MEGA JOURNAL OF CASE REPORTS

Case Report

Compiled Date: March 29, 2023

Intracranial Solitary Fibrous Tumor Misdiagnosed before Operation: Case Report and Literature Review

Zhu Yajun, Sun Xiaochuan and Guo Zongduo*

Department of Neurosurgery, The First Affiliated Hospital of Chongqing Medical University, China

***Corresponding author**: Guo Zongduo, MD, PhD, Department of Neurosurgery, The First Affiliated Hospital of Chongqing Medical University, No. 1, Youyi Road, Yuanjiagang, Yuzhong District, Chongqing, China, Tel: +86 13594118695; E-mail: stonegzd@163.com

Abstract

Objective: To analyze and summarize the clinical data of 2 patients with intracranial solitary fibrous tumor (ISFT), and improve the clinicians' understanding and diagnosis of the disease.

Methods: The clinical manifestations, imaging data, pathological characteristics and other data of 2 patients with ISFT admitted to our hospital were analyzed retrospectively. The clinical characteristics were summarized, and the relevant literature was retrieved through PubMed with the "intracranial solitary fibrous tumor "keyword.

Results: In both patients, the tumors occurred in the supratentorial region, one in the left occipital region and the other in the right temporal and occipital region. MRI: In case 1, the lesion showed a mixed low signal on T1WI and a mixed high signal on T2WI. The lesion was accompanied by cystic change and necrosis. After enhancement, uneven enhancement was seen, including the "black-white sign" and "black-white reversal sign"; in case 2, the lesions showed equal signals on T1WI and T2WI and no cystic change or necrosis in the lesions. After enhancement, the lesions were significantly and uniformly enhanced, and a "dural tail sign" could be seen; No flow void signal was seen in both. MRS: The N-acetyl aspartic acid (NAA) peak was decreased and the choline (Cho) peak was increased in 2 patients; The lactate (Lac) peak increased in case 1, and the alanine (Ala) peak increased in case 2. Immunohistochemistry: CD34, Bcl-2, and CD99 were positive, S-100 was negative. EMA was positive in case 1, but negative in case 2. Ki-67 proliferation index 30% (+) in case 1, 8% (+) in case 2.

Conclusion: ISFT is an extremely rare intracranial extracerebral tumor. Brain MRI ("black-white sign" and "black-white reversal sign") and MRS can assist in the diagnosis of this disease, but the gold standard for diagnosis remains pathology and immunohistochemistry.

Keywords: Solitary fibroma; Intracranial extracerebral tumors; Imaging findings; Pathological characteristics

Introduction

The Solitary Fibroid Tumor (SFT) is a rare tumor originating from mesenchymal tissue. Most of them are benign tumors, and few can be malignant. Some studies have shown that SFT is a spindle cell tumor, which originates from dendritic mesenchymal cells expressing the CD34 antigen, and can occur, in multiple parts of the human body, mostly in the pleura **[1-4].** Scholars initially believed that STF only occurred in the visceral pleura **[5].** With the accumulation of cases and clinical progress, STF in extra-pleural sites has also been reported. However, Intracranial Solitary Fibrous Tumor (ISFT) is extremely rare, accounting for 2.5% of meningeal tumors and less than 1% of intracranial tumors **[6,7].** ISFT imaging features are similar to meningioma, glioma, and other tumors. Therefore, it is easy to misdiagnose the tumor at its early stage and difficult to make a differential diagnosis because of the lack of specificity of the clinical features and imaging manifestations of ISFT. At present, the gold standard for the diagnosis of ISFT is pathology and immunohistochemistry. The clinical manifestations, imaging characteristics, pathological features, immunophenotypes, and other data of 2 cases with ISFT were analyzed retrospectively, and we searched PUBMED with the keyword “intracranial solitary fibrous tumor”, and found a few papers published.

