

Case Report

A case of a malignant solitary fibrous tumor in the maxillary sinus extending into the intracranial space

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JapanE-mail: cooninh@zb4.so-net.ne.jpReceived: January 22nd, 2018Accepted: January 25nd, 2018Published: January 26th, 2018**Citation**Fujiwara-Tani R, Yamada M, Kitagawa S, Nakagawa H, Kuniyasu H. A case of a malignant solitary fibrous tumor in the maxillary sinus extending into the intracranial space. *Case Rep Rev Press*. 2018; 2(1): 84-86. doi: 10.28964/RevRepPress-2-113**Copyright**

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KEYWORDS: Solid fibrous tumor; Paranasal sinus; STAT6;**ABBREVIATIONS:** SFT: Solid Fibrous Tumor; CT: Computed Tomography; MRI: Magnetic Resonance Images; HE: Hematoxylin and Eosin; HPF: High Power Magnificent Field; BCL-2: B-cell Lymphoma 2; EMA: Epithelial Membrane Antigen; NCAM: Neural Cell Adhesion Molecule; SMA: Smooth Muscle Actin; EGR: Early Growth Response; ALDH: Aldehyde Dehydrogenase; MMP: Matrix-Metallo Proteinase.**INTRODUCTION**

A solitary fibrous tumor (SFT) arising from the sino-nasal region extending into the intracranial spaces is a rare tumor.¹ Histopathological diagnosis of a SFT is based on nuclear STAT6 expression by immunohistochemistry,² which is associated with formation of NAB2-STAT6 fusion gene.^{3,4} A malignant phenotype is histopathologically diagnosed by dense cellularity and frequent mitotic characteristics.⁵ We experienced a case of malignant SFT arising from maxillary sinus expanding into intracranial region. In the case, the nuclear STAT6 expression was a key finding for diagnosis.

CASE REPORT

A 67-year-old woman was admitted to Tokushu-kai Nozaki Hospital complaining of a headache. A head computed tomography (CT) scan revealed a solid tumor-measuring 5.5 cm in diameter-in the left maxillary sinus that had extended into the left inferior part of the brain (Figure 1A). The tumor appeared hypointense on T2-weighted magnetic resonance images (MRI) (Figure 1B). The patient subsequently underwent a craniotomy for tumor resection.

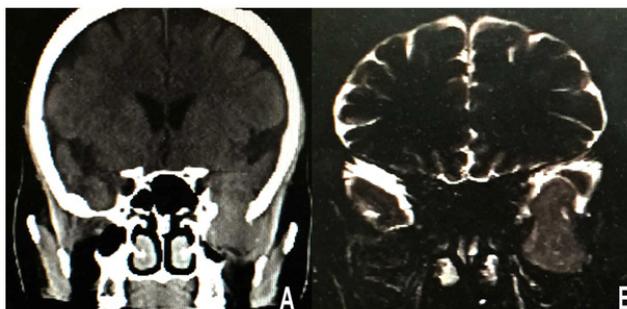


Figure 1: (A) Coronal view of the computed tomography image showing a solid tumor in the left maxillary sinus measuring 5.5 cm in diameter. The tumor had extended into the intracranial-extradural space of the left lower brain and compressed the lower surface of the left frontal lobe (B). The tumor was hypointense on T2-weighted magnetic resonance images (MRI).

