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Research Article

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# Clinical and Imaging Misdiagnosis of Intracranial Fibrosarcoma

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**Established Facts**

Primary Intracranial Fibrosarcoma (PIF) is an uncommonly invasive tumor, accounting for 0.5-2.7% of all central nervous system tumors. There is no specificity in clinical symptoms, but headache or epilepsy are often the initial symptoms.

**Novel Insights**

1. Primary Intracranial Fibrosarcoma (PIF) is an uncommonly invasive tumor, increased awareness of the CT characteristics of this rare tumor may broaden the radiologist's knowledge base.
2. Understanding the imaging features of Primary Intracranial Fibrosarcoma is contribute to diagnose and further to treat it.

## Abstract

**Introduction:** To improve the clinical and imaging understanding of Primary Intracranial Fibrosarcoma (PIF) and reduce the misdiagnosis rate. Case Presentation 4 patients with PIF confirmed by pathology were collected and their clinical manifestations, CT and MRI findings were analysed. The main clinical manifestations were headache. Two cases had slightly higher CT findings and isodense mass shadow. Both cases had iso-low T1T2 mass signal on magnetic resonance imaging. Pathological findings showed tumor cells. **Conclusion:** The clinical manifestations of PIF patients were not specific. Most of the CT findings were round or lobulated masses with different densities, which were related to the collagen components of the masses. Most of the MRIs showed low T1, T2 signals.

**Keywords:** Primary intracranial fibrosarcoma; Imaging; Misdiagnosis; Computed tomography

## Introduction

Primary Intracranial Fibrosarcoma (PIF) is an uncommonly invasive tumor, accounting for 0.5-2.7% of all central nervous system tumors. There is no specificity in clinical symptoms, but headache or epilepsy are often the initial symptoms. Previous studies mostly discussed the clinical and pathological perspectives, but were hardly related to CT or MRI analysis. In this paper, four cases of intracranial fibrosarcoma, confirmed by surgery and pathology from October 2012 to June 2018, were retrospectively analysed to explore the CT and MRI image features and improve the understanding of this disease.

## Data and Methods

### Clinical Data

Among the cases of intracranial fibrosarcoma, there were two males and two females, 3 cases were aged between 20 and 36 years old (with an average age of 27.0 ± 6.7 years), and one case was 59 years old. In turn, the affected parts were right temporoparietal region, right frontotemporal region, left optic canal segment to the cavernous sinus segment and left temporoparietal. The clinical symptoms are non-specific, which is mostly related to the area of

the tumor and characterized by headache, dizziness, blurred vision.

### Methods of Examination

In this group of four patients, two were examined with Siemens 128-slice spiral CT, four received MRI plain scan + enhancement examination. Among them, one patient received MRS (3.0T Trio, Siemens, Erlangen, Germany). The sagittal plane, coronal and axial position T1-weighted enhanced images were obtained by intravenous injection of gadopenamide. The dose was 0.2 mmoL/kg and the rate was 2.0-3.0 ml/s (American Medard double-barrel high-pressure syringe).

### Image Analysis

Two senior radiologists (engaged in radiological diagnosis of the head and neck system for more than five years) were selected to diagnose the lesion. They separately used the double-blind method to evaluate the location, shape, size, boundary, density, signal and enhancement characteristics with the focus. Finally, a consensus diagnosis was reached after consultation.

### Surgical Pathology

All 4 patients were treated with surgical total resection and the specimens were excised with HE staining and immunohistochemical markers, including Vimentin, transmembrane glycoprotein (CD34, CD99), pan CK, Desmin, Epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), isocitrate dehydrogenase (IDH1), s-100 protein, smooth muscle actin (SMA) and bcl-2, cell proliferation index (ki-67).

## Results

### CT Performance

CT examination in two cases were all plain scanning. In one case, the right frontotemporal intracranial plate showed a slightly circular high-density shadow with a wide base, which was about 70 mm x 50 mm in size. High-density haemorrhagic shadow was seen in the edge and interior of the focus with CT value of 25~35HU and edematous shadows could be seen in the marginal brain tissue, Compression narrowing of right ventricle and displacement of midline structures (Figure 1a). In another case, from the left optic canal segment to the cavernous sinus segment, a long strip of isotone shadow was seen. The orbital apex was enlarged, communication with the intracranial area. The edge was clear, the size of which was about 32x11mm. The average CT value was 26.3HU. There was no obvious absorption and destruction of the adjacent bone (Figure 2a).

