

Endoscopic surgical treatment with nasal cavity approach for chondrosarcoma of paranasal sinus and the skull base

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Abstract

Background Chondrosarcoma(ChSa) is a rare malignant tumor. But it's necessary to discuss the clinical characteristics and treatments for ChSa of paranasal sinus and the skull base. **Methods** The clinical characteristics of ChSa of paranasal sinus and skull base in 10 patients (6 males and 4 females) from 2001 to 2017 were analyzed. They all underwent by endoscopic surgery . The patients' age ranged from 18 to 47 years, with a median of 35 years. Clinical symptoms: stuffy, nose bleeding, runny, headache, diplopia, eye outreach limited, blurred vision and even blindness. Surgery methods—under nasal endoscopy, after the attachment sites of the tumors to normal tissues were confirmed, the tumors were peeled off along the clear boundary between the tumors and normal tissues, and the potential residual tumor tissues on bones were cleared by a drill. **Results** All patients were treated with endoscopic surgery— followed up postoperatively for 24 to 108 months, with a median of 36 months. 8 of 10 patients were no recurrence, 2 were alive with tumor. **Conclusion** ChSa of paranasal sinus and skull base can be treated by nasal endoscopic surgery with good clinical results.

Background

Chondrosarcoma(ChSa) is a rare malignant tumor composed of transformed chondrocytes and commonly occur within the axial or appendicular skeleton[1]. ChSa account for approximately 3 new cases per 10⁶ population per year[2]. There are three subtypes of ChSa: conventional, mesenchymal, and myxoid. The vast majority of ChSa is conventional ChSa and classified by the World Health Organization in three grades. Approximately 90% of these are low- to intermediate-grade tumors (grade I or II) which have indolent clinical behavior and low metastatic potential. Only 5–10% of conventional ChSa is grade III which can often lead to metastasis and aggressive local recurrence. The 5-year recurrent rate of grades I, II, III, mesenchymal, and myxoid subtype have been reported to be 15%, 16%, 33%, 63% and 16%, respectively, and the 5-year survival rates are 90–95%, 81–90%, 43–75%, 46%, and 94%[3, 4]. So the prognosis for the majority of patients with ChSa is favorable and correlates with histologic grade and adequate surgical margins[5].

About 10% of all ChSa is located in the head and neck. Most of them occur within skull base. Only a few cases of primary chordoma of the nasal cavities and paranasal sinuses have been reported in the literatures[6, 7]. The majority of skull base ChSa is low grade(50% of skull base ChSa was WHO Grade I tumour and 90% was WHO Grades I and II collectively) and exhibit an indolent growth pattern[8]. So most ChSa of the nasal skull base in its early stages is hard to be diagnosed, because its early clinical symptoms are not obvious. The tumor is typically erodes the outer cortical bone, lower cranial nerves and internal carotid artery(ICA)[6]. They are generally very slow growing and erode the bones of the skull base; however higher grade tumours occur, characterised by rapid growth and early [metastasis](#). not sensitive to radiotherapy and chemotherapy. The overall current consensus for initial treatment of ChSa is maximal safe surgical resection. Gautam et al. pointed out surgery is the primary therapeutic option and endoscopic endonasal approaches have been proposed for some skull base ChSa[9]. The extent of

tumour resection is very important to skull base ChSa prognosis. This was based on a literature review of multiple cases that showed an increased progression free survival with more extensive resections. Total or near total resection has good disease specific survival rates of 85–100% at 5 years[10-12]. However, surgery is at risk of not only removing the tumor completely but also damaging peripheral structure. Worse still, when it was diagnosed, the bone structure often has been damaged that made the anatomical structure more complex and the operation more difficult. To reduce the risk, we retrospectively reviewed 10 patients with endoscopic endonasal resection of skull base ChSa since 2001. The study was as follows.