Case Presentation

Case 1

A male 38 years old patient was admitted to the hospital with "post-traumatic headache for one month". The main manifestation was recurrent headaches after trauma, without any other accompanying symptoms. Imaging examination suggested: 1. MRI (Figure 1a-c): left occipital cystic solid mass, hemorrhage in the mass, suspected glioma; 2. MRS (Figure 2a): The peak of N-acetyl Aspartic Acid (NAA) in the focus area decreased, the peak of choline (Cho) increased, and the inverted lactate (Lac) peak was visible. It is more likely to be diagnosed as left occipital glioma upon admission. Then the intracranial space-occupying lesions were resected surgically. During the operation, the lesions were cystic and solid with rich blood supply; The cystic part is grayish white, with general blood supply and obvious boundary with the surrounding brain tissue. Postoperative pathology showed a solitary fibroid tumor (Figure 4a). Immunohistochemical results are as follows: KI67 10% (+), CD34 (+), B-cl2 (+), CD99 (+), EMA (+), S100 (-). One month after the operation, the patient received radiotherapy. The patient was regularly followed up for one year. No tumor recurrence or new growth was found in the head MRI (Figure 1d-f). At present, the patient is still being followed up.

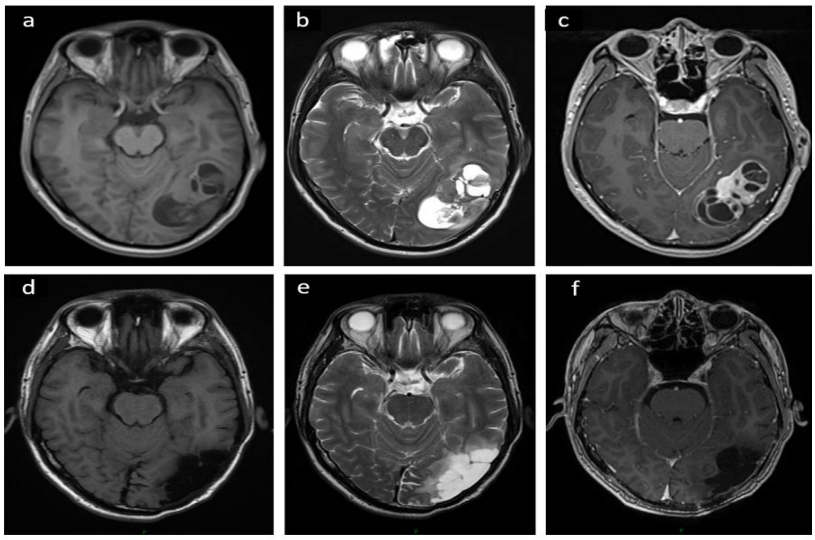

Figure 1: MRI of case 1 before and 1 year after operation; a-c) "Black-white sign" and "black-white reversal sign" were seen before the operation. d-f) No tumor recurrence was found 1 year after the operation, and cerebrospinal fluid signals were seen in the operation area.

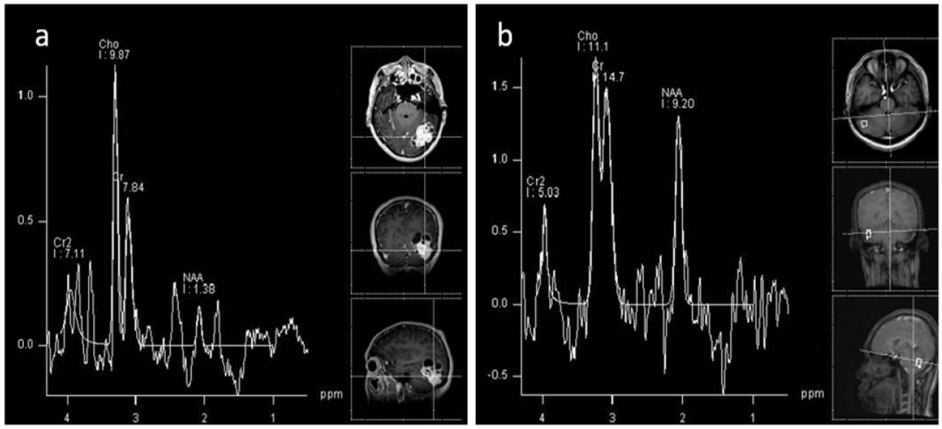

Figure 2: a) MRS of case 1 showed that the NAA peak decreased, the Choline peak increased, and the lactic acid peak was inverted. b) MRS of case 2 showed that the NAA peak decreased, the Choline peak increased, and the Ala peak inverted.

