

Single Case – General Neurology

Malignant Solitary Fibrous Tumor of the Right Cerebellum: A Case Report

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Keywords

Solitary fibrous tumor · Central nervous system · Immunohistochemical · Pathology

Abstract

Solitary fibrous tumor is a very rare mesenchymal tumor that occurs mostly in the pleura, and there are few reported cases of a presence in the central nervous system, particularly in the cerebellum. In 2016, the WHO classified solitary fibrous tumors into grade I. In this article, we present a case of malignant solitary fibrous tumor recurring 8 years after surgery in a 63-year-old male. Magnetic resonance imaging showed low to intermediate mixed signal intensity on T1W1. Immunohistochemical staining positivity for Vimentin, CD99, CD34 and Bcl-2, it is consistent with the immunohistochemical characteristics of solitary fibrous tumor. We resected the patient's tumor, and the patient was followed up for 3 months with no signs of recurrence. Solitary fibrous tumors are very rare in the central nervous system. Immunohistochemical staining positivity for CD34 and Bcl-2 is strongly expressed in most solitary fibrous tumor. Surgical resection is the preferred treatment. Due to the small number of cases, the biological behavior and prognosis of this tumor need to be further explored.

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Introduction

Solitary fibrous tumor (SFT) is a rare mesenchymal tumor that occurs mostly in the pleura. It was first reported and named by Klemperer and Rabin in 1931 [1]. The 2016 WHO classification of central nervous system tumors combines intracranial solitary fibrous tumor with hemangiopericytoma (HPC) into the same category, using the combined term SFT/HPC [2]. In 2016, the WHO classified SFT/HPC into grade I, corresponding to the original SFT, HPC, and inter-variant HPC [2]. SFT can occur anywhere in the body, but it is very rare in the central nervous system [3], particularly in the cerebellum. A 63-year-old man was admitted to our hospital for walking instability. According to the patient's previous diagnosis, it could be recurrence of the tumor. The patient's diagnosis, treatment process and related literature review are reported as follows.

Case Presentation

This patient was a 63-year-old Chinese man who was admitted to our hospital for 1 week of walking instability. Eight years ago, this patient suffered from headache and dizziness symptoms and was diagnosed with atypical solitary fibrous tumor. The patient was treated twice with gamma knife radiosurgery after the surgery, and no adjuvant radiotherapy was used. In addition to walking instability, there were no symptoms such as nausea and vomiting. Moreover, there was no other medical history, including hypertension and diabetes. A brain magnetic resonance imaging (MRI) scan revealed a $5.1 \times 4.1 \times 2.6$ cm homogeneous mass located in the right cerebellum (Fig. 1).

After discussion in the department, the patient underwent a craniotomy and resection of tumor. During the surgery, the tumor's color was yellow-white, soft in texture, rich in blood supply, and there are obvious boundaries between tumor and surrounding tissues. Postoperative pathological report suggested a malignant solitary fibrous tumor, the tumor cells were positive for Vimentin, CD99 (Fig. 2), bcl-2 (Fig. 3) and CD34 (Fig. 4), EMA, S100 and GFAP were negative. Ki-67 immunostaining showed a 30% proliferative index (Fig. 5). Histological examination showed that the tumor was composed of spindle cells (Fig. 6). The patient was discharged with no symptoms such as limb paralysis. Three months after the operation, MRI showed that the tumor was completely removed and no recurrence was found (Fig. 7).

