Surgical Treatment of Primary Intracranial and Extracranial Communicating Leiomyosarcoma with One Case Recurred

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Case report

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Abstract

background

To analyze and summarize the clinical features and diagnosis and treatment experience on primary intracranial and extracranial communicating leiomyosarcoma to deepen clinicians'understanding of the rare disease.

Cases presentation

The clinical data and recurrence of a patient who was diagnosed with primary intracranial and extracranial communicating leiomyosarcoma admitted to the Neurosurgery Department, The Affiliated Ganmei Hospital of Kunming Medical University in May 2015 were retrospectively analyzed. According to the patient data, two successful surgical operations were performed and the surgeries went well, with more than 2-year and 8-month follow-up so far. No obvious complications were found after hospital discharge, and follow-up was continued.

Conclusions

Surgical removal is the most important and effective treatment means for primary intracranial and extracranial communicating leiomyosarcoma currently.

Background

Smooth muscular tumor is a rare malignant tumor derived from the tissue of the middle embryo layer, which usually has no obvious symptoms in the early stages, and the late symptoms mainly depend on the site and volume of the tumor. Primary intracranial-extracranial communication leiomyosarcoma can invade into the skull, and its anatomical structure is complex, which is very rare. The treatment method is generally determined according to the patient's age, tumor location, pathological characteristics and distant metastasis, including surgical resection, radiotherapy, chemotherapy and biological therapy. At present, surgery is the most commonly used treatment. In this paper, a patient admitted to the affiliated Ganmei hospital of Kunming Medical University who relapsed and re-operation after primary intracranial-extracranial communication leiomyosarcoma was regarded as the research object. Combined with domestic and foreign literature, its related clinical characteristics and diagnosis and treatment measures were analyzed.

Case Presentation

First admission

The patient, 37 years old, married, male, was admitted to the hospital for the first time on May 10, 2015 because of "a subcutaneous mass on the top of the left forehead was found in January". Main complaint: The subcutaneous mass gradually increased in January. Physical examination: The left frontal subcutaneous mass is about 4 cm×5 cm, flexible in texture, no tenderness to the touch, clear border, no redness, no skin ulceration, no obvious sense of movement, 0.7 cm higher than the normal skin margin, and no vascular murmur in the mass is heard. CT prompts: The local skull on the left forehead showed worm-eaten changes, and low-density shadows such as fusiform were seen inside and outside the skull. Part of the brain tissue is slightly compressed and moved inward, its internal density is not uniform, the CT value is about 38 HU, and the uneven edges are enhanced. MRI prompts: On the left frontal parietal bone, there is a fusiform T_1 long T_2 signal shadow, about 2.2 cm×3.5 cm×4 cm, growing around the skull, the corresponding skull absorption becomes thinner, and the bone destruction is seen in it, showing uneven low signal. After the enhancement, the position is abnormally and unevenly enhanced, and the delayed scan shows obvious enhancement, and the adjacent brain parenchyma is slightly compressed and moved inward (Fig.1).

Preoperative CT showed that the local skull was worm-eaten, and the outer side of the skull showed low-density shadows such as fusiform, and its internal density was uneven(a); Preoperative MRI (T1WI) signal uneven density shadow(b); Preoperative MRI (T2WI) is contour signal(c); Preoperative MRI enhancement showed ring enhancement(d); A re-examination of the brain CT showed that the tumor was completely removed one week after the operation(e).

Operation Procedure: During the operation, the left forehead mass was seen to be fish-like, tough and without obvious adhesion to the scalp. It was frozen during the operation. After completely exposing the tumor outside the skull, drill a hole in the skull, bite the skull along the tumor, and cut the dura mater in a circular shape. There is no adhesion between the tumor and the brain, and the tumor is completely excised (Fig.2); After intraoperative freezing indicated a malignant tumor, the surrounding normal meninges and skull tissue were visible to the naked eye about 2 cm, and the artificial dura mater and titanium mesh were repaired.

