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Solitary Fibrous Tumor of the Sella Mimicking Pituitary Adenoma: An Uncommon Tumor in a Rare Location—A Case Report

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Abstract Solitary fibrous tumor (SFT) is rarely located in the central nervous system, and sella turcica involvement was reported in only two patients. We report the case of a 28-year-old man with a SFT of the sella turcica mimicking a pituitary nonfunctioning macroadenoma. He presented with optic nerve compression caused by a heterogeneous tumor located in the sellar and suprasellar area. At surgery, the tumor was hard and infiltrated the sellar diaphragm, so that resection resulted in a cerebrospinal fluid fistula. His postoperative course was also complicated by complete central diabetes insipidus, hypopituitarism, and two episodes of meningitis. After surgical resection, the diagnosis of SFT was reached on the basis of histological and immunohistochemical studies. He was discharged after 49 days. Ten months after surgery, he was clinically well, and magnetic resonance images showed no evidence of residual or recurrent tumor. SFT should be considered in the differential diagnosis of sellar and parasellar tumors.

Keywords central nervous system · diabetes insipidus · optic chiasm · sella turcica · solitary fibrous tumor

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Introduction

Solitary fibrous tumors (SFT) are spindle-cell neoplasms that histologically and immunohistochemically resemble pleural-based SFTs, and are characterized immunohistochemically by CD34, vimentin, and bcl-2 reactivity [1–4]. They are rare in the central nervous system (CNS). A MEDLINE search in July 2008 for "solitary fibrous tumor" and "central nervous system" identified 30 articles describing 65 patients with non-spinal SFT of the CNS [1–31]. Involvement of the pituitary fossa was described only in two other patients [13, 22]. A SFT in the parasellar region was also described [18]. We report a case of SFT of the pituitary fossa and suprasellar area that presented with compression of the optic chiasm, mimicking a nonfunctioning pituitary macroadenoma.

Case Report

History and Examination A 28-year-old white man presented with visual disturbance of 2 weeks duration. Clinical examination disclosed a well-developed man with no other medical complaints, except for nocturia once or twice a night in the last few years. His weight and height were, respectively, 84 kg and 1.80 m. Fasting serum aspartate

aminotransferase, alanine aminotransferase, creatinine, urea nitrogen, glucose, alkaline phosphatase, calcium, and phosphorus levels, as well as blood hemoglobin and hematocrit, were normal. The total 24-h urine output volume varied from 2.5 to 3.5 L on three different days. Urine had a density of 1,007. Results of serum hormone levels are shown in Table 1.

Visual fields were evaluated by perimetry, disclosing right temporal hemianopsia and left inferior temporal quadrantanopsia. Radiological studies showed a heterogeneous mass inside and above the pituitary fossa with involvement of the optic chiasm; there was no calcification (Figs. 1, 2, 3, 4, and 5). The patient was diagnosed with a nonfunctioning pituitary macroadenoma.

Operation Endoscopic transnasal transsphenoidal microsurgery was performed, and a hard, fibrous tumor was removed. The sellar diaphragm was involved by the lesion, and resection resulted in a cerebrospinal fluid (CSF) fistula. The sphenoid sinus was packed with fat and biological glue, and a lumbar drainage via percutaneous catheter was performed.

Histopathological Findings The specimen contained a small amount of adenohypophysial parenchyma and a highly cellular spindle-cell proliferation with a dense, hyalinized collagenous stroma and dilated vascular spaces, some showing a staghorn-like appearance. Areas of cellular pleomorphism and increased cellularity were present, but mitoses were not identified (Fig. 6A). Immunohistochemistry showed strong immunoreactivity for vimentin (Fig. 6B), CD34 (Fig. 6C), and BcL-2; vessel walls stained for factor VIII. No specific staining for GFAP or S100 protein was found, and the tumor was negative for keratins, EMA, smooth muscle actin, and muscle-specific actin as well as myoD1, melanoma cocktail, and CD99. Cyclin D1 was localized in only a few scattered nuclei (less than 5% in the most positive area) and MIB-1 labeling had a similar pattern with an index of <5%. Stains for p53 were weakly positive in the majority of tumor nuclei. The features were considered to be those of SFT.

