities, the connection with the dorsum sellae and the lack of contrast enhancement allow the clinician to easily rule out a pituitary adenoma or an intrapituitary cystic lesion. Deviation of the pituitary stalk, deformation of the pituitary gland, and the relation of the optic chiasm can also be evaluated by MRI.

One report described successful surgical resection of sellar spine in a patient with headache and bitemporal visual field defects, leading to resolution of these symptoms.<sup>6</sup> However, because of the rarity of symptomatic sellar spine, no definite treatment guideline exists.

In conclusion, we have herein reported a unique case in which sellar spine was incidentally detected during imaging evaluation of a patient who was strongly suspected to have ACTH-dependent Cushing's syndrome and was initially misdiagnosed as having a pituitary adenoma on MRI. Radiologists and neurosurgeons should be aware of sellar spine, a rare anatomical variant, and be familiar with its imaging features to avoid unnecessary invasive surgery or delayed diagnosis.

### Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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