Pathological diagnosis: leiomyosarcoma; Irregular cells such as round shape can be seen in the tumor, the nucleus is abnormally enlarged, the nucleus is deeply stained and markedly atypia, a few vacuolar cells are seen, the cytoplasm is rich in eosinophilia, nuclear divisions are seen, diffuse and sheet-like, some interstitial mucinous degeneration(Fig.3); Silver staining (+), B lymphoma-2 (Bcl-2) (+), CD68 (scattered +), CD163 (scattered +), smooth muscle actin (SMA) (+), FLI-1 (+), FN (+), Ki-67 (about 30%+); The edge of the specimen is normal tissue.

Immunohistochemistry SMA(+)(a); Immunohistochemistry Ki-67(+)(b)

Postoperative follow-up: The patient was discharged from the hospital 9 days after extensive tumor resection. After the operation, the tumor was completely removed and the tumor was not sensitive to radiotherapy and chemotherapy, so radiotherapy and chemotherapy were not performed. Routine follow-up visits for 2 years and 3 months, no recurrence was found until August 2017, and no recurrence or distant metastasis was found during PET-CT examination. Two years and four months after surgery, a subcutaneous mass appeared on the left frontotemporal roof, and leiomyosarcoma recurrence was considered.

Second admission

The patient was re-admitted to the hospital on September 27, 2017 because of "the left frontotemporal parietal mass was found for 20 days". Physical examination: The original surgical scar can be seen at the top of the left frontotemporal area, and a subcutaneous mass about 3 cm in diameter can be palpable at the top of the left temporal top. The texture is flexible, no tenderness to the touch, clear borders, no redness, swelling and skin ulceration, and no obvious sense of movement, 0.5 cm higher than the normal skin margin, no blood vessel murmur in the mass was heard. Multiple enlarged lymph nodes can be palpable on both sides of the neck, with a maximum diameter of about 3 cm. CT prompts: The left frontal-temporal parietal bone is congenitally absent, and a wide basal segment with low-density shadows can be seen on the left top. The boundary is about 3.2 cm×1.3 cm, and the CT value is about 39 HU. MRI prompts: The left frontal mass and the frontal bone changed postoperatively. An irregular abnormal lesion was seen in the inner plate of the left parietal bone, showing a slightly longer T_2 signal with a length of 3.5 cm×1.6 cm×3.5 cm. After the enhancement, the enhancement is uneven and the space-occupying sign is obvious (Fig.4).

Preoperative CT wide basal segment low-density shadow, clear boundary(a); Preoperative MRI (T_1WI) soft tissue showed low density shadow(b); Preoperative MRI (T_2WI) is contour signal(c); Preoperative MRI enhancement showed uneven enhancement(d); A re-examination of the brain CT showed that the tumor was completely removed 1 week after the operation(e)

Operation process: During the operation, there was a 3 cm×4 cm mass attached to the three-dimensional titanium mesh on the top of the left forehead, which was not separated from the surrounding tissues, partial resection was performed. After removing the titanium mesh, the space occupied was seen at the top of the left temporal area, with a diameter of 4 cm, and a fish-like appearance with clear boundaries. Remove the artificial dura mater, tumor tissue and the affected skull at this site, and it was found that the tumor originated from the top meninges. 3 cm of skull, dura mater, periosteum and subcutaneous tissue were resected toward the top, and proliferative tissue, artificial dura mater, brain tissue, and skull were resected to the frontotemporal area 4 cm from the tumor. After the pathological biopsy of the resection margin showed negative, the resection range continued to be expanded by about 3-4 cm. At the same time, a 2 cm×5 cm skin flap at the site of the tumor was removed, and one-stage artificial dura mater and titanium mesh were performed.

Pathological diagnosis: leiomyosarcoma (WHO grade Ⅱ); In the tumor, irregular cells such as fusiform and round-like are seen, the nuclei are deeply stained and huge, and are significantly atypia, accompanied by a few vacuolar cells, mitoses are obvious, flaky and diffuse, and some interstitial mucinous degeneration (Fig.5); Silver immersion staining (+), SMA (+); no tumors were found in the surrounding tissues; no tumor cells were found in the neck lymph node needle aspiration cytology biopsy.

Postoperative follow-up: The patient was discharged 10 days after another extended resection. Leiomyosarcoma recurred without metastasis. No radiotherapy or chemotherapy was performed before or after surgery. Regular review after discharge. As of June 1, 2020, the patient has been reviewed for more than 2 years and 8 months without any obvious complications. Follow-up is continuing.