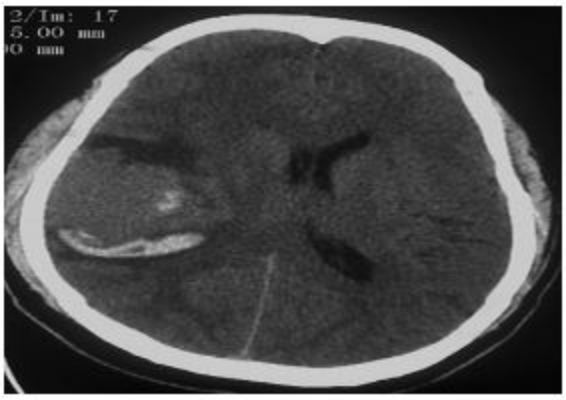



Figure 1a

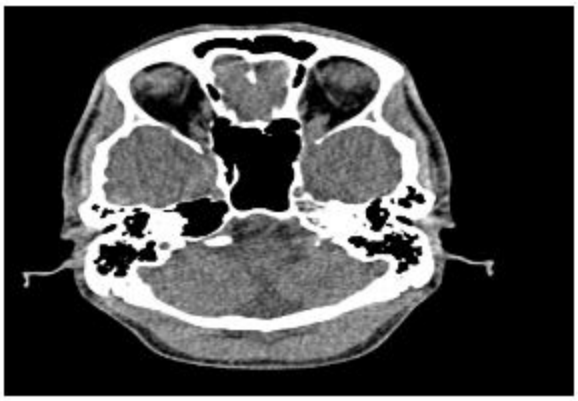


Figure 2a

### MRI Performance

MRI plain scan showed localized masses with T1WI and T2WI signals, two of the focuses showed patchy short T1 signal and patchy long T1 long T2 signal with haemorrhage and cystic change signal, small peduncular edema was seen around (Figure1b,1c), DWI showed an equal and slightly high signal, ADC showed a low signal (Figure1d,1e). The tumors in three cases were connected with the skull and Wide base of dura mater, and were clearly decomposed with brain tissue. The sulcus and cerebral fissure were not deepened obviously, the ventricular system was not enlarged evidently, and the midline was slightly displaced. A long “8” shaped mass was found between the left optic canal segment and the cavernous sinus segment in one case (Figure 2b,2c). Four cases all got obvious strengthened after enhancement (Figure1f,1g,2d), Peripheral edema was not enhanced and without the meningeal tail sign. One case of MRS showed that NAA (N-acetyl aspartic acid peak) was in a low position in the mass, Cho (choline) and Cr (creatine) peaks were slightly reduced, Lip peaks were significantly increased, and inverted Lac peaks were not seen (Figure1h).