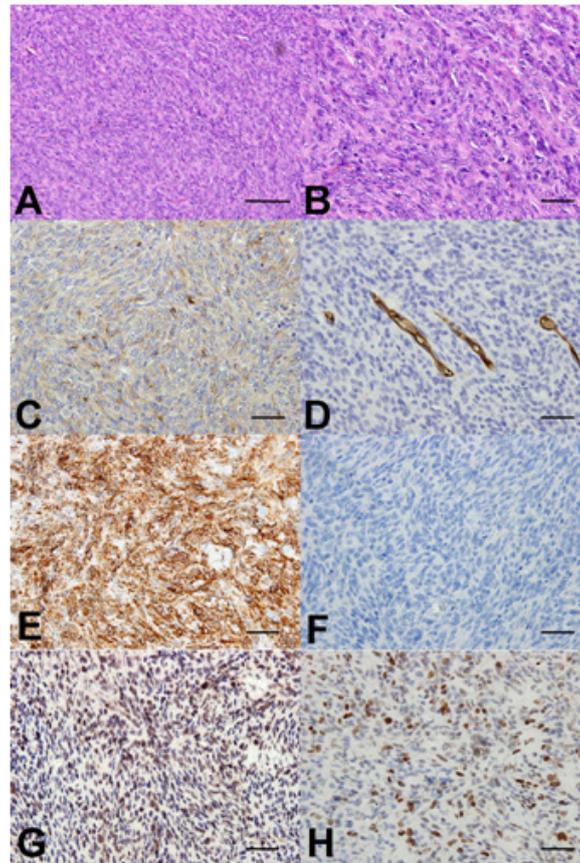


Figure 2. (A) Hematoxylin & eosin (HE)-stained medium magnification photomicrogram (20x). The tumor comprised highly proliferating bland spindle cells. Scattered collagenous stroma was found. (B) HE-stained high magnification photomicrogram (40x) showed mitotic figures (3/HPF). (C) Epithelial membrane antigen (EMA) was weakly positive. (D) CD34 was only positive in vascular endothelial cells, and was negative in the tumor cells. (E) BCL-2 was strongly positive in most of tumor cells. (F) CD99 was negative in the tumor cells. (G) STAT6 was positive in the nuclei of most of tumor cells. (H) Ki-67 was positive in around 30% of the tumor cells. Bar=100 μ m.

Hematoxylin and eosin (HE) staining revealed a complicated pattern of PAS-negative short spindle shape tumor cells in the extirpated tissue, in which mitotic cells were recognized (2 to 5/10 HPF). Epithelioid features and staghorn-shaped blood vessel were not observed. The tumor cells were positive for vimentin, BCL-2 and EMA, but were negative for CD99, CD34, S100, NCAM, synaptophysin, SMA, and desmin. Around 30% of the tumor cells showed positive staining for Ki-67. Based on these findings, a differential diagnosis of malignant solitary fibrous tumor (SFT), synovial sarcoma, and anaplastic hemangiopericytoma was confirmed. On further assessment, positive immunostaining for STAT6 was detected in the nucleus of most tumor cells. Based on these findings, this tumor was finally diagnosed as a malignant SFT (Figure 2).

DISCUSSION

Since histopathological diagnosis based only on HE is difficult, immunostaining for molecular markers is essential to obtain a relevant diagnosis. Although SFT is usually positive for CD34, BCL-2, CD99, EMA⁶; both CD34 and CD99 were negative in

the present case.

Recently, studies have shown that STAT6 expression in the nuclei of tumor cells was an important factor for the definitive diagnosis of SFT.^{2,6} Positive staining for STAT6 could accurately confirm a definitive diagnosis for SFT without depending on the CD34 status.² Moreover, STAT6 is usually negative in the tumors that mimic SFT, involving synovial sarcoma or fibromyxoid sarcoma.²

STAT6 overexpression has been reported to be caused by the formation of its fusion gene NAB2-STAT6 that results in the former's overexpression in the nuclei.^{3,4} The fusion gene accelerates cellular proliferation and the activation of early growth response (EGR)-responsive genes.³ Thus, this gene might be a potential target of therapeutic strategies for SFT.⁷

Usually in the case of sino-nasal SFT, the tumors are confined to the extracranial regions and their proliferative activity is not very high. At this stage the SFT presents an indolent clinical image.⁸ In contrast to previous reports, the tumor in the present

case demonstrated more mitotic cells and high Ki-67-positivity. The presence of more than 4/10 HPF mitotic cells was reported as a marker for malignant SFT.⁵ Therefore, the present case was classified as a malignant SFT. Our results were further corroborated by another case that reported a high number of mitotic cells (>10/10 HPF) and intracranial extension in the basifrontal region in the sino-nasal malignant SFT.⁹

Although enhanced expressions of the stem cell-related proteins ALDH1¹⁰ and CD44¹¹ were also reported, their expressions were not associated with malignant properties as opposed to matrix-metalloproteinase-2 (MMP2) expression.¹¹

The first-line treatment for sino-nasal SFT is surgical excision.¹ Accordingly, in the present case, the tumor was immediately resected following its discovery. Since the tumor had extended widely into both the intracranial and paranasal sinus spaces, complete resection was difficult to achieve. Local recurrence is found in approximately 25% of cases with deep expansion of the tumors.¹ As post-operative radiotherapy is also recommended for the treatment of malignant SFT,¹² the same is planned for our patient as well. Close follow-up of more than 10 years is recommended after surgery for the earlier detection of recurrence.¹

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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