Methods

2.1 Clinical data:

10 patients with skull base ChSa were enrolled in this study. They were all treated by endoscopic endonasal resection from 2001 to 2017, including 6 males and 4 females, aged from 18 to 47 years, with a median of 35.2 years. And the mean duration of follow-up was 43.8 months (range 24-108 months). Patient characteristics are reported in Table 1. Clinical symptoms of the patients: seven of ten patients were nose bleeding, runny, stuffy, four of ten patients were headache, two of ten patients diplopia, eye outreach limited, blurred vision, and one patient was even blindness. Clinical examination: tumor was gray or pink, easy bleeding, and occupying one side, bilateral nasal cavity and even the entire nasopharynx. Imaging examination: range of tumor invasion to the single or bilateral nasal cavity, anterior skull base, nasal septum, the entire nasopharyngeal cavity, maxillary sinus, ethmoid sinus, orbital paper plate, sphenoid sinus, anterior cranial fossa, bilateral eyes, bilateral optic canal, part of the ICA bone and slope area. In 3 of the 10 cases, tumor invaded anterior cranial base, and in the remaining 7 cases, tumor invaded middle cranial base. Besides, in 2 cases, tumor invaded ICA and optic nerve bone canal. No tumor invasion of cerebrum was observed during endoscopic surgery in all cases. Before operation, three of the patients were misdiagnosed of ossifying fibroma, fibrous dysplasia, fibrous vascular tumor, olfactory neuroblastoma and cartilage tumors in other hospitals' department of image and pathology.

2.2 Surgical methods

All cases with skull base ChSa were operated by endoscopic endonasal resection under general anesthesia.

First of all, we used the adrenalin gauzes to shrink nasal mucosa to reduce bleeding and used long gauze to push the tumor to the bottom of nasal cavity to check the situation of tumor. Then we cut all or part of

the middle turbinate and inferior turbinate. Inserted nerve dissector along the incision and separating it up and down while pushing the tumor towards to nasal cavity, used turbinate scissors to remove the upper and lower parts of the maxillary sinus wall, exposed the maxillary sinus internal angle, and used tissue forceps to trim the maxillary sinus medial wall. Besides we pulled the sliver back downwards to expose the front of the middle turbinate, used bipolar electrocoagulation to expose the bone at the front of the middle turbinate, and placing a wide white gauze in the incision of electrocoagulation. According to the differences among tumor tissue, normal soft tissue and bone texture, we used 5 mm suction as a nerve dissector, while sucking with a suction device and protected with a sliver, to peel the tumor back and down. We can also use tonsil dissector, while stopping the bleeding with electrocoagulation treatment, to peel the tumor back and down. The entire tumor, middle turbinate, and some normal ethmoidal tissues were dissected from the anterior skull base and sent to the posterior nostril, nasopharyngeal cavity, and even the oropharynx. Clear up soft tissue that adheres to normal skull base bone and papyraceous lamina. Examine the size of the anterior cranial bone destruction zone and the condition of the meninges carefully. High-speed drill was used to grind suspected invaded anterior skull base bone. Because the tumor grew expansively and the tumor boundaries are clear. When the base of skull bone was destructed, tumor is likely adhensive to endocranium. In order to reduce the meningeal hemorrhage when stripping off tumor, using bipolar electrocoagulation coagulate around the meninges which were adhere to the tumor. The tumor was dragged and spun off by vise and then removed. Tumors may invade other non-functional anatomy regions, such as papyraceous lamina, posterior part of nasal septum and the bottom of the junction between medial wall of maxillary sinus with the palate. We should removed the organizational structure as far as possible and removed tumor from the oropharynx, filling nasal cavity with iodine spinning.