Case 2

A male 30 years old patient was admitted to the hospital with" sudden disturbance of consciousness and limb twitching for 1 day". The patient had a sudden loss of consciousness and limb twitching without inducement, which lasted for several minutes and then relieved. No obvious positive signs were found in the nervous system examination. Imaging examination suggested: 1. MRI **(Figure 3a-c):** irregular mass shadow under the skull inner plate in the right temporo-occipital junction area, meningioma may be present; 2. MRS **(Figure 2b):** NAA peak decreased, the Cho peak increased, and inverted Alanine (Ala) peak was seen in the lesion area. Considering diagnosis of right temporo-occipital meningioma. Then the lesion was removed by surgery. During

the operation, it was found that the lesion was buried in the brain parenchyma, which was solid, with a rich blood supply and unclear boundary with the surrounding brain tissue. Postoperative pathology also showed a solitary fibroid tumor **(Figure 4b).** Immunohistochemical results are as follows: Ki67 8% (+), CD34 (+), Bcl-2 (+), CD99 (+), EMA (-), S100 (-). The patient did not receive chemotherapy after the operation and was followed up regularly for 5 years. No tumor recurrence or new growth was found in the head MRI **(Figure 3d-f).**

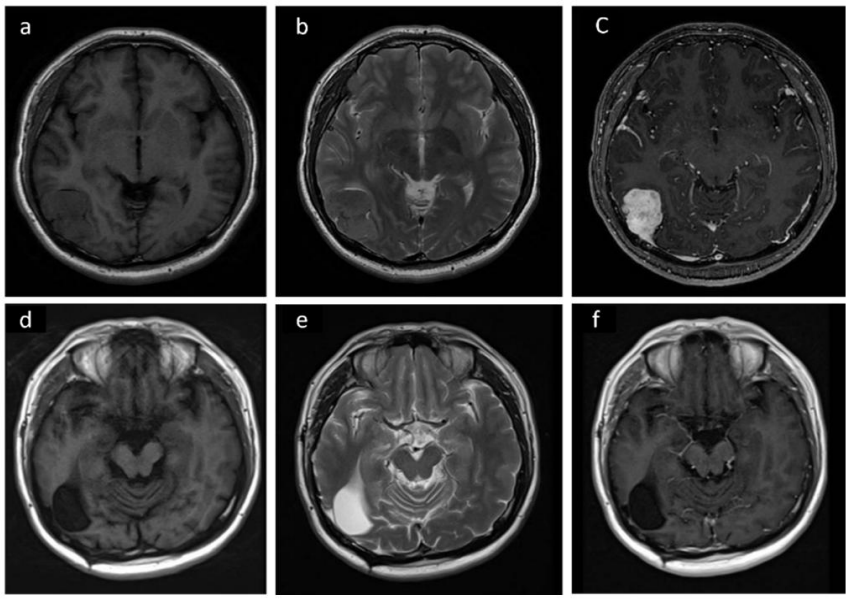

Figure 3: MRI of case 2 before and 5 years after operation; a-c) A mass shadow in the right temporoparietal occipital junction area before operation. It was significantly and evenly enhanced, and the "dural tail sign" was seen after enhancement. d-f) No tumor recurrence was found 5 years after the operation.

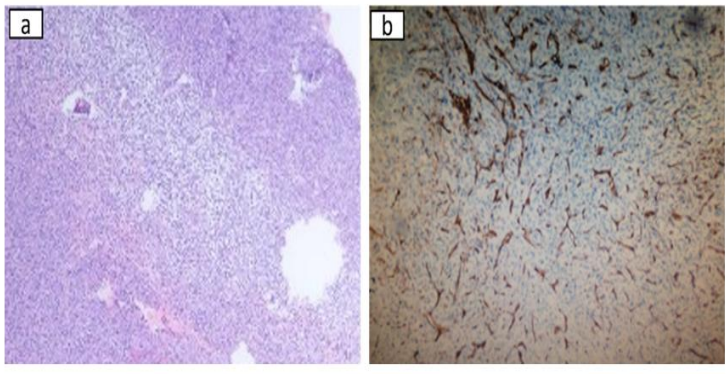

Figure 4: Postoperative pathological sections of case 1 (a) and case 2 (b).