Discussion

SFTs are rare in the central nervous system. In 1996, the SFT of the central nervous system was first described by Carneiro et al. [4]. SFTs occur in individuals between the ages of 60 and 70 years. The incidence rate in men and women is roughly the same, and there is no obvious gender difference [5]. But the intracranial SFTs are more common in women around 50 years of age [6]. Most SFTs are benign, but about 10–15% of cases have malignant features such as recurrence or metastasis [7]. Patients with intracranial SFTs may have several non-specific symptoms associated with location of tumor or increased intracranial pressure, such as dizziness, headache, hemiplegia, gait disturbance, hearing or mental disorder [8]. Intracranial SFTs usually need to be differentiated from some intracranial tumors, such as fibrous meningioma, hemangiopericytoma, etc. [9]. However, it is generally difficult to identify SFTs

and other tumors before surgery. Because the characteristics of SFTs in imaging are not clear. In MRI, it usually appears as a low to intermediate mixed signal intensity present on T1WI and a high or mixed signal on T2WI [10]. Therefore, it is difficult to perform a preoperative diagnosis of SFTs by imaging examination alone, and it is still necessary to combine the postoperative pathological examination to make it clear. Pathologically, solitary fibrous tumor cells usually show positive for CD34, CD99 and bcl-2, which has a good diagnostic value for SFTs [11], and this patient's pathological diagnosis is consistent with the pathological characteristics of SFTs. It is generally believed that CD34 has certain specificity and accuracy for the diagnosis of SFTs [12]. Bisceglia et al. [13] suggested high Ki-67 (>5%) as a factor for poor prognosis of SFT of the central nervous system. The pathological results of this patient showed that Ki-67 was about 30%. Therefore, the possibility of recurrence is high and regular follow-up is necessary.

Despite the high recurrence rate after surgical resection, there is no doubt that surgical resection is still the preferred treatment [14]. Total resection is significantly better than subtotal resection [15, 16]. Whether or not adjuvant radiotherapy is needed after tumor resection is still undecided, and adjuvant radiotherapy has no significant effect on improving overall survival of patients [17]. The average time to recurrence of SFTs is about 3 years [18]. It is worth mentioning that 8 years ago, the patient underwent surgery and was followed by gamma knife radiosurgery. Since then, no radiotherapy or drug treatment has been carried out. It can be concluded that postoperative gamma knife radiosurgery may be effective for SFTs, but to definite this viewpoint is difficult due to the limited number of cases.

Conclusion

SFTs of the central nervous system are very rare, and have slow growth, recurrence, and metastatic tendencies. Immunohistochemical staining positivity for CD34 and Bcl-2 is strongly expressed in most SFTs, it could be helpful for the diagnosis of SFTs. Surgical resection is the preferred treatment. Whether it is adjuvant radiotherapy is still inconclusive. Although most SFTs are benign, long-term follow-up is still necessary. Due to the limited number of cases, the biological behavior and prognosis of this tumor need to be further explored.

Statement of Ethics

This patient has given his written informed consent for publication of data and images. The study protocol conformed to the ethical guidelines of the 2013 Declaration of Helsinki.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All authors were involved in the preparation of the manuscript. Yang Liu, Li-Gang Chen, and Da-Ping Song designed the study. Ye-tao Zhu, Yang Liu, Li-Gang Chen, and Da-Ping Song analyzed the preoperative data. Ye-Tao Zhu, Yang Liu and Da-Ping Song analyzed the surgical and pathological findings. Postoperative follow-up and data analysis were conducted by Ye-Tao Zhu, Yang Liu and Li-gang Chen, and Yang Liu revised the manuscript. All authors read and approved the final manuscript.

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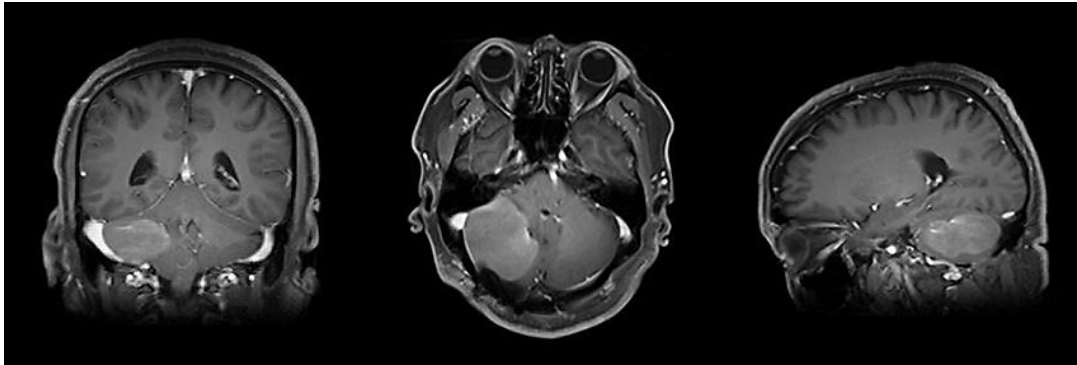


Fig. 1. MRI showing the right cerebellum space-occupying lesion on T1-weight image.