If the tumor invade middle cranial fossa, such as the saddle area, clivus regions, ICA, optic canal and so on, in addition to the above-mentioned steps, we resected anterior wall of sphenoid sinus by drill or bone rongeur. The sphenoid sinus cavity and adjacent tumors were separated bluntly, such as twining the front part of the nerve dissector, peeling the tumor towards oropharynx and so on. We must pay attention to the direction of the ICA and the bony duct of the optic nerve, the anatomic site of the cavernous sinus, and the extent of bone erosion in the clivus, to avoid damage the important anatomical structure and major complications. When we were dealing with the invaded optic nerve or internal carotid arterial bone tube, because the tumor has a clear boundary with the normal tissue and the connection is loose, we can use medical gauze and nerve dissector separated them easily. Fragmented soft tissue strips could be slowly removed by an electric cutter. The direction of tumor separation should be consistent with the direction of nerves and arteries. When tumor has been detached and the optic nerve or ICA has been exposed, we could remove the surrounding tumor tissue with a small nerve dissector that wraps two to three layers of gauze on the head and then bited with the rongeur to remove the tumor gently. High-speed drilling was used to deal with parts of the suspected invaded bone were as far as possible to achieve contours or expanded contours, such as the optic nerve, ICA bone canal, clivus and pterygoid bone. After ICA bone canal was open, fascia was covered to protect artery. And then we must be careful not to damage the

joint capsule of atlanto-axis when dealing with the bone of the lower clivus. Otherwise, it would affect the stability of the skull.

2.3 Typical case

A male patient, aged 38, hospitalized for “repeated headaches, left eye pain with blurred vision four weeks” in April 2007. Clinical examination: left eye vision 1.0, right eye vision 1.5, bilateral eyes move normally, no external nose deformities, the forward edge of bilateral choana and the anterior wall of sphenoid sinus was ectasia and hyperemia. Bilateral middle and inferior turbinates were neither hypertrophy nor shrink. Magnetic resonance imaging (MRI) examination: an irregular massive lump could be found in plain cross-section T1WI, T2WI, and in sagittal plane, T1WI. The lump signal is not uniform, the boundary is clear, the edges are irregular and slightly lobulated. Signal of the lump shows slightly long T1, and equal T2 signal. The inside of the lump showed irregular long T1, long T2 signal liquefaction and necrosis area. Adjacent to the mass, the ethmoid bone, the bilateral orbit, the slope, and the skull base are compressed and absorbed. On both sides of the lump, medial rectus, optic chiasm, pituitary, ICA, top wall of nasopharynx, posterior turbinate and right maxillary sinus medial wall were under a little pressure. Enhanced scan shows non-uniform enhancement in heterogeneous lump, no enhancement in liquefaction necrosis. The tumor was about 3.6cm × 4.5cm × 3.8cm. No abnormal signal was found in the brain parenchyma. These MRI revealed: 1. The tumor located in ethmoid and sphenoid sinus is most likely to be olfactory neuroblastoma. 2. Paranasal sinusitis. Operation range: right around the ethmoidal cells, posterior nasal septum, sphenoid sinus, left orbital apex, the left optic bone pipe, clivus (Figure 1). Pathological diagnosis in other hospital was consistent with cartilage tumors. In February 2008, the patient was admitted to our hospital because of left eye vision and sharp decline in visual acuity for 10 days. His speciality examination showed that his left eye has no light perception, right eye vision is 0.2. White secretions can be observed in the left nasal cavity, with pale red neoplasia at the top of nasal cavity, upper septum, and nasopharyngeal top. Preoperative MRI T2WI scans showed a 5.2cm × 5.2cm × 6.3cm lobulated soft tissue lump in the anterior cranial fossa, ethmoid, sphenoid sinus and nasal cavity. The signal around the lump was high and the central signal was low. Bone had been destroyed in anterior cranial fossa, ethmoid, sphenoid sinus wall, nasal septum, bilateral orbital wall and upper middle turbinate. Normal tissue had been invaded in bilateral orbital and the right maxillary sinus (Figure 2). Range of operation: the former base of the skull, bilateral frontal margin around the tip of bilateral orbital, maxillary sinus, clivus and bilateral optic bone tubes. During the operation, it was observed that the bone of the right ICA was partially destroyed, the defect of anterior skull base bone was about 3.0 cm × 2.5 cm, and the endocranium was not damaged. The tumors that could be seen with the naked eye during the operation were resected and there was no residue in the postoperative radiological examination. (Figure 3). The surface of the tumor was as smooth as jade, while texture was like cartilage (Figure 4). Pathology diagnosis: well-differentiated ChS. After follow-up, recurrence was found in 32 months, and the patient has been living with tumor. (Figure 5).