Discussion And Conclusions

Clinical features

Leiomyosarcoma is a rare malignant tumor that originates in mesoderm tissue. Its prevalent sites include the uterus, gastrointestinal tract and retroperitoneum. Leiomyosarcoma in the head and neck accounts for about 1% to 4% of the total leiomyosarcoma[1, 2]. Since the tumor is highly malignant and aggressive, most of it is caused by the metastasis of distant primary malignant tumors, mostly gastrointestinal and female reproductive system tumors, with high mortality [3]. Leiomyosarcoma that originates outside the skull is called extracranial leiomyosarcoma, and leiomyosarcoma that originates inside the skull is called intracranial leiomyosarcoma. Partial damage to the skull can lead to intracranial and extracranial communication lesions. Because this situation is particularly rare, patients are easily overlooked when they go to the doctor, so most of the patients have advanced to the advanced stage when the disease is diagnosed[4, 5]. The tumor can occur at any age without obvious gender differences. It is usually seen in individuals with weakened immune function[6], especially human immunodeficiency virus-positive patients carrying Epstein-Barr virus[7]. Clinically, the intracranial location of this tumor mostly occurs in supratentorial, and a very small part is found in the cerebellum or spinal cord. Symptoms and signs vary depending on the location of the tumor. Extracranial masses are usually used for treatment, while the common intracranial symptoms are headache, vomiting, and secondary epilepsy. This case was treated with a subcutaneous mass and there were no related intracranial symptoms.

Imaging and pathological features

Primary intracranial-extracranial communication leiomyosarcoma is very rare, mostly derived from meningeal mesenchyme, and has nothing to do with meningeal epithelial cells, it is a new type of tumor[8]. At present, there are no clear diagnostic criteria for it in clinical practice, and the diagnosis is mostly based on the imaging findings and pathological immunohistochemistry of the tumor. The CT findings of this tumor are mostly medium-sized soft tissue masses with clear boundaries, a small part of which can be seen infiltrating and growing, and the boundary is blurred; while the MRI findings are generally not uniform, some accompanied by cystic degeneration and necrosis, and often the edge of the enhancement is ring-shaped enhancement[9]. In this case, the leiomyosarcoma part of the lesion is clearly demarcated from the normal brain tissue, with only mild edema at the edge, and the internal signal is uneven. After the MRI is enhanced, the uneven enhancement is obvious, and there is a patchy non-enhanced area. Pathological examination: The intracranial-extracranial communication leiomyosarcoma is the same as the leiomyosarcoma in other soft tissues, and is mostly arranged in irregular spindle cell bundles. Under the microscope, the peripheral cell tumor-like structure can be seen in some parts, and the nucleus is slender and changeable, the color is dark, the mitoses are easy to see, the cytoplasm is acidophilic or lightly stained, and vacuoles are common.

Differential diagnosis

At present, primary intracranial-extracranial communication leiomyosarcoma does not have a unified standard for diagnosis, and the clinical symptoms are not specific. It belongs to extracranial neoplastic lesions, and generally requires related intracranial and extracranial lesions, such as benign meningioma, intracranial hemangiopericytoma, bone eosinophilic granuloma, skull osteosarcoma, glioma and fibrosarcoma, etc., are differentiated, see Table 1.

Tale 1 primary intracranial-extracranial communication leiomyosarcoma Related differential diagnosis criteria