Postoperative Course He developed hypopituitarism, as shown in Table 1, and complete central diabetes insipidus. Glucocorticoids, DDAVP, levothyroxine, and testosterone were administered. His postoperative course was also complicated by two episodes of meningitis, and he was discharged 49 days after the surgery. Two months after the surgery, his eyesight had improved markedly, and the total 24-h urine output volume varied from 1.5 to 2.0 L, while he was treated with intranasal DDAVP. The patient was clinically well, and MRI of the brain performed 10 months after the surgery showed no evidence of tumor.

Discussion

Pituitary adenomas comprise approximately 90% of sellar masses [32]. In a large series of 911 patients who underwent transsphenoidal surgery for sellar masses, 83 had lesions other than pituitary adenoma, including cysts, other primary tumors such as craniopharyngioma, chordoma, meningioma, or lymphoma, metastatic carcinoma, inflammatory lesions such as granulomas, infection or lymphocytic hypophysitis, and mucocele or aneurysm [32]. SFT is not usually included in this differential diagnosis.

Pituitary adenomas can manifest excess hormone production, visual disturbances, or hypopituitarism. Anterior pituitary failure, visual loss, and visual field abnormalities do not distinguish pituitary adenoma from a non-adenomatous lesion [32, 33]. In contrast, certain clinical findings, such as diabetes insipidus and cranial nerve palsies, are more commonly associated with non-adenomatous sellar masses. At presentation, our patient had mild polyuria and nocturia that he attributed to a habit

Table 1 Serum hormone levels

	Before surgery	After surgery	Normal values
Free T4 (ng/mL)	1.4	0.7	0.7–1.9
TSH (mU/L)	1.07	0.02	0.3–4.5
FSH (mU/mL)	3.6		2–17.5
LH (mU/mL)	3.1		1.4–7.7
Testosterone (ng/Dl)	417	<5	132–813
Prolactin (ng/Ml)	3.9		2–14.5
Cortisol (μg/Dl)	10	<1	5–20
ACTH (pg/Ml)	21.8	<10	10–52
IGF-1 (ng/Ml).	187		117–321
PTH (pg/mL)	31		16–65

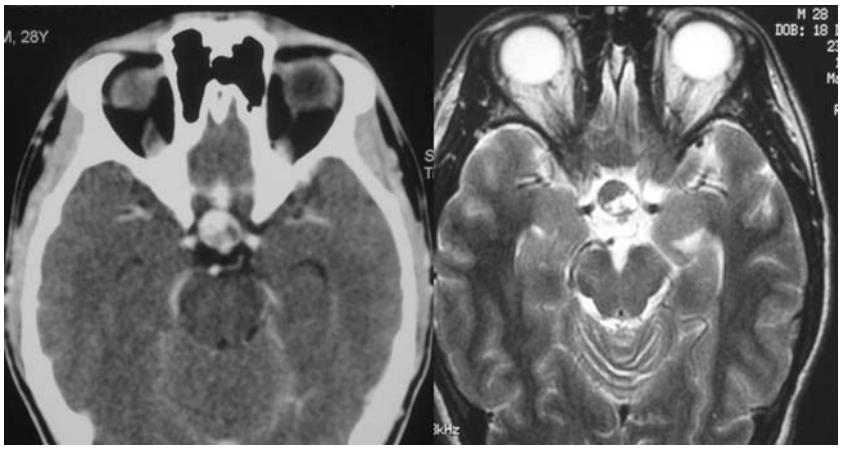

Fig. 1 Contrast-enhanced computed tomography (*left*), and magnetic resonance T2WI axial (*right*) images, at the suprasellar level, showing tumor above the pituitary gland with heterogeneous contrast enhancement and T2WI signal in the same fashion. There is no calcification