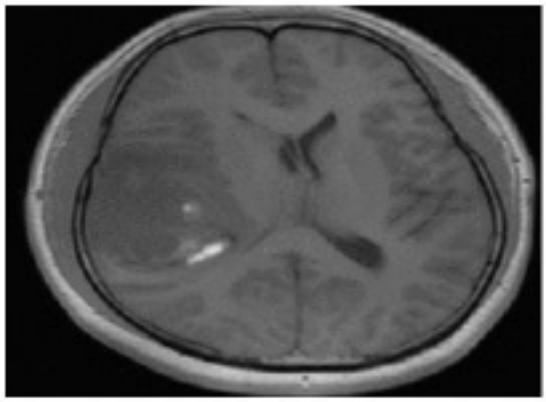


**Figure 1b**

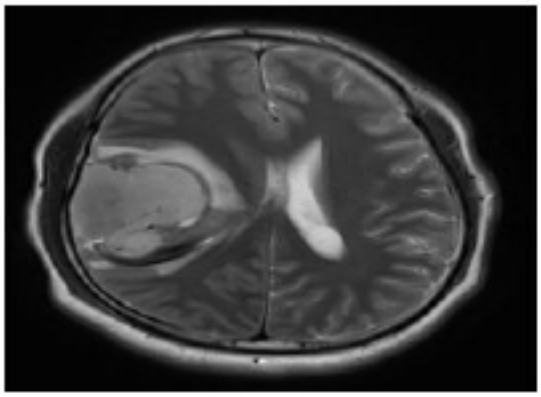


**Figure 1c**

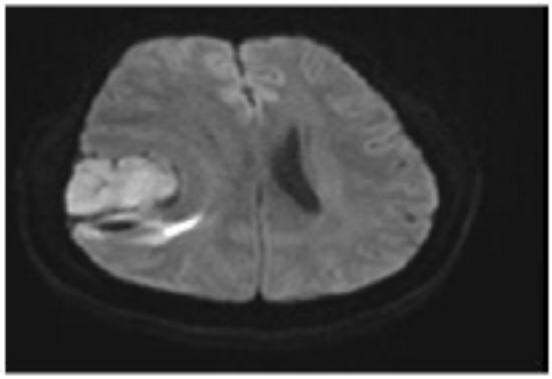
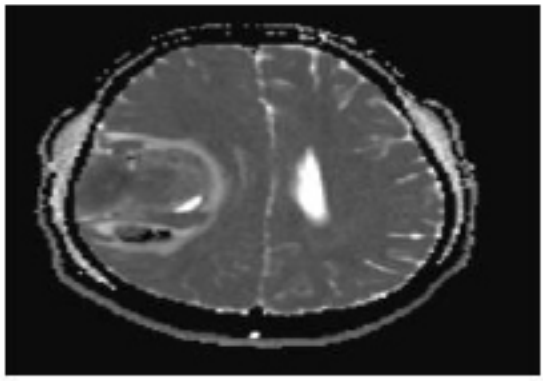


**Figure 1d**



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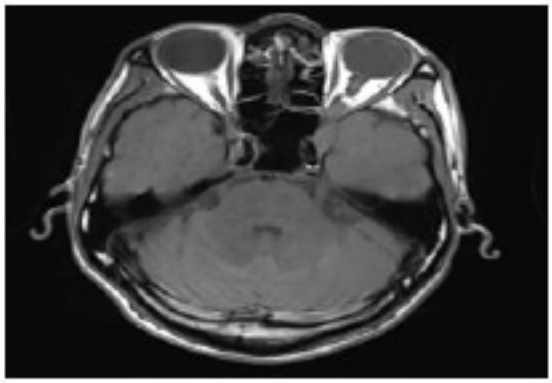


**Figure 2b**

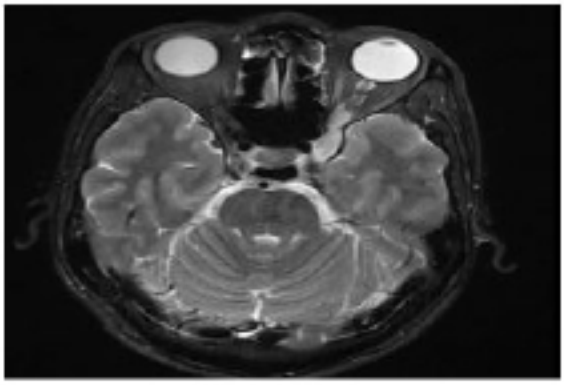
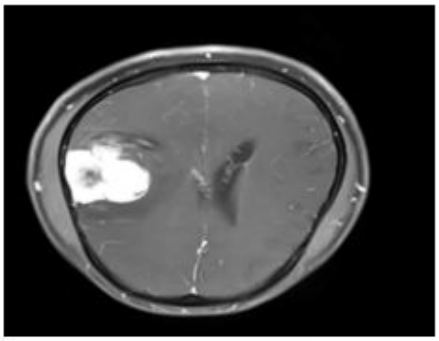


**Figure 2c**



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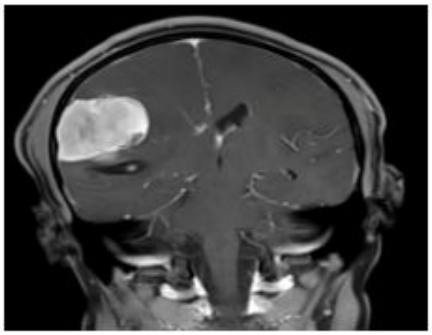