Results

All 10 patients' tumors that could be seen with the naked eye during the operation were resected by endoscopic endonasal resection surgery and there was no residue in the postoperative radiological examination. Postoperative follow-up results of patients out of hospital from 24 to 108, with a median of 36 months, showed 8 patients were no recurrence and 2 patients were living with tumor. In which 8 cases of recurrence-free patients, one case had recurrence after conventional surgery and had two radiotherapy failures, then decided to accept endoscopic surgery. In the two patients who were living with tumor, one patient underwent the 2nd operation because of recurrence 8 months after the first surgery. After that, the patient's tumor recurred again 32 months after the first operation, and the tumor had violated the intracranial tissue. Therefore, the patient refused to undergo reoperation and were living with tumor now. In another case, recurrence occurred 15 months after endoscopic surgery, followed by 2 consecutive cranio-nasal approaches, and finally relapse was confirmed in October 2009. In this group of patients undergoing endoscopic surgery, intraoperative blood loss was 200 to 2000 ml. There was no ICA rupture, optic nerve damage, and cerebrospinal fluid leakage and other complications.

Conclusions

ChSa mainly occurs in the lower extremities, upper limbs, trunk and so on, rarely seen in the nose. In the head and neck region, it mostly occurs in the sphenethmoid, Spheno-occipital(clivus), temporal-occipital junction and other cranial cartilage[13]. Rosenberg et al[7] retrospectively analyzed the characteristics of 200 cases of skull base ChSa, including 12 cases of sphenethmoid(6%), 56 cases of Spheno-occipital (clivus) (28%),and 132 cases of temporal-occipital(66%). All cases are closely related to clivus. Due to the site is concealed, the early symptoms of nasal ChSa of skull base were not obvious. When the tumor was found, it had been large and destroyed the surrounding bone structure. The tumors showed expansive and aggressive growth, but its border is relatively clear. The imaging examinations of these 10 cases in the group were in accordance with the above. Skull base ChSa also has the characteristics of easy recurrence but rare metastasis. These 10 cases in this group came from multiple hospitals, some are bulky and repeatedly relapsing, some had long history. Nonetheless, no patient had metastasis.

For the diagnosis and differential diagnosis of nasal skull base ChSa, some scholars believe that ChSa has typical features in imaging, such as ChSa in the X-ray showed bone destruction, ambiguous lobulated translucent area, where irregular plaque-like calcifications, and the greater the number of calcifications are, the lower the malignancy of the tumor is. These features could help identify with other tumors. ChSa in CT plain scan showed a soft-tissue mass with equal or slightly lower density at the base of the skull with calcified or ossification of the plaque. The tissue between the calcified lesions showed equal or lower density with clear boundaries and some are connected to the base of the skull by a wide base. It can invade the inside and outside of the skull. The sphenoid, sphenoid sinuses and clivus are most often involved. After the injection of contrast agent, Non-calcified areas are slightly unevenly enhanced. In MRI T1-weighted image ChSa showed equal and low signal mixed shadows, and in T2-weighted image showed high and low mixed signals. The tumor borders are clear, and intratumoral calcification is a no-signal area. After the enhanced scan, the tumor was mildly heterogeneous and the calcified area was not enhanced[14]. However, ChSa is not easy to distinguish from chordoma in

