

A primary atypical solitary fibrous tumor of the sella mimicking nonfunctional pituitary adenoma: a case report

Weining Yin · Chunxiao Ma · Jian Wu · Bowen Cai · Chao You

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Abstract We reported a 32-year-old male with a sellar solitary fibrous tumor who presented with headache and affliction in the left eye. Serum biochemical examination revealed hypoglycemia. The tumor was assumed to be a nonfunctional pituitary adenoma preoperatively. A subtotal resection of the tumor was performed. Immunohistochemically, atypical solitary fibrous tumor was established. The residual tumor had no progression or distant metastasis at a 44-month follow-up after gamma-knife stereoradiotherapy.

Keywords Solitary fibrous tumor · Sella · Immunohistochemistry

Abbreviations

SFT Solitary fibrous tumor
HPC Hemangiopericytoma
EMA Epithelial membrane antigen

Introduction

Solitary fibrous tumors (SFTs) of the meninges were first described by Carneiro and co-workers in 1996 [3]. Conventional SFT could be defined by alternating hypercellular and hypocellular areas and diffuse immunoreactivity for CD34 antigen [3]. The recent study, however, showed that the SFT had a wider spectrum including hemangiopericytoma (HPC) which was conventionally a specific entity [7]. The lesion located in the sellar region is

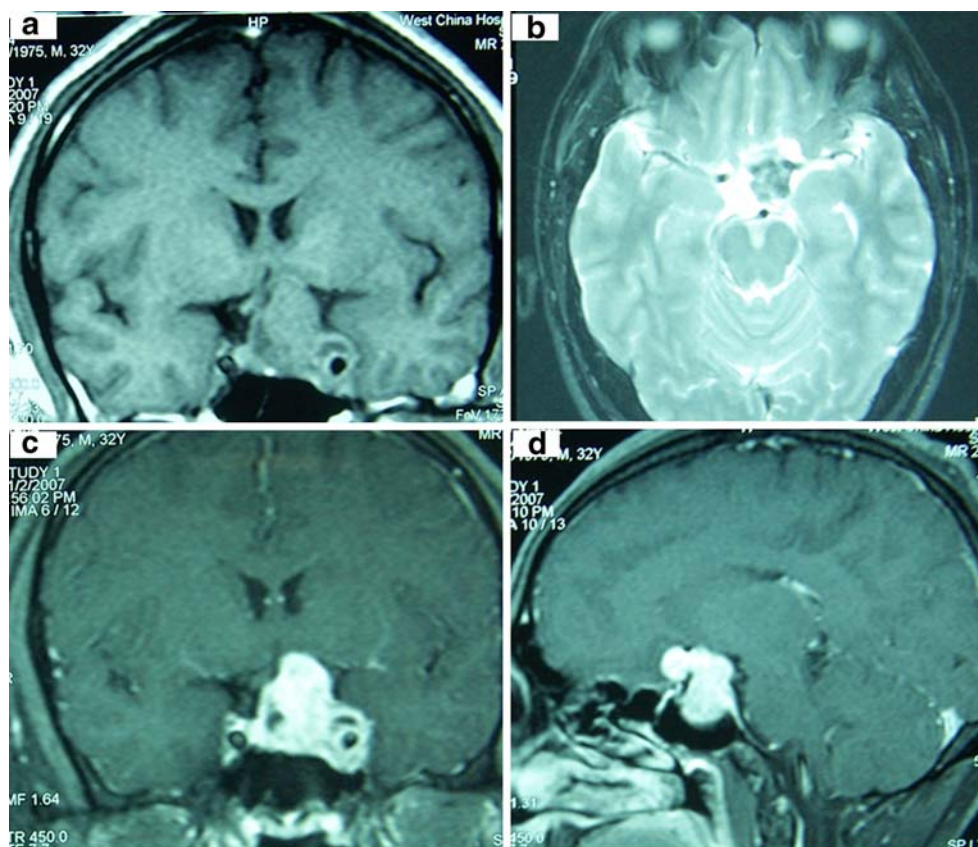
uncommon and is seldom diagnosed preoperatively. To our knowledge, only 1 case of sellar SFT and five cases of sellar hemangiopericytoma have been reported in recent English language literatures [6, 8, 10, 11, 13, 16]. We report the experience of a case and discuss the diagnosis and treatment of the tumor at this unusual location.