Discussion

The incidence rate of ISFT is extremely low, accounting for about 0.4% of all intracranial tumors **[7].** In recent years, with the development of imaging and pathology, clinicians gradually recognized ISFT cases. The tumor is predominantly benign with a malignant transformation rate of 10% - 15% **[6].** It is insidious and slowly growing, and there are no clinical symptoms in the early stage. In the later stage, with the increase of the tumor, it causes symptoms of high intracranial pressure and space-occupying symptoms at the corresponding lesion location, or it starts with epilepsy. The two cases reported in this paper had headache and seizures as the main clinical manifestations respectively, which lacks specificity. Therefore, they are prone to be missed and misdiagnosed in the early stage of the disease.

Imaging is somewhat helpful for diagnosing ISFT, especially MRI. Previous studies have shown that ISFT has certain characteristics on MRI **[8-10].** Although the shape and signal of the lesion area were varied, the "black-white sign" with alternating high and low signals can be seen on T2WI in some cases. What's more, the" black-white reversal sign" can be seen in the low signal area under T2WI after enhancement, which is the characteristic manifestation of the tumor in MRI and may be of great significance for diagnosis. Because the mucus necrosis or degeneration area showed a high signal, the tumor cell area showed a slightly high signal, and the low cell area containing dense collagen fibers showed a low signal, ISFT showed a mixed signal on T2WI **[10].** It is obvious that case 1 can see a typical "black-white sign" and "black-white reversal sign", which is very consistent with the characteristics of ISFT previously reported. In addition, the differential diagnosis between ISFT and meningioma has been reported in some literatures **[11-13],** because they have some similarities in MRI. Among them, the "dural tail sign" is the main cause of the preoperative misdiagnosis of ISFT. In case 2, the "dural tail sign" was seen on MRI before the operation, which was misdiagnosed as meningioma. In the past, a few cases have found that ISFT shows a "dural tail sign" on MRI, as in case 2, so the "dural tail sign" is not unique to meningioma or ISFT. It indicates that ISFT not only originated from the mesenchyme but also meningeal epithelial cells. MRS may play a role in differentiating meningioma from ISFT. Some scholars have suggested that Glutamic Acid (Gla) and Ala peak elevation is seen on MRS of meningiomas, but not on ISFT **[14-16].** However, in addition to the decrease in the NAA peak and increase in the Cho peak seen with ISFT, the Lac peak and lipid peak were seen in some lesions. The above characteristics may be a preoperative differentiation method between ISFT and meningioma. In both cases, the NAA peak decreased and the Cho peak increased, and the Lac peak was also seen in case 1, which may be helpful for the diagnosis of ISFT. However, in case 2, MRI imagining showed the "dural tail sign" and the Ala peak, leading to preoperative misdiagnosis. So preoperative imaging findings can only be used as an auxiliary diagnosis, and the final diagnosis is still based on pathological biopsy findings.

The pathology of ISFT is characterized mainly by fascicles of spindle cells or irregularly arranged around hyalinized vessels with bland collagen fibers, and its morphologic appearance is similar to that of other spindle cell tumors, making the differential diagnosis difficult. Its diagnosis requires further immunohistochemical examination. In the present study, most ISFT expressed CD34, Bcl-2, CD99, S100, and vimentin in all **[13,17].** Among them, CD34 and Bcl-2 are characteristic immune markers. The 2 cases herein were compatible with the above observations. Although some studies consider EMA to exhibit high specificity for meningiomas **[18],** the EMA results of cases 1 and 2 were opposite: case 1 showed positive but case 2 was negative. So it was precisely the negative expression of EMA in case 2, which excluded the possibility of its meningioma. What's more, after

the World Health Organization (WHO) classified ISFT in 2016, it is generally believed that the Ki-67 proliferation index of Class III ISFT is usually ≥ 10% **[19].** In case 1, the Ki-67 proliferation index of the tumor is 30%. And the tumor was irregular in shape with cystic change and necrosis, a high possibility of malignancy, which was the reason for receiving postoperative radiotherapy. Therefore, it can be speculated based on the Ki-67 proliferation index between benign and malignant to guide the postoperative radiotherapy treatment.

Conclusion

ISFT is a kind of intracranial extracerebral tumor with an extremely low clinical incidence rate. It lacks specificity and is easy to be misdiagnosed. The possibility of ISFT should be considered when the "black-white sign" on T2WI of MR and/or the "black-white reversal sign" on the low signal area of T2WI after enhancement, the decrease of NAA peak and the increase of Cho peak indicated by MRS. Pathology and immunohistochemistry are still the "gold standard" for the diagnosis of ISFT.