imaging[15]. At the same time, we also need to differentiate it from craniopharyngioma, nerve sheath tumors, meningioma and so on. Three patients in this group were misdiagnosed pre-operation as diagnosis of cartilaginous tumors, angiofibroma, ossifying fibroma or fibrous dysplasia, neuroblastoma. This suggests that the tumor has certain characteristics in imaging, but because of the low incidence, imaging doctors lack of diagnostic experience, it is easy to misdiagnose; and if the tissue for pathology is not sufficient, it is also easy to mislead the diagnosis of pathology. Although radiotherapy may play a supporting role in the treatment of skull base ChSa, surgery is the most effective method to eradicate or prolong the patient's life because the tumors' character which rarely metastasizes, shows expansive and invasive growth, and has clear borders[1, 9, 16].

However, this tumor has the characteristics of easy recurrence, combined with its specially anatomic location. Therefore how to maximize the removal of the tumor under the premise of ensuring important anatomical structures is the goal pursued by surgeons and patients. Battaglia et al[17] considered that purely endoscopic endonasal technique may provide a minimally invasive and safe approach to radically resect selected tumors such as ChSa. However, with the developing of the scope of endoscopic surgical treatment, in skull base region of endoscopic sinus surgery, there is no need for additional head and face incisions, the field of surgical view is extended and multi-angled, "Four Hands" (the master surgeon and assistant co-operation) and other characteristics can be used in both sides of the nostrils. It creates a new way for the complete resection of skull base ChSa. Above all, micro-surgery could be better than open surgery. Zhang et al[18] reported for the first time in 2005 in China that nasal endoscopic surgery was used for the treatment of skull base ChSa, followed up for 6 months without recurrence. All 10 cases of patients in this group underwent endoscopic surgery. Although, the tumors were completely resected by postoperative imaging and visual observations, the follow-up results show that in this group of 2 patients with recurrence. Their tumors were largely invasive, especially invasive the area with complex anatomical structures such as important neurovascular and blood vessels crossing regions, suggesting that tumor growth sites that are too close to vital nerve vessels have a high recurrence rate. The reason for recurrence may be related to the insufficient use of surgical instruments. For example, failed to use a high-speed skull base drill to completely remove the non-functional anatomical structures adjacent to the skull base and the tumor, failed to achieve the contour or enlargement of the skull base, and failed to ensure maximum possible safety margins. Besides, in the area where the tumor and the meninges are adhering, it may be necessary to remove the tumor together with part of the meninges. Finally, it can also be related to the factors that the operator chooses to protect the patient's safety and avoid touching high-risk tissue structure. The follow-up results of this group of patients also confirmed that the surgeon should remove as large a range of tumors and surrounding tissues as possible to reduce non-visible tumor residues. At the same time, patients should be reminded that they must be closely followed up after surgery in order to deal with asymptomatic recurrence lesions as soon as possible. Of course, with the application of new equipment, the improvement of surgical operation techniques, and the accumulation of surgeons' surgical experience, the advantages of nasal endoscopic surgery in the treatment of skull base tumors are increasingly prominent.

Abbreviations

Chondrosarcoma=ChSa; internal carotid artery=ICA; Magnetic resonance imaging=MRI

Declarations

Declarations

The authors report no conflicts of interest. All authors agree to submit the manuscript, and the manuscript has not been submitted to other magazines at the same time. The authors alone are responsible for the content and writing of the manuscript.

Ethics approval and consent to participate

Not applicable.

Consent for publication

All patients gave written informed consent before participation in this study.

Availability of data and material

All data generated or analysed during this study are included in this published article.

Competing interests

The authors declare that they have no competing interests

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Authors' contributions

All authors participated in the elaboration of the manuscript. All authors read and approved the final manuscript.