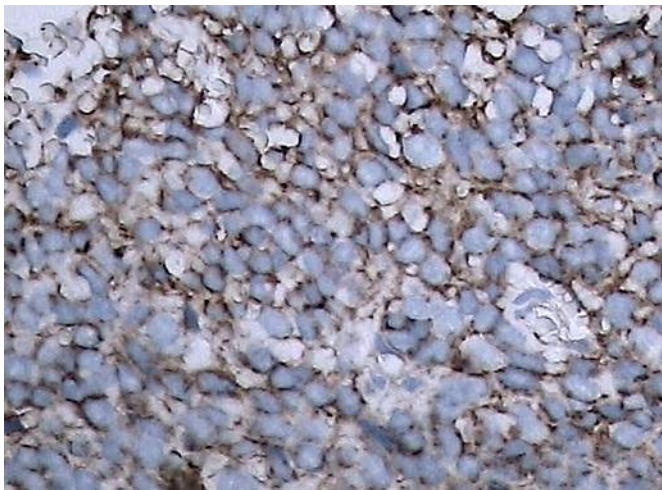


Fig. 2. Tumor cells showing diffuse immunohistochemical positivity for CD-99.

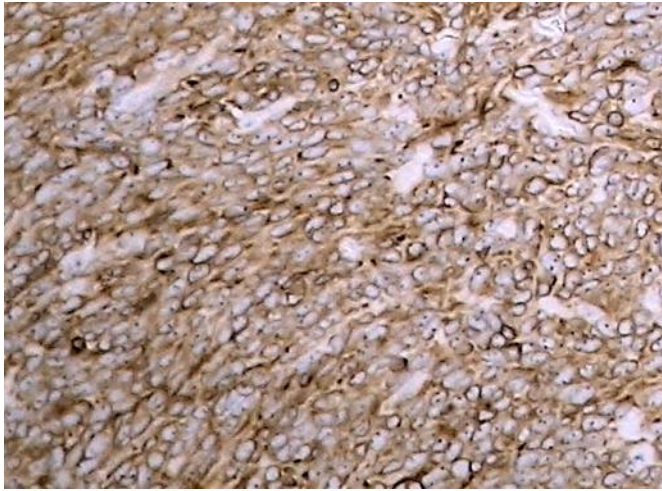


Fig. 3. Tumor cells showing diffuse immunohistochemical positivity for Bcl-2.

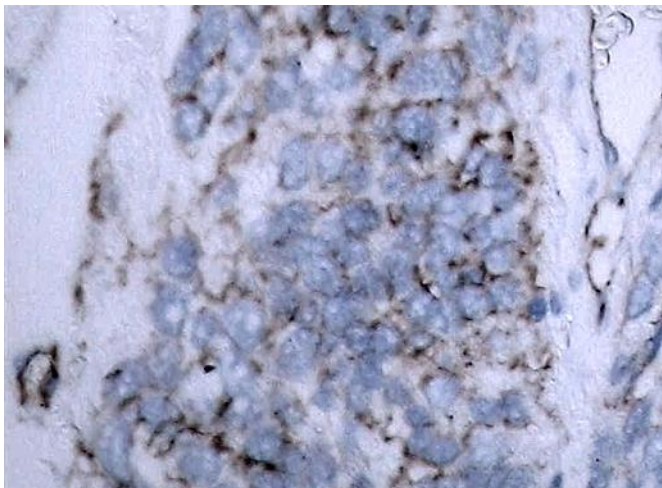


Fig. 4. Tumor cells showing diffuse immunohistochemical positivity for CD-34.