**Figure 1g.**

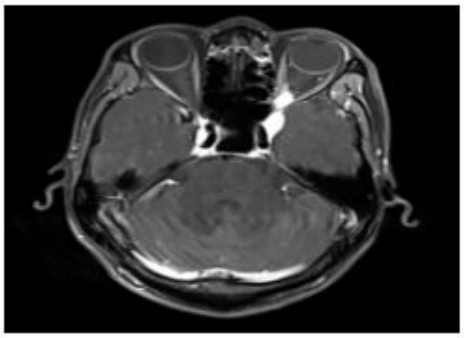

**Figure 2d.**

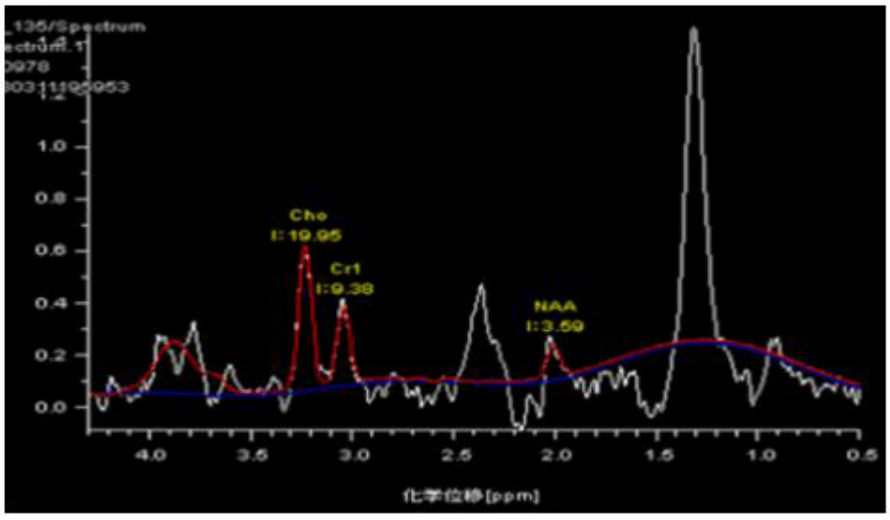


**Figure 1h:** showed that NAA (N-acetyl aspartic acid peak) was in a low position in the mass, Cho(choline) and Cr(creatine) peaks were slightly reduced, Lip peaks were significantly increased, and inverted Lac peaks were not seen.

**Figure 1a~1h:** Show a male of 20 years old who suffers from the intracranial fibrosarcoma. the right frontotemporal intracranial plate showed a slightly circular high-density shadow with a wide base, High-density hemorrhagic shadow was seen in the edge and interior of the lesion, On T1WI, the signal was uniform and slightly low, on T2WI, it was mainly iso-and slightly high, on DWI sequence, it was iso-and slightly high and on ADC, it was iso-and slightly low, Small patches of short T1 signal and long T1 long T2 signal were seen around them. MRS.

**Figure 2a-2d:** Show a male of 36 years old who suffer from Left optic canal - intracranial fibrosarcoma, CT showed equal-density long strips of mass, 8-shaped shape, enlarged apical foramen, MRI plain scan (b, c) showed T1WI, T2WI When the signal is clear, the edge of the tumor is clear, the anterior segment of the optic nerve is slightly elevated, the blood vessels in the muscle cone are slightly thickened, and the abnormal blood flow in the saddle area is not seen. The enhanced scan (d) is markedly uniform and enhanced.

### Pathological results

The nuclei of tumor cells are spindle-shaped, tapered at both ends, arranged in bundles, intersecting at acute angles, forming herringbone-like or fishbone-like structures. Immunohistochemistry showed Bcl-2 (+), CD99 (+), Vim (+), CD34 (+), pan-CK (-), Des (-), EMA (-), GFAP (-) IDH1 (-), S100 (-), SMA (-).Among them, Grade II differentiation in 2 cases, Ki-67(30%+); Grade III differentiation in 1 case, Ki-67(50%+); Grade II differentiation in 1 case, Ki-67(80%+).

## Discussion

### Summarize

PIF accounts for 0.5-2.7% of intracranial tumor and it is difficult to diagnose correctly. Besides, it is easily misdiagnosed as malignant fibrous histiocytoma [1]. PIF can occur at any age, which often leads to poor prognosis. In our study group, there were 3 patients with an average age of 27 years old who was basically consistent with the literature [2-4]; Two cases recurred respectively 3 months and 6 months after operation, one case did not show signs of recurrence during the course of re-examination, and one case did not follow up in our hospital. Some scholars believe that PIF originates from mesenchymal cells on the dura, pia mater, adventitia, choroid plexus or choroid, usually located on the supratentorial and superficial surface, and adheres to the durometer [2]. Among our group, three cases of intracranial fibrosarcoma were located in the frontal and parietal part of the brain. The rest one case was located in the intracranial and extra cranial communication of the left optic canal segment to the cavernous sinus segment, which is relatively rare. PIF preferred surgical treatment, early postoperative radiotherapy combined with radiotherapy may help to control tumour recurrence and improve prognosis. In order to prevent the possibility of distant metastasis, postoperative systemic chemotherapy can be appropriate after surgery.