Pathological features	Leiomyosarcoma[10]	Benign meningioma[11]	Intracranial hemangiopericytoma[12]	Eosinophilic granuloma of bone[13]	Skull Osteosarcoma[14]	Gliosarcoma [15]	Fibrosarco [16]
Age	Mostly in middle-aged people aged 40 to 60	Mostly in middle-aged people aged 40-50	Most common in people aged 30 to 45	Most commonly in children aged 5-10	Mainly in young people under 20	Mostly in middle-aged people between 40 and 60 years old	Mostly in young peo aged 20-4(
Gender	Mostly in female	The incidence of female is higher than that of male, and the ratio of male to female is 1:2	Slightly more common in male	The incidence rate is male: female 2:1	Higher incidence in female	More common in male, male to female ratio 2:1	Slightly mo common i male
Course time	Slightly shorter	Long	Slightly shorter	Long	Short	Short	Short
Skull changes	Partially visible erosive destruction of the skull	May involve the skull, causing local skull deformation	Mostly without bone destruction	Local osteolytic bone destruction	With significant bone destruction	Mostly without bone destruction	Bone destructior may occur when the skull is involved
CT findings	Low-density shadows, squeezing the brain tissue inward, obviously occupying the space, uneven enhancement appears after enhancement, clear boundaries, edema around the tumor is not obvious, some tumors can erode the skull, penetrating lesions, and occasionally calcification.	The round shape is slightly denser, and the surrounding boundary is clear. After the enhancement, the tumor usually shows a uniform and moderate enhancement, with obvious space-occupying signs and large differences in peritumoral edema.	Irregularly lobulated, with high, uniform, and low density on plain scan, unclear borders, common cystic degeneration and necrosis, peritumoral edema is not obvious, and uniform and significant enhancement after enhancement.	Obvious bone destruction can be seen, usually involving the inner and outer plates of the skull at the same time, with variable and irregular shapes, and osteolytic bone destruction with clear boundaries.	Irregular soft tissue masses are often accompanied by bone destruction. Tumor bones can be seen in the masses.	Mixed density shadow, with large lesions, involving multiple brain lobes, necrosis of the cyst can be seen in the focus, often accompanied by edema around the tumor, clear boundaries after enhancement, irregular and moderate enhancement	Round or lobulated mass with uneven density, fu border, ma be accompan by hemorrhag necrosis, cystic degenerati mild to moderate peritumora edema, uneven enhancem after enhancem
MRI findings	There are often uneven signal, accompanied by cystic degeneration, necrosis, and occasional bleeding; there is often marginal ring enhancement after enhancement	Isometric or slightly longer T_1 and longer or slightly longer T_2 signals, which are obviously and evenly enhanced after enhancement, showing "meningeal tail sign"	T_1WI and T_2WI showed equal and slightly low signal, necrosis of the cyst can be seen in the lesion, accompanied by empty vascular shadow, the tumor boundary is clear, and slight edema can be seen around the lesion	The lesions on T_1WI showed isointensity, the lesions on T_2WI showed slightly hyperintensity, with some dots and slightly lower signals, and T_2 Flair showed medium or high intensity, with uneven enhancement	T_1WI is mostly equal signal, T_2WI is mostly iso-equal high signal, and it is often clearly identified by the typical Codman triangle	T_1WI showed irregular long T_1 with slightly low signal, T_2WI showed mixed high signal, and showed uneven substantial enhancement after enhancement	T_1WI show low or equ signal, T_2 showed m confoundi signal, anc showed obvious fla enhancem after enhancem

Treatment and prognosis

At present, the clinical treatment of leiomyosarcoma is still mainly surgical excision, supplemented by radiotherapy, chemotherapy. In the early stage, the main treatment method is complete surgical resection, mainly to remove the tumor visible to the naked eye, and try to ensure that the intraoperative histopathology of the resection margin is negative. Then, expand the resection of normal tissue larger than 2 cm. In the advanced stage, the degree of tumor malignancy is generally high, the disease-free survival period is short, usually 6 to 24 months [17], and it is difficult to complete resection after surgery, so radiotherapy can be supplemented before or after surgery[18]. Since this type of tumor is not sensitive to radiotherapy and chemotherapy, radiotherapy and chemotherapy are used less frequently. Although preoperative or postoperative radiotherapy and chemotherapy can reduce the probability of recurrence and distant metastasis

to a certain extent, it has little effect on the survival and prognosis of patients[2]. Studies have shown that bevacizumab can treat some soft tissue sarcomas and is effective[19], but there is still a lack of clinical data to support primary intracranial-extracranial communication leiomyosarcoma. At present, the prognosis of the disease is extremely poor[20]. This case is primary intracranial-extracranial communication leiomyosarcoma, which has bone destruction. The effect of the first extended resection is satisfactory, and there is no sign of recurrence in 2 years and 3 months. After the recurrence, the patient was re-operated to ensure that the surgical margin was negative, and the resection range of the lesion was expanded by 3~4 cm. During the follow-up, it was found that there was no recurrence in 2 years and 8 months before publication. It can be seen that surgical expansion of the tumor is still the main treatment for the tumor. At present, there is no uniform standard for the scope of surgical resection. Because recurrence still occurs when the tumor is resected more than 2 cm, it is recommended to quickly complete the histopathological examination of the resection margin during the operation. If the result is negative, continue to expand the resection by more than 2 cm.