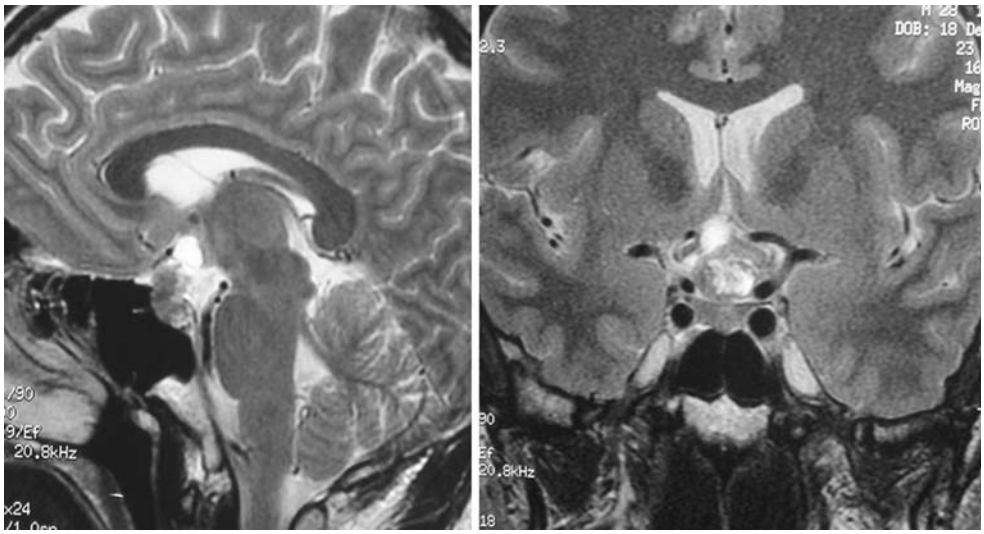

Fig. 2 MRI sagittal (*left*) and coronal (*right*) T2WI views. In the sagittal view, there is a mass inside the pituitary fossa, with compressed hypophyseal tissue seen in the coronal view

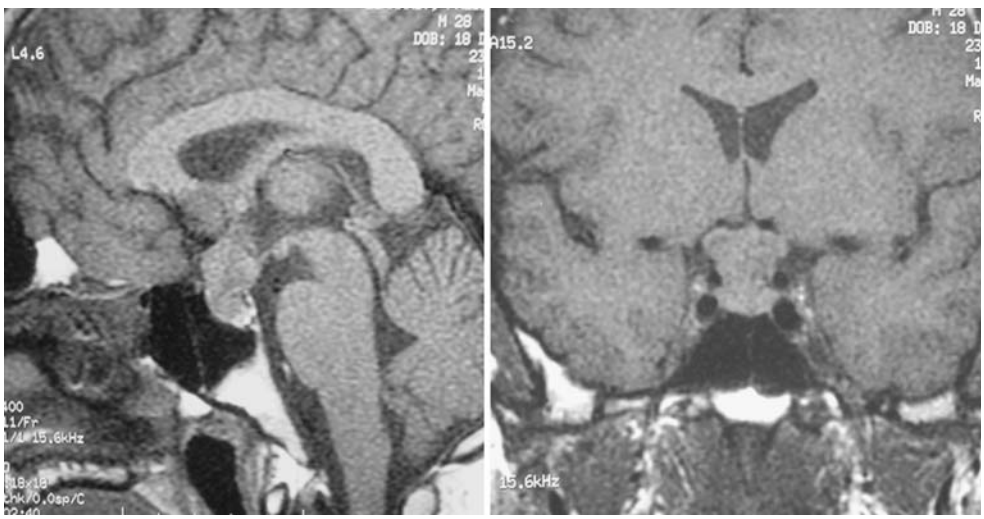

Fig. 3 MRI sagittal (*left*) and coronal (*right*) T1WI views. Note involvement of the optic chiasm

