

## An MDT treatment of extremely proptosis caused by the recurrent meningioma

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### Abstract

Meningiomas are the most common primary tumors of the central nervous system and are more common in female patients. Most meningiomas are benign, but the tumor infiltration can often lead to recurrence. We report a case of recurrent meningioma at the skull base in a juvenile, involving multiple sites including cranial and facial tissue, the prognosis is good after complete surgical resection.

### Keywords

Meningiomas; Recurrent; Surgical resection; Reconstruction.

### Abbreviations

MDT: Multi-Disciplinary Team; RAPD: Relative Afferent Pupillary Disorder; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; WHO: World Health Organization

### Introduction

Meningiomas originate from the meningeal covering of the central nervous system and are the most common primary tumor of the central nervous system with an annual incidence of about 5 per 100,000 people. Meningiomas are usually benign and rarely have malignant manifestations [1,2].

Meningiomas are more common in women (sex ratio 2-4:1) and the incidence increases with age [3].

Its clinical manifestations vary depending on the site of involvement and the degree of tumor infiltration. Surgery and radiotherapy are currently accepted first-line treatments, but combined treatment of tumors should also be considered in refractory cases[3].

## Case Presentation

A 16-year-old girl presented to us because of a huge tumor on the face for 8 years and decreased visual acuity in both eyes for 2 years. The patient had a history of nasal cranial communication tumor resections twice 10 years ago, and the postoperative pathological diagnosis was meningioma.

Ocular examination showed the tumor size was about 10 x 10 cm, with granular hyperplasia on the surface, accompanied by increased hair and pigmentation; The tumor is located in the middle of the face and the entire alar of the nose is completely destroyed (Figure 1); light perception was exist only in the left eye; proptosis cannot be measured due to the huge tumor size; eye movement are severely restricted in all directions in both sides; the inferior bulbar conjunctiva of both eyes is highly edematous and prolapsed outside the palpebral fissure; the corneal epithelium has a large defect area and the stromal layer is infiltrated in both eyes; the anterior chamber is normal and bilateral pupil diameter were about 5-6 mm with RAPD+ and fundus details were blurred in both sides.

CT and MRI showed that the tumor involved the anterior skull base, sphenoid and the ethmoid sinus, and invaded the inferior orbital wall, the pressure on the tumor caused the protrusion of the eyeball, and the optic nerve was pulled into a straight line.

The patient was a child with low body weight; the huge recurrent tumor has a wide range of involvement; the tumor has a rich blood supply, and the risk of anesthesia caused by intraoperative hemorrhage is high; it is difficult to determine whether the eyeball can be retracted after tumor resection; if the tumor can be completely removed, then how to reconstruct the huge defect area was faced by the surgeon.

The surgical approach for this patient was designed by the MDT multidisciplinary consultation: the otolaryngologist removed the main tumor in the middle of the face and the nose, the orbital surgeon removed the orbital-infiltrating part, loosened its adhesions, and retracted the eyeball and bulbar conjunctiva into the orbit, then palpebral fissure was sutured (Figure 2). The neurosurgeon continued to remove the basicranial tumor. The defect area was reconstructed with a 3D-printed titanium mesh (Figure 3), and the skin and subcutaneous tissue were reconstructed with a free vascular pedicle flap from the lower leg to anastomose with the facial artery (Figures 4,5). The postoperative pathological diagnosis was the same as the previous - it was recurrent meningioma. HE staining showed that the tumor had invaded the skin, skin appendages, striated muscle, and lymph nodes. Immunohistochemical staining showed CD34(-), EMA (+), GFAP (-), Ki67(+,5%), PR (+), S-100(+), SSTR2(+), STAT6(-), VIM (-), CD21(-), CD35(-) (Figure 6).

The long-term follow-up showed that the skin flap survived well, and the visual acuity improved to LP in one side and HM in other side.

## Discussion

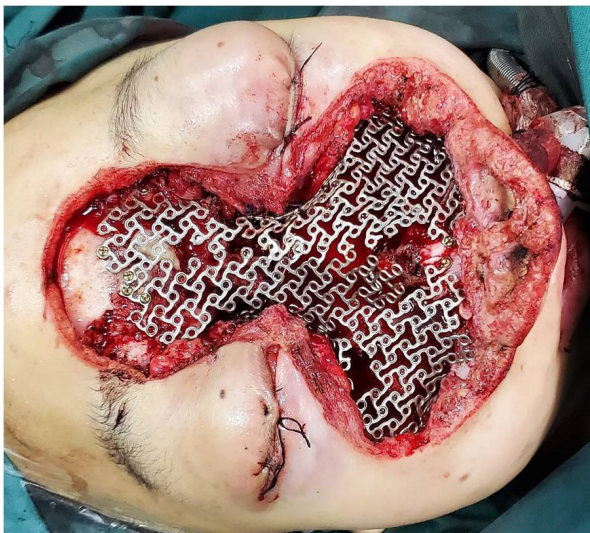
Meningiomas are often diagnosed due to neurological symptoms (neurological deficits, seizures, increased intracranial pressure) or tinnitus or headache. Magnetic Resonance Imaging (MRI) is often used to make an initial diagnosis and to precisely locate and measure tumor size. The currently identified risk



**Figure 1:** The appearance of the patient before the surgery.



**Figure 2:** Part of the excised tumor morphology during the surgery.



**Figure 3:** The implanted titanium mesh during the surgery.



**Figure 4:** Appearance of the patient immediately after surgery.

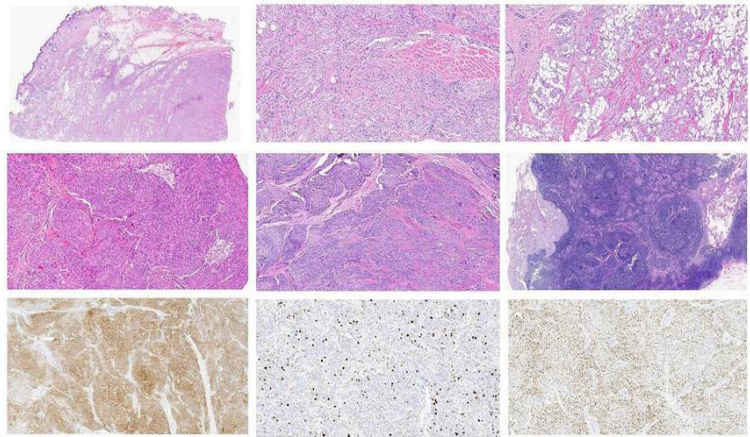
factors associated with meningioma development are radiation therapy and hormone intake (cyproterone acetate), and both have a dose-response relationship. Studies show that stopping hormone therapy intake even reduces tumor size [4-6].

According to the 2000/2007/2016 WHO classification of meningiomas into 15 subtypes and various invasive criteria (mitotic, necrotic, cellular aspect) based on their histological appearance [7], Overall, more than 80% are grade I benign tumors [3] while atypical grade II includes 4-15% of meningiomas, and malignant grade III accounts for 1-3% [8]. While histological classification is currently the gold standard for the diagnosis and treatment of meningioma, there is still much debate about its relevance. Tumors are very heterogeneous within each grade moreover, some diagnostic criteria are vaguely defined and subject to a high interobserver bias [9-11]. The 5-year survival rate is 70% for benign meningiomas and 55% for malignant meningiomas [12], and prognostic factors include age, male gender, low Karnofsky performance status, high grade, high mitotic rate, subtotal surgical resection, and optic nerve involvement[13-16].





**Figure 5:** Appearance