References

1. Yilmaz C, Kabatas S, Ozen OI, Gulsen S, Caner H, Altinors N. Solitary fibrous tumor. J Clin Neurosci. 2009;16(12):1578-81.
2. Masmoudi M, Hasnaoui M, Dgani I, et al. Solitary Fibrous Tumor of the External Auditory Canal. Ear Nose Throat J. 2021:145561321997539.
3. Sheikhy A, Fallahzadeh A, Ahmadi-Tafti SH, et al. Intrapericardial solitary fibrous tumor: A case report and review of literature. Echocardiography. 2021;38(6):1052-6.
4. Gopakumar S, Srinivasan VM, Hadley CC, et al. Intracranial Solitary Fibrous Tumor of the Skull Base: 2 Cases and Systematic Review of the Literature. World Neurosurg. 2021;149:e345-e359.
5. Gengler C, Guillou L. Solitary fibrous tumour and haemangiopericytoma: evolution of a concept. Histopathology. 2006;48(1):63-74.
6. Sung KS, Moon JH, Kim EH, et al. Solitary fibrous tumor/hemangiopericytoma: treatment results based on the 2016 WHO classification. J Neurosurg. 2018;1-8.
7. Kinslow CJ, Bruce SS, Rae AI, et al. Solitary-fibrous tumor/hemangiopericytoma of the central nervous system: a population-based study. J Neurooncol. 2018;138(1):173-82.
8. Clarencon F, Bonneville F, Rousseau A, et al. Intracranial solitary fibrous tumor: imaging findings. Eur J Radiol. 2011;80(2):387-94.
9. Kim HJ, Lee HK, Seo JJ, et al. MR imaging of solitary fibrous tumors in the head and neck. Korean J Radiol. 2005;6(3):136-42.
10. Liu Y, Tao X, Shi H, Li K. MRI findings of solitary fibrous tumours in the head and neck region. Dentomaxillofac Radiol. 2014;43(3):20130415.
11. Cheng L, Ni H, Dai Y. Intracranial solitary fibrous tumor mimicking meningioma: A case report. Medicine (Baltimore). 2020;99(50):e23504.
12. Xiao D, Shi J, Zhou M, et al. Tumor volume and the dural tail sign enable the differentiation of intracranial solitary fibrous tumor/hemangiopericytoma from high-grade meningioma. Clin Neurol Neurosurg. 2021;207:106769.

13. Ashizawa K, Ogura K, Nagase S, et al. A collision tumor of solitary fibrous tumor/hemangiopericytoma and meningioma: A case report with literature review. Pathol Int. 2021;71(10):697-706.
14. Righi V, Tugnoli V, Mucci A, Bacci A, Bonora S, Schenetti L. MRS study of meningeal hemangiopericytoma and edema: a comparison with meningothelial meningioma. Oncol Rep. 2012;28(4):1461-7.
15. Zhang T, Yu JM, Wang YQ, Yin DD, Fang LJ. WHO grade I meningioma subtypes: MRI features and pathological analysis. Life Sci. 2018;213:50-56.
16. Mama N, Ben Abdallah A, Hasni I, et al. MR imaging of intracranial hemangiopericytomas. J Neuroradiol. 2014;41(5):296-306.
17. Jeong AK, Lee HK, Kim SY, Cho KJ. Solitary fibrous tumor of the parapharyngeal space: MR imaging findings. AJNR Am J Neuroradiol. 2002;23(3):473-5.
18. Yamashita D, Suehiro S, Kohno S, et al. Intracranial anaplastic solitary fibrous tumor/hemangiopericytoma: immunohistochemical markers for definitive diagnosis. Neurosurg Rev. 2021;44(3):1591-600.
19. Li S, Zhou Q, Zhang P, et al. The relationship between the apparent diffusion coefficient and the Ki-67 proliferation index in intracranial solitary fibrous tumor/hemangiopericytoma. Neurosurg Rev. 2022;45(2):1625-33.

Citation of this Article

Yajun Z, Xiaochuan S and Zongduo G. Intracranial Solitary Fibrous Tumor Misdiagnosed before Operation: Case Report and Literature Review. Mega J Case Rep. 2023; 6: 2001-2007.

Copyright