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Surgical Treatment of Intracranial Solitary Fibroma Tumors

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Abstract: Objective: To summarize the clinical presentation, diagnosis, and therapy of solitary fibrous tumors (SFT) individuals. Methods: Broad literature research has been conducted as well as the author's own clinical experienced has been reviewed. Results: Articles from the most recent 30 years have been collected while a typical case of our own has been presented. Based on the most updated evidence, the therapy of SFT has been proposed. Conclusion: For intracranial SFT, it's rare and similar to common intracranial tumors in imaging examination, the misdiagnosis rate is high. Total surgical resection and postoperative radiotherapy have a certain effect on the prognosis of patients, and gene sequencing can guide the pathological molecular typing of patients and the next step of treatment.

Keywords: Central nervous system; Solitary fibrous tumors; Surgical treatment.

1. Introduction

Solitary fibrous tumors (SFT), mainly derived from mesenchymal spindle cells, are a relatively rare soft tissue tumor, which is common in middle-aged people. The most common site is the pleura, but also in other parts of the head and neck, including the orbit, nasal cavity, thyroid gland, parotid gland, etc. [1]. The most common location of intracranial SFT is supratentorial, followed by posterior fossa, lateral ventricle and fourth ventricle [2]. At present, the diagnosis of SFT is mainly through pathological diagnosis. Imaging examination, such as head MR or CT, often lacks specificity and is easy to be misdiagnosed [3]. A patient with intracranial solitary fibroma was admitted to the First Affiliated Hospital of Jinan University in January 2022. Preoperative magnetic resonance imaging revealed meningioma, but the final pathological result showed solitary fibroma. Now this case is reported as follows.

2. Clinical Presentation

A 27-year-old female patient was admitted to the hospital because of "repeated dizziness for one month with weakness of the right lower limb for one day". Physical examination: clear mind, answer to the point, right lower limb muscle strength 4, other limb muscle strength 5, limb muscle tension normal. Magnetic Resonance Imaging (MRI) showed an irregular mass shadow at the top of the left side, about 6.5x5.6x5.4cm in size. The focus showed low signal intensity (T1WI) and slightly high signal intensity (T2WI). Small patches of low signal intensity (T1WI) and high signal intensity (T2WI) were seen in the interior. Banded cerebrospinal fluid signals were seen around the periphery. Large patches of edema shadow were seen around. Enhanced scanning showed uneven enhancement of the focus, and meningeal tail sign (Figure 1A-1C) was seen. It was considered as meningioma. Cerebral angiography was performed on January 2022, it showed that the space occupying stain on the top of the left forehead was mainly supplied by bilateral middle meningeal artery, left anterior cerebral artery and left posterior cerebral artery, and no abnormality was found in other blood vessels. Subsequently, the left and right middle meningeal artery tumor feeding arteries were embolized. Operative treatment was performed in next day, during the operation, the Dural tension was high, the tumor was close to the cerebral falx, soft in texture, rich in blood supply, and there was an arachnoid space between the tumor and normal brain tissue. The base of the tumor was located at the side of the cerebral falx. The tumor was reduced first, and then the remaining tumors were completely removed under the microscope. Postoperative pathology showed solitary fibrous tumor (WHO Grade II). During the follow-up one year after operation, the patient did not complain of limb fatigue, and there was no recurrence sign in the reexamination of head MRI (Fig2A-2E).

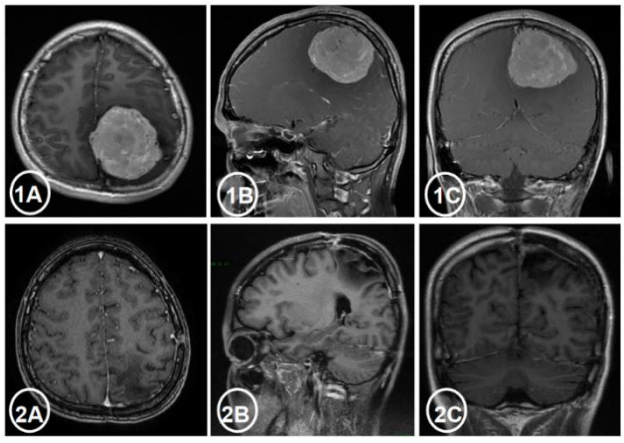

Figure1. 1A-1C Preoperative imaging, A: axial, B: sagittal, C: coronal ;2A-2C Postoperative imaging, A: axial, B: sagittal, C: coronal