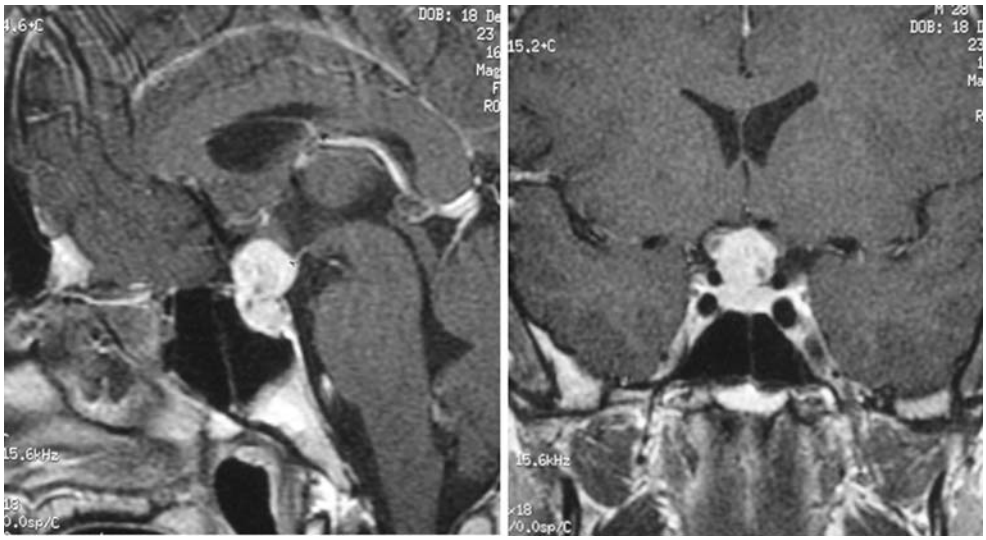

Fig. 4 MRI sagittal (*left*) and coronal (*right*) T1WI views with IV gadolinium showing strong heterogeneous contrast enhancement

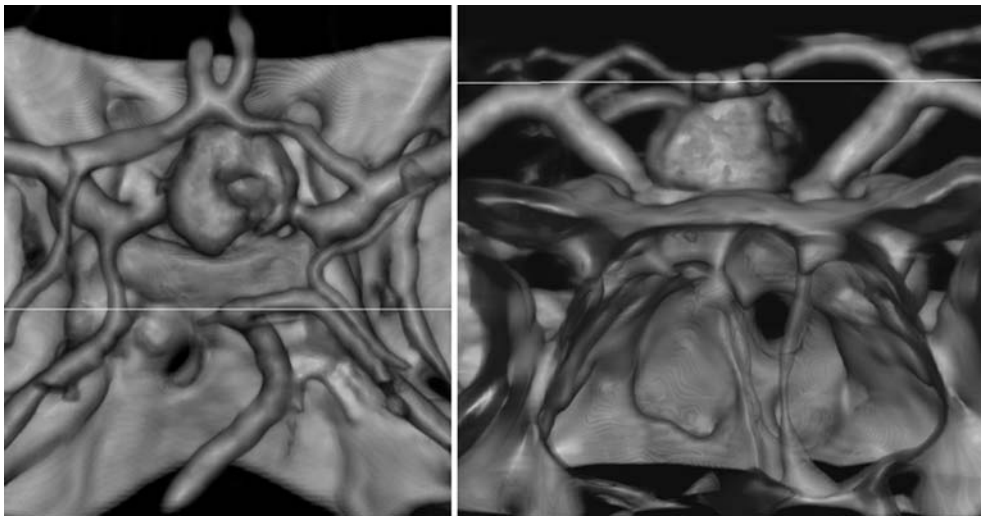

Fig. 5 Multislice contrast-enhanced computed tomography rendering images of the tumor in the superior (*left*) and the anterior (*right*) views to surgery plan