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Tables

Table 1 Patient characteristics

Patients	Sex	Age [Y]	Location	Fellow- up Time [M]	WHO Grande	Clinical symptoms	invaded ICA
1	F	18	middle cranial base[ethmoid sinus][sphenoid sinus][slope area	108	0	stuffy, nose bleeding, runny	
2	M	41	middle cranial base[ethmoid sinus][sphenoid sinus][bilateral optic canal][slope area	30	0	stuffy, nose bleeding, runny[headache	
3	M	44	middle cranial base[sphenoid sinus][slope area	32	0	stuffy, nose bleeding, runny	
4	M	47	bilateral nasal cavity[anterior cranial base][maxillary sinus][bilateral optic canal	24	0	stuffy, nose bleeding, runny[headache][diplopia][blindness	√
5	F	32	the entire nasopharyngeal cavity[middle cranial base][sphenoid sinus][slope area	56	0	stuffy, nose bleeding, runny	
6	M	21	Single nasal cavity[middle cranial base][sphenoid sinus][slope area	26	0	stuffy, nose bleeding, runny	
7	M	37	nasal septum[anterior cranial base	78	0	stuffy, nose bleeding, runny	
8	M	47	ethmoid sinus[anterior skull base][optic canal	29	0	Diplopia[eye outreach limited, blurred vision	√
9	F	31	middle cranial base[ethmoid sinus][slope area	30	0	headache	
10	F	34	middle cranial base[sphenoid sinus][slope area	25	0	headache	

Figures

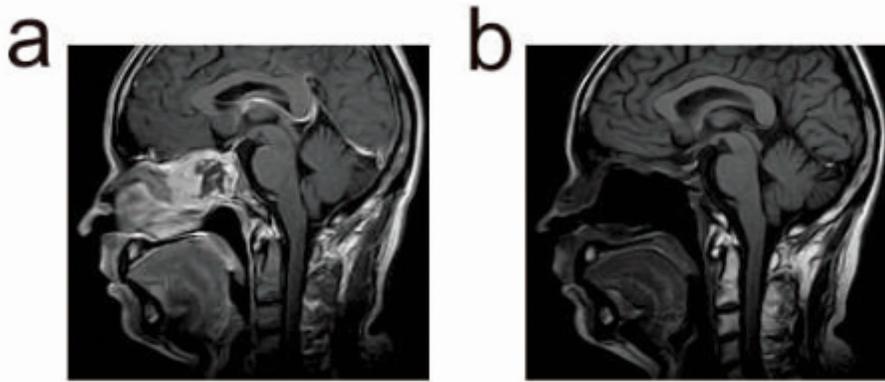


Figure 1

preoperative and postoperative MRI before the first nasal endoscopic surgery. a: preoperative MRI; b: postoperative MRI at 16 days.

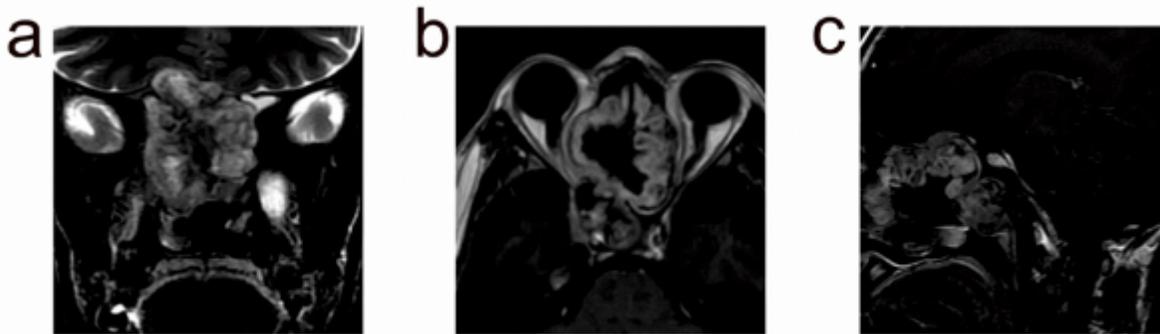


Figure 2

the patient's second preoperative MRI: there is a soft tissue mass in the nasal cavity with high signal and low central signal. The mass boundary is clear, and the anterior cranial fossa is involved. a: coronal scanning; b: horizontal scanning; c: sagittal scanning.