Abbreviations

CT
Computed tomography
MRI
Magnetic Resonance Imaging
SMA
smooth muscle actin
WHO
World Health Organization
HE
hematoxylin-eosin

Declarations

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Not applicable

Authors' contributions

Wang ZY reviewed the relevant literature and participated in the drafting of the manuscript. Pu KR and Zhang WC participated in the revision of the manuscript. Chen P and Shao DC participated in the information collection. Zhao N reviewed the manuscript and are contributors in writing it. The authors read and approved the final manuscript.

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Availability of data and materials

Not applicable.

Ethics approval and consent to participate

Not applicable as this is a case report, not a clinical study.

Consent for publication

Patient's consent for publication was obtained.

Competing interests

The authors declare that they have no competing interests or financial ties to disclose.

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Figures

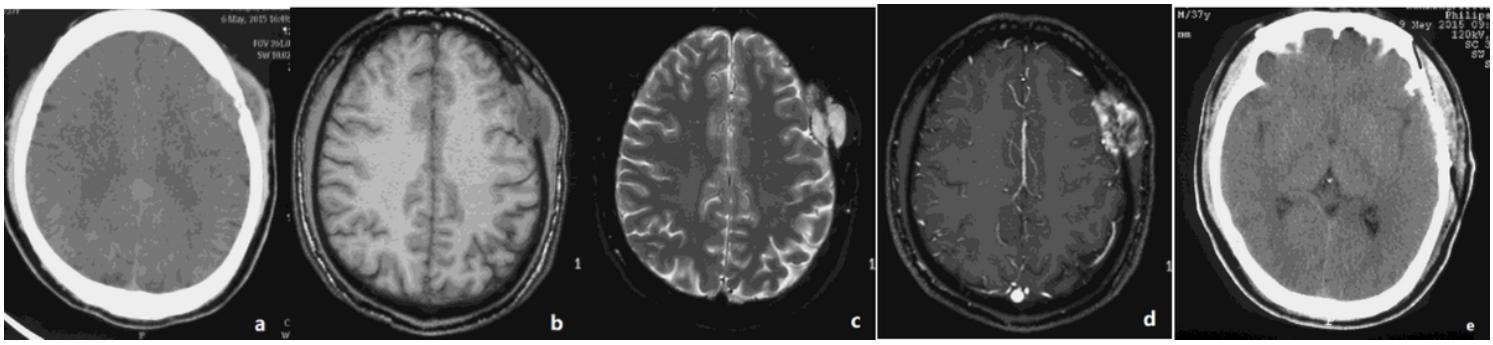

Figure 1

The first preoperative CT, MRI plain scan, enhancement and postoperative CT findings of the patients. Preoperative CT showed that the local skull was worm-eaten, and the outer side of the skull showed low-density shadows such as fusiform, and its internal density was uneven(a); Preoperative MRI (T1WI) signal uneven density shadow(b); Preoperative MRI (T2WI) is contour signal(c); Preoperative MRI enhancement showed ring enhancement(d); A re-examination of the brain CT showed that the tumor was completely removed one week after the operation(e).