### Clinical Pathology

The appearance and texture of the tumor are different depending on their collagen content: In a cell-rich area, it is the appearance of medullary tissue of brain; the tissue texture of colloidal fibrosarcoma is hard and rich in fibre. Therefore, there is no aponeurosis lesion like fibroid tumour in general. Meanwhile, Calcification focus, mucous degeneration, hose-like changes and osteoid formation could appear in the tumour tissues. According to its histological and cytological characteristics, intracranial fibrosarcoma can be divided into three types: fibrous, spindle cell and undifferentiated or polymorphic cell fibrosarcoma [4]. Intercellular variation, mitosis index, the amount and density of collagen production are important indicators to assess the malignancy of fibrosarcoma. Grade I differentiation is the best differentiation and the malignant degree is the lowest. It is characterized by fewer cells, slightly larger nuclei than normal fibroblasts, overdyeing, less mitosis and abundant collagen, which is so similar to fibroid tumors. The differentiation of grade IV is the worst. The cells with a high degree of anaplasia and peculiar shape have more mitosis and fewer collagen fibers, this is highly malignant and most like the malignant fibrous histiocytoma. The most common and typical changes of fibrosarcoma are grade II and III. Immunohistochemistry showed that Vimentin was widely expressed, but not epithelial, muscle, endocrine and neurogenic markers. Besides, there are different numbers of single spindle cell components and herringbone structure in grade II and III.

### Clinical Symptoms

It is not specific; the most common symptom is the transient headache. Other common symptoms include generalized tonic clonic seizures and scalp swelling and there is no neural defect in general. These tumors could show symptoms of occasional neurological dysfunction and paralysis in extreme conditions [3]. It can also show symptoms such as visual field, olfactory or auditory dysfunction and limb dyskinesia associated with tumor placeholder. The clinical manifestations are various and nonspecific in this group.

### Imaging Features

The location of the intracranial fibrosarcoma is usually superficial. The images are varied due to the content of different components, the more collagen, the denser the tissue, the higher the CT value, and MRI showed low signals such as T1WI and T2WI. CT mainly shows as class round or lobulated Soft tissue density and mass shadow, the mass is connected to the skull and dura, and the boundary of brain tissue is clear or unclear. The surrounding brain parenchyma is displaced by compression,

with uniform or uneven density, and spotted calcification, cystic change, etc.; Intertumoral hemorrhage and skull bone destruction are rare [2]. DWI presented equal and slightly higher signal, with obvious homogeneous enhancement but no meningeal caudal sign. The spectrum showed that NAA was in a low position in the mass, which indicates the absence of neuronal tissue in this region, Cho and Cr slightly decreased, Lip peak increased significantly, and the inverted Lac peak appeared, which suggests that ischemia and anoxia occur when the tumor grows fast, and then the Lip peak appears.; Cho being reduction is related to tissue necrosis and regeneration [6].