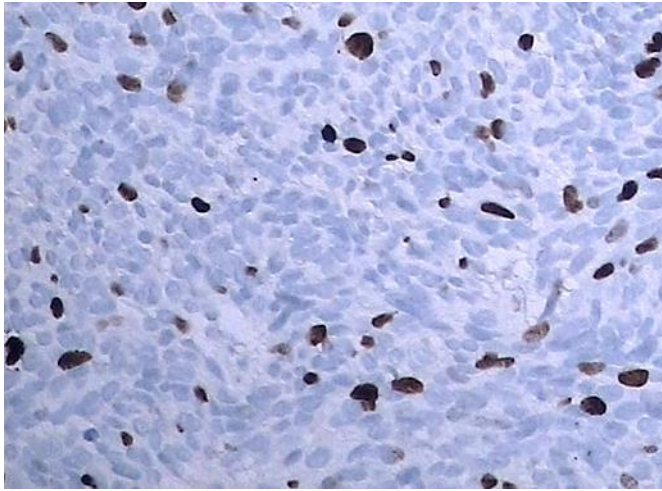


Fig. 5. Ki-67 immunostaining showed a 30% proliferative index.

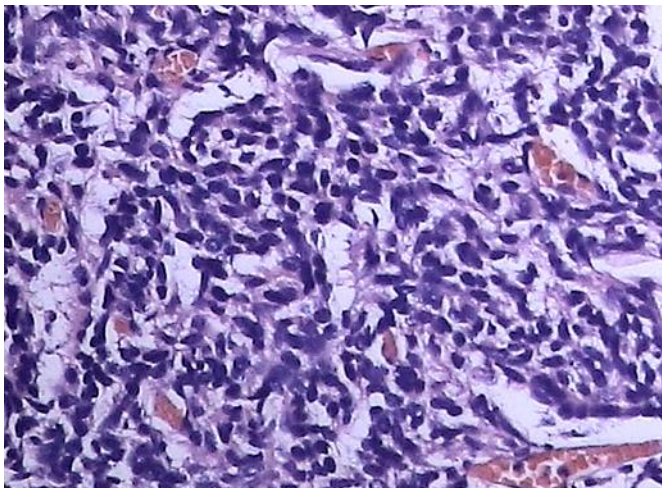


Fig. 6. Histological examination showed that the tumor was composed of spindle cells (H&E staining).

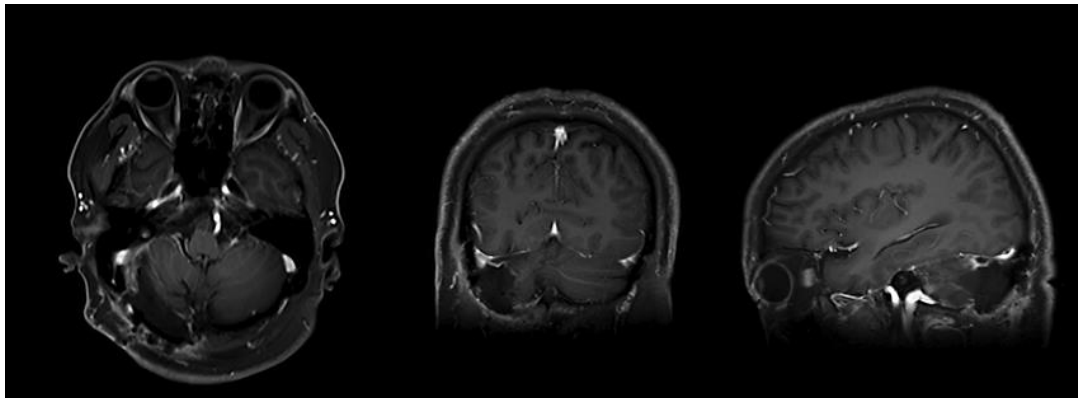


Fig. 7. MRI showed that the tumor was completely removed.