3. Discussion

SFT of the central nervous system was first reported by Cameiro in 1996 [4]. It is difficult to distinguish it from

meningioma in clinical and imaging aspects. Its imaging features include: MRI often indicates that most of T1WI shows equal signal, T2WI signal depends on the proportion of tissue components constituting the lesion, tumor cells are loose and contain a lot of collagen fibers, showing low signal, tumor cells and vascular stroma are rich, so they show equal or slightly high signal, mucus degeneration or necrotic cyst changes show high signal. On CT, it showed uniform or high density. After enhancement, it was obviously enhanced uniformly. Some of them showed uneven enhancement. Those without enhancement were rare. In some reports, calcification was seen in the mass. In a word, intracranial SFT is rare and it mostly presents as a solitary mass with regular shape and clear boundary, the signal or density is relatively uniform. After enhancement, it is significantly enhanced, and empty vessels can be seen in it.

The genetic characteristics of solitary fibroma include the presence of NAB2-STAT6 gene fusion on chromosome 12q13 and the presence of STAT6 nuclear positive in IHC as a specific marker of SFT [5]. It may originate from Zimmerman's outer skin cells on the capillaries, which are spindle cells arranged close to the reticular fibrous membrane of the capillaries, and may be variant smooth muscle cells, and have multi-directional differentiation potential; SFT is invasive, easy to recur, and can cause tumor metastasis [6]. In this patient, our detection did not find the molecular variation of isolated fibrous tumor: NAB2-STAT6 gene fusion and TERT promoter mutation, suggesting that this patient may not be a potential beneficiary of immunotherapy.

SFT is difficult to be separated from other intracranial tumors on imaging and has a high misdiagnosis rate. Plain CT scan showed that the lesions were round or nearly round, with clear boundaries, and were equal, slightly high and high-density, which may be related to different tissue composition [7]. Slightly high density may contain some collagen fiber components, and high density may be related to abundant spindle cells and their arrangement. Most of the signals on T1WI were isogonal, while those on T2WI where is signal [8]. Depending on the proportion of tissue components that constitute the lesion, tumor cells with loose and large amount of collagen fibers show low signal, tumor cells rich in tumor cells and vascular interstitial show equal or slightly high signal, and mucous degeneration or necrotic cyst change show high signal.

In immunohistochemical analysis, CD34 positivity is found in 80% to 100% of SFT [9], although CD34 expression may be weak in more aggressive lesions, which facilitates differentiation from other spindle cell tumors and is the most important marker for the diagnosis of SFT. In addition, STAT6 expression is important in the expression and classification of ISFT [10]. As for the differential diagnosis of intracranial SFT, the main differential diagnosis of ISFT is fibrous meningioma. Unlike the classic thickening of bone near meningiomas, SFT shows erosion of the adjacent skull. It can also mimic schwannomas when located in the cerebellopontine horn cisterna. Compared with ISFT, schwannomas exhibit more cystic degeneration and the resulting T2 hypersignal.

Intracranial SFT should be distinguished from acoustic schwannoma, meningioma, epidermoid cyst, and angioblastoma [11]. Auditory schwannoma shows thickening of auditory nerve tracts on the diseased side and enlargement of internal auditory tract. CT scan of meningioma shows calcification, adjacent bone often accompanied by hyperplasia, and the "meningeal tail sign" around the lesion can be distinguished. Epidermoid cysts were characterized by long T1 and long T2 signals, and fluid attenuation inversion recovery signals [12]. Diffusion-weighted imaging (DWI) was significantly limited, with no enhancement after enhancement. The characteristic manifestations of hemangioblastoma are large cyst and small nodules. The wall nodules of hemangioblastoma are significantly enhanced, but the cystic part is not enhanced, and empty vascular flow signals can be seen.

At present, the treatment of SFT is mainly through surgical resection, most SFT can be totally removed by surgery, but tumors located near the sagittal sinus or deep in the skull base can only be subtotal removed. Some studies have shown that temozolomide combined with bevacizumab has a good tolerance and clinical efficacy in SFT patients [13]. In addition, some studies have shown that gamma knife radiosurgery is a feasible auxiliary means to treat SFT [14]. The malignant degree of SFT is closely related to the expression of Ki-67. Ki-67>5% indicates poor prognosis, and more than 10% of the patients are malignant [15]. In this case, the patient’s Ki-67 nearly 10%, which is consistent with our gene sequencing results and have potential malignant potential. For such patients, we recommend long-term follow-up and regular review. If there is a recurrence, gamma knife treatment is required, and if necessary, whole brain radiotherapy is added.

4. Conclusion

SFT is rare and similar to common intracranial tumors in imaging examination, the misdiagnosis rate is high. Total surgical resection and postoperative radiotherapy have a certain effect on the prognosis of patients, and gene sequencing can guide the pathological molecular typing of patients and the next step of treatment.

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