Case report

A 32-year-old man was admitted to the Department of Neurosurgery with a 2-month history of intermittent headache, ophthalmalgia, intolerance of light, and progressively decreasing visual acuity. On admission, the ophthalmologic examination disclosed moderate chemosis and conjunctive congestion, atrophy of optic papilla, impairment of the visual field with complete temporal hemianopia of the left eye. The corneal reflex was normal and the reaction to light was prompt on the left side. The direct or indirect light reflex was normal. General examination including biochemistry, chest X-ray, and electrocardiogram of the patient was otherwise unremarkable. Endocrine investigations disclosed normal hormone secretion of the pituitary. Serum biochemical examination revealed hypoglycemia (3.2 mmol/l, normal range: 3.9–5.9). Magnetic resonance imaging (MRI) showed a $3.2 \times 2.6 \times 2 \text{ cm}^3$ lobulated mass located in the pituitary fossa. The left optical nerve and chiasma were compressed by the lesion. The lesion exhibited isointensity with respect to gray matter on the T1-weighted scans (Fig. 1a) and hyperintensity with focal hypointensity on the T2-weighted sequences (Fig. 1b). Gadolinium enhanced T1-weighted images demonstrated homogeneous enhancement of the tumor (Fig. 1c, d). Based on the clinical and endocrine investigation, the tumor was assumed to be a nonfunctional pituitary

W. Yin · C. Ma · J. Wu · B. Cai · C. You (✉)
Department of Neurosurgery, West China Hospital,
Sichuan University,
Chengdu, China
e-mail: youchao5837@163.com

Fig. 1 **a** Magnetic resonance imaging (MRI) revealed isointensity with respect to gray matter on T1-weighted scans. **b** Hyperintensity with focal hypointensity on T2-weighted sequences. **c** Axial contrast T1-weighted sequences demonstrated homogeneous enhancement of the tumor. **d** Sagittal contrast T1-weighted sequences demonstrated homogeneous enhancement of a lobular mass in the pituitary fossa



adenoma. Operatively, a highly vascular, red-tan and hard tumor was seen in the suprasellar region. The resection was difficult and caused torrential hemorrhage. A subtotal resection was performed. The histological examination demonstrated that the tumor consisted of monotonous hypercellular areas (Fig. 2a) and thin-walled ‘staghorn’ branching vessels (Fig. 2b) arranged in a collagenous background. Atypical cells were readily to be found (Fig. 3a). Clusters of spindle cells were surrounded by reticulin fibers (Fig. 3b). Immunohistochemical examina-

tion demonstrated positive for bcl-2 (Fig. 3c), CD99 (Fig. 3d) and vimentin whereas negative for S-100 protein and epithelial membrane antigen (EMA). The CD34 immunoreactivity was less intense. There was no immunopositive evidence for adenohypophysial hormones. According to the immunohistochemical examination, the diagnosis of atypical SFT was established. Postoperatively, the visual acuity in his right eye remains stable, but that in the left is still poor. One week later, gamma-knife stereoradiotherapy was performed. The postoperative course was uneventful.