### The Distinction of Diagnoses

Intracranial fibrosarcoma usually occurs outside the brain, but it can also be found inside the brain parenchyma because the fibrosarcoma can ordinate from the brain parenchymal cells [7]. To make sure the tumor in the brain or out of the brain is very helpful for qualitative analysis. The main manifestations of being able to confirm and prompt extracerebral space occupying include: cerebrospinal fluid-perivascular space; white matter collapse sign and cortical compression sign; broad base connected with dura mater; adjacent bone changes, such as osteoporosis or destruction, compression and deformation of bone, enlargement of adjacent nasal sinuses; invasion of extracranial soft tissue tumors such as adjacent skull muscles; venous sinus obstruction and encapsulation of extracranial artery [8]. Usually, it should be differentiated from the following tumors: (1) meningioma [9]. Growth was slow, the boundary was clear, the lesions were uniformly enhanced and "Dural tail sign", there was much calcification, and MRS showed characteristic alanine (Ala) peak. (2) Peritoneal cell tumor [10]. also called peritoneal cell tumor, originated from perivascular cells surrounding capillaries and micro veins after capillaries, which is a kind of rare and invasive tumor. The changes of MRI signal are very complex, peritumoral edema is more obvious, common vascular flow empty shadow, uniform and obvious enhancement, generally without "dural tail sign". MRS has obviously increased Cho peak, slight lipid peak, lacking Ala peak and Glx peak. (3) Central nervous system lymphoma [11]. A short course, mostly within half a year; T1WI showed equal and slightly low signal intensity, with homogeneous signal、obvious peripheral edema and homogeneous enhancement of the lesion. Typical "notch sign", "sharp angle sign" and "clenching fist sign" could be found, especially in the lesions near the subarachnoid space. (4) Neurilemmoma: One case of PIF with left optic canal communicating with cavernous sinus, which should be differentiated from orbital neurilemmoma. The latter can communicate with intracranial neurilemmoma to form a dumbbell shape with uneven multi-signal and uneven enhancement of lesions. (5) Metastatic tumor: Most of them are founded in middle-aged and elderly people. They are usually located at the corticomedullary junction. The surrounding edema is obvious, showing uniform or circular enhancement. The presence of primary cancer supports the diagnosis of intracranial metastases.

To sum up, intracranial fibrosarcoma is easily confused with meningioma before the operation, especially in young people, fibrosarcoma cannot be completely excluded. We should be alert to the possibility of intracranial fibrosarcoma if we found a tumor outside the brain that shows MRI enhanced scan was significantly enhanced without meningeal tail sign and MRS showed significantly higher Lip peak and inverted Lac peak.

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## Author Contributions

Chun Zhou: designed the study performed the research and wrote the paper.

Wenqian Jiang and Qingyu Wu: designed the study performed the research and analysed data.

Yongming Tan: performed the research and wrote the paper.

### Statement of Ethics

The subjects have given their informed consent and that the study protocol has been approved by the institute's committee on human research.

## References

1. Bisogno G, Roganovic J, Carli M, Scarzello G, Calderone M, et al. (2002) Primary intracranial fibrosarcoma. Childs Nerv Syst 18: 648-651.
2. Lee JG, Song SW, Koh YC, Cho J, Choi JW, et al. (2013) Primary intracranial fibrosarcoma presenting with hemorrhage. Brain Tumor Res Treat 1: 91-94.
3. Giridhar P, Mallick S, Haresh KP, Gupta S, Julka PK, et al. (2016) Intracranial fibrosarcoma treated with adjuvant radiation and temozolomide: Report of a case and review of all published cases. J Egypt Natl Canc Inst 28: 111-116.
4. Okeda R, Mochizuki T, Terao E, Matsutani M (1980) The origin of intracranial fibrosarcoma. Acta Neuropathol. 52: 223-230.

**5.** Wang CZ, Luo J, Sun W, Dai JD (2009) Fibrosarcoma Caused by Postoperative Radiotherapy for a Pituitary Adenoma Report of a Case and Review of the Literature. Journal of Capital Medical University. 30: 243-248.

**6.** Liu ML, Han T, Lei J, Guo J, Lian KX, et al. (2010) Intracranial primary fibrosarcoma: a case report. Chinese Journal of Radiology. 44: 220-222.

**7.** McDonald P, Guha A, Provias J (1997) Primary intracranial fibrosarcoma with intratumoral hemorrhage: neuropathological diagnosis with review of the literature. J Neurooncol 35: 133-139.

**8.** Tao YE, Ke LI (2009) Imaging Diagnosis of Meningioma and Analysis (+of) the Cause of Misdiagnosis. Chinese Computed Medical Imaging.15: 507-511.

**9.** Li H, Wang YJ, Wang JJ, Wang ZQ, Guo GH (2017) Combination of MRI and CT in the diagnosis of micro meningioma. Journal of Practical Radiology. 12: 1507-1509.

**10.** Han D, Chen YY, Jia YJ, Guang-ming MA, Ren CL, et al. (2016) One case: the intracranial hemangiopericytoma. Journal of Practical Radiology. 32: 1982-1983

**11.** Liu L, Xiao JH, Wei Y, Yang WJ (2011) MRI Diagnosis of Primary Central Nervous System Lymphomas and Pathological Features. Chinese Journal of Medical Imaging.19: 43-47.