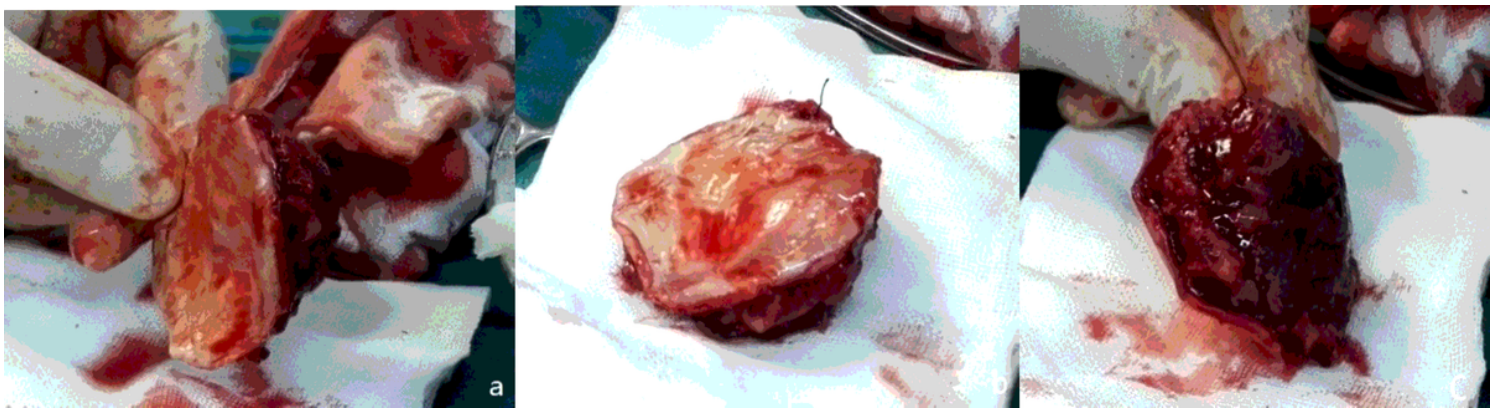

Figure 2

Leiomyosarcoma tissue (a), affected meningeal surface (b) and inner surface of the skull (c) removed during the first operation

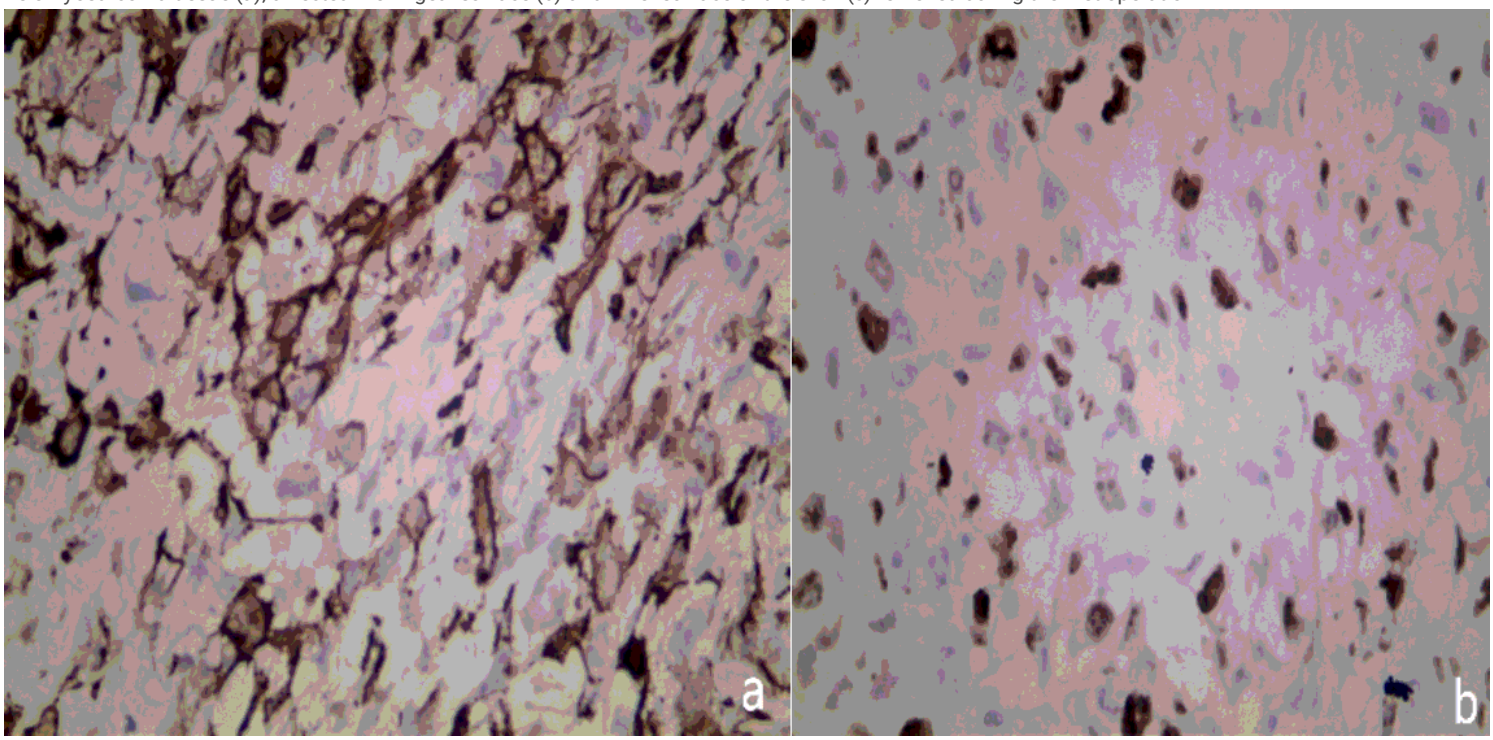

Figure 3

After the first operation, under the microscope, tumor cells were found to be irregularly arranged in a spindle shape, with enlarged nuclei, and some cells were vacuolated (magnification,×400). Immunohistochemistry SMA(+)(a); Immunohistochemistry Ki-67(+)(b)

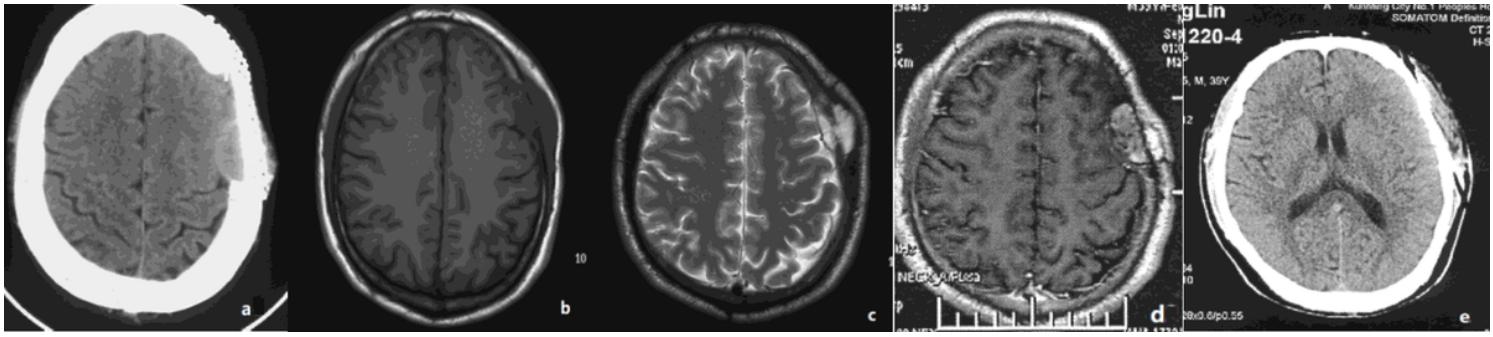

Figure 4

The second preoperative CT, MRI plain scan, enhancement and postoperative CT findings of the patients. Preoperative CT wide basal segment low-density shadow, clear boundary(a); Preoperative MRI (T1WI) soft tissue showed low density shadow(b); Preoperative MRI (T2WI) is contour signal(c); Preoperative MRI enhancement showed uneven enhancement(d); A re-examination of the brain CT showed that the tumor was completely removed 1 week after the operation(e)

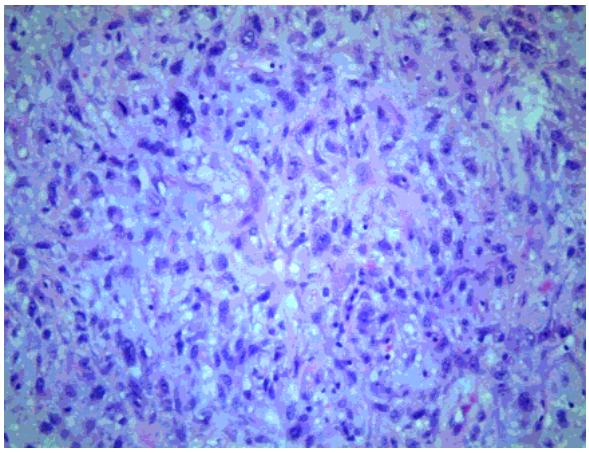

Figure 5

Under the second postoperative microscope, the tumor cell nucleus was deeply stained, markedly atypia, and mitotic figures (HE staining, magnification, ×400)