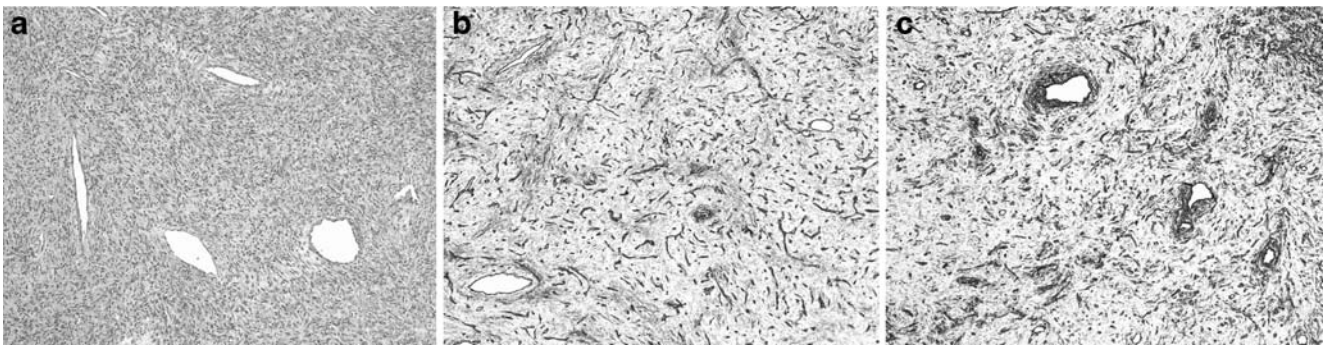

Fig. 6 Spindle-cell tumor consistent with solitary fibrous tumor. **a** Highly cellular spindle-cell proliferation with a dense, hyalinized collagenous stroma and dilated vascular spaces, some showing a staghorn-like appearance. Areas of cellular pleomorphism and increased cellularity were present, but mitoses were not identified. (H&E ×50). **b** Strong immunoreactivity for CD34 (×50). **c** Strong immunoreactivity for vimentin (×50)

of drinking tea at night. After surgery, total 24-h urine volume decreased and his nocturia resolved, while treated with DDAVP. It is possible that he had mild partial central diabetes insipidus before the surgery, but unfortunately, he was not tested to prove that hypothesis.

The initial diagnostic impression was of a nonfunctioning pituitary macroadenoma. The tumor was heterogeneous in T1-weighted images (T1WI), and T2-weighted images (T2WI), and after gadolinium, and in contrast-enhanced computed tomography, as shown in Figs. 1–4. The heterogeneity could have been due to cystic degeneration and hemorrhage that are not uncommon in larger pituitary adenomas [34]. However, histopathology confirmed this lesion to be a SFT.

Two SFTs involving the pituitary fossa have been previously described. In 2003, Cassarino et al. described a patient with an aggressive tumor invading the sphenoid sinus, cavernous sinus, and pituitary fossa, extending into the nasopharynx; this lesion was only partially resectable because of extensive bony invasion and encasement of the carotid artery [22]. Their first impression was of a malignant tumor originating in the pituitary fossa. In 2005, Pakasa et al. reported a patient with visual field loss in the left eye and a lesion in the pituitary fossa that was radiologically consistent with a pituitary adenoma with suprasellar extension. The tumor resection was incomplete, and it recurred twice in the same location within 3 years [13]. Both of these were shown to be SFT.

The origin of these lesions is not known, SFT in the CNS, although rare, are usually well-defined, solid, dura-based masses, occurring in much at the same sites as do meningioma [3, 20]. We speculate that SFT of the sella could have had its origin in the dural-based fibroblast or dendritic cells of the sellar diaphragm or in the mesenchyma of the vasculature of the pituitary stalk [3]. In this regard, they join a group of rare mesenchymal lesions of the pituitary including glomangioma [35] and other stromal tumors [36] that can mimic pituitary adenoma.

Unlike the other two reported cases, our patient has had a benign course, in accordance with the histopathological grade of the tumor. There was no evidence of invasion except for the sellar diaphragm that could be the site of origin of the tumor. Surgical resection was complete, and there was no evidence of tumor residua or recurrence in 10 months of follow-up surveillance.

We conclude that SFT can mimic pituitary adenoma and should be considered in the differential diagnosis of sellar and suprasellar masses.

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