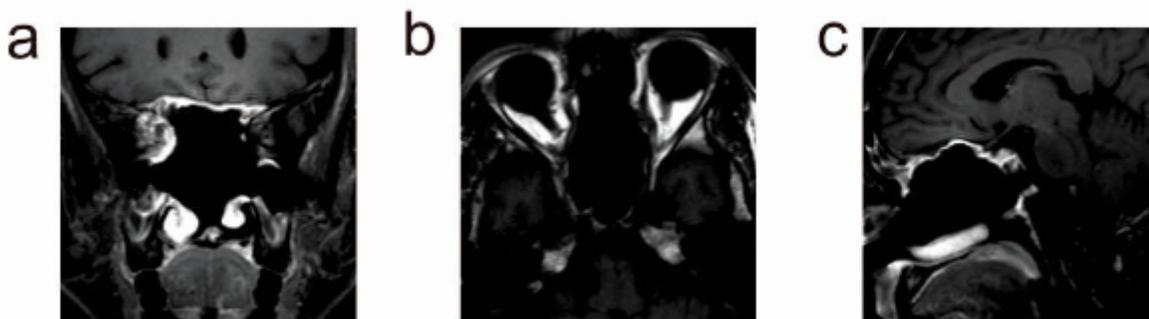


Figure 3

15days after the patient's second surgery. postoperative MRI image shows: The tumor has been excised and no tumor remains in the field. a: coronal scanning; b: horizontal scanning; c: sagittal scanning.



Figure 4

The appearance of chondrosarcoma.

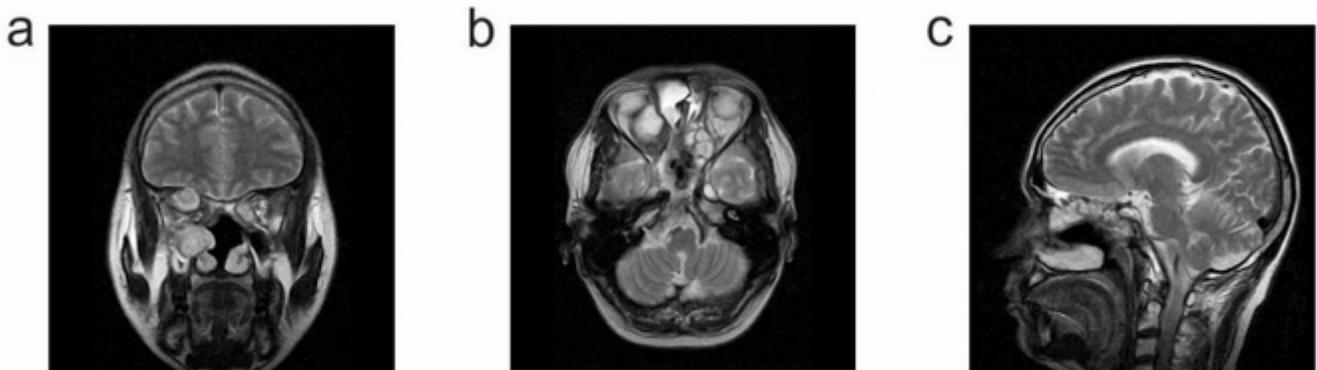


Figure 5

MRI after the 2nd surgery in 32 months reveals: the tumor had been recurred at the base of the fossa and the right maxillary sinus. In the right anterior cranial fossa, the lesion was prominent at the right orbit, and the left lesion was prominent at the left orbital medial wall. a: coronal scanning; b: horizontal scanning; c: sagittal scanning