Fig. 2 **a** The tumor consisted of monotonous hypercellular areas in a collagenous background (200 \times). **b** Thin-walled ‘staghorn’ branching vessels were focally evident (75 \times)

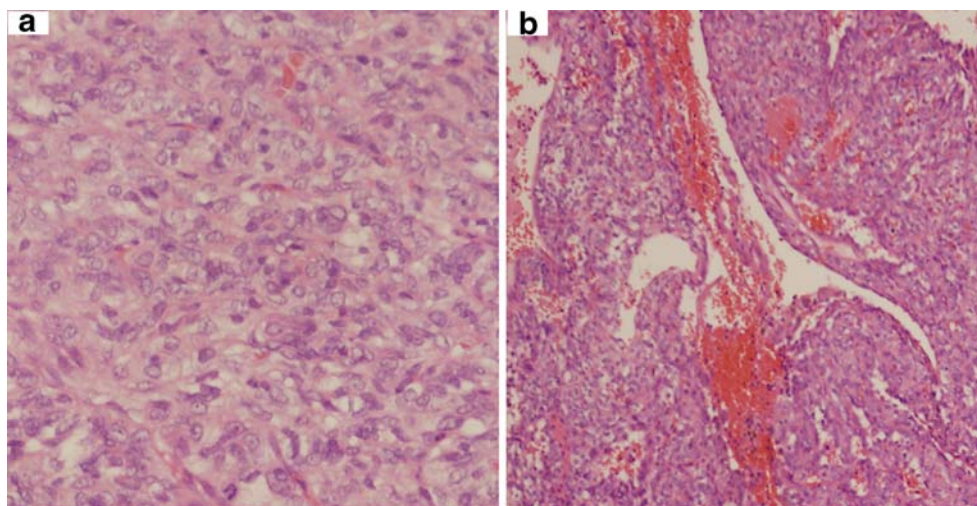
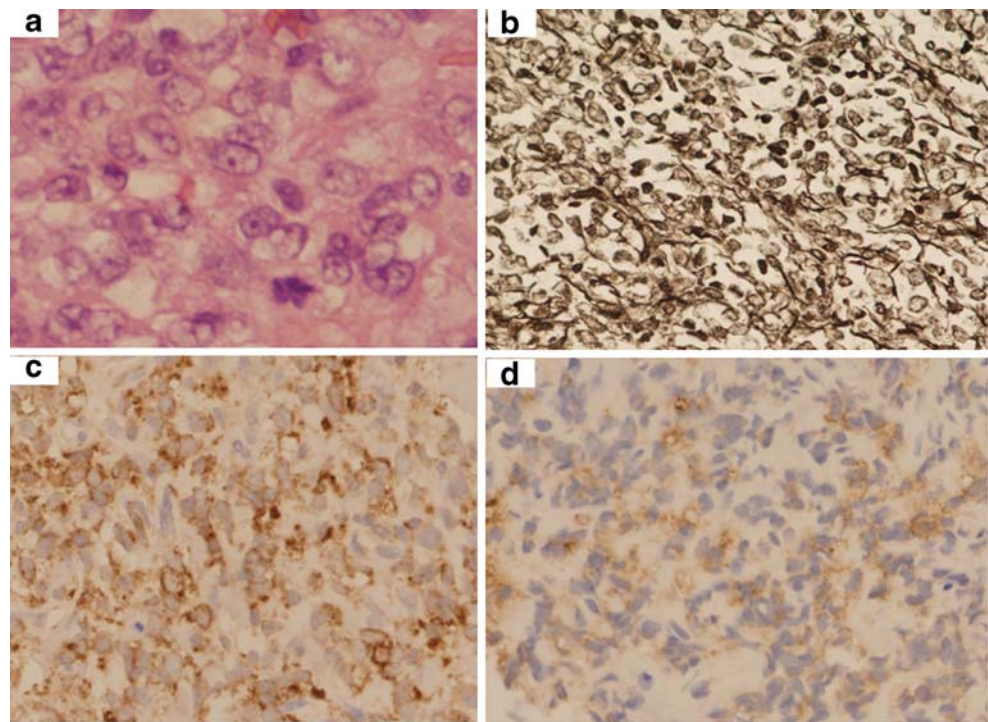


Fig. 3 **a** Atypical cells were readily to be found (600×). **b** Clusters of spindle cells were surrounded by reticulin fibers (300×). **c** Immunohistochemical examination demonstrated positive for bcl-2(100×). **d** Immunohistochemical examination demonstrated positive for CD99 (100×)



Blood glucose level was restored to normal (4.1 mmol/l, normal range: 3.9–5.9). The residual tumor had no progression or distant metastasis at a 44-month follow-up.

Discussion

Solitary fibrous tumors are rare spindle cell tumors, commonly arising from the visceral pleura [1]. However, these tumors have now been demonstrated in various locations outside the thoracic cavity [2]. On the basis of limited information, meningeal SFT mostly occurred in the posterior fossa (26%) and spine (25%) [4]. The lesion located in the sellar region is uncommon and is seldom diagnosed preoperatively. To our knowledge, only one case of sellar SFT and five cases of sellar hemangiopericytoma have been reported in recent English language literature [6, 8, 10, 11, 13, 16]. Clinically, the sellar SFT may present with local symptoms related to the tumor site. Local mass effects including visual disturbance, headache and radiographic abnormalities are clinically identical to pituitary adenoma. Endocrine examination disclosed normal or abnormal anterior pituitary hormone secretion. The sellar SFTs are prone to be diagnosed as pituitary adenoma preoperatively [6, 8, 10, 11, 13, 16]. The hypoglycemia symptom, as a paracarcinoma symptom, may be attributed to the secretion of insulin-like growth factor [9]. On MRI, the SFTs are usually isointense with normal brain parenchyma on the T1-weighted images and they had high or low signal intensity on the T2-weighted images. Homoge-

neous or heterogeneous enhancement of the tumor was demonstrated after contrast administration [6, 8, 10, 11, 13]. Radiologically, it is currently difficult to differentiate a sellar SFT from a pituitary adenoma or fibrous meningioma.

Histologically, the feature of SFTs exhibited alternative hypercellular and hypocellular region set in a collagenous background. A patternless pattern is frequently described for this lesion [3, 6]. Immunohistochemical examination showed strong and diffuse reactivity for CD34, bcl-2, vimentin and CD99, but negative for EMA and S-100 protein [6, 8, 10, 11, 13]. The histological and detailed immunohistological findings would exclude the diagnosis of meningioma, schwannoma, and pituitary adenoma [6, 8, 10, 11, 13, 16]. In our case, the SFT showed monotonous high cellularity and thin-walled ‘staghorn’ branching vessels histologically. The CD34 immunoreactivity was weak in cytoplasm. All of those features are not typical SFT but hemangiopericytoma (HPC)-like tumor. Some authors [7], however, proposed that SFT and HPC were a single entity and the HPC was a variant of SFT based on the absence of pericytic differentiation in most HPCs and the morphological and immunohistochemical similarities to SFTs. True HPCs correspond to entities such as myopericytoma, infantile myofibromatosis, and HPC-like lesions of the sinonasal tract showing myoid differentiation [7]. These lesions are the only ones that deserve to be called HPC. All other HPC-like lesions are best considered as a variant of SFT [7]. According to the above-mentioned criteria, all the five cases of sellar HPC should be included in the category of SFT.

The malignant histological features of the pleura could be applied to meningeal SFTs. These malignant features include high cellularity, increased mitotic index, necrosis areas, and nuclear pleomorphism [15]. It seems that there is no strict correlation between morphology and biology in SFTs [5]. The most important fact affecting prognosis was the extent of removal instead of the histological features [12]. Nevertheless, when the lesion cannot be totally resected, radiotherapy should be considered [8, 11, 13]. Nakahara et al. [14] reported a shrinkage of an occipital residual SFT at a 4-year follow-up after gamma-knife stereoradiotherapy. In our case, the residual tumor had no progression or distant metastasis, and the blood glucose level was restored to normal at a 44-month follow-up after gamma-knife stereoradiotherapy. This fact implied that postoperative gamma-knife stereoradiotherapy may be helpful to control the residual tumor of SFT.

The previous reports and the presented case taught us important new lessons regarding sellar lesions. First, SFT should be considered in the differential diagnosis of sellar masses. Second, the optimal way to manage a sellar SFT may be safe maximal surgical resection and postoperative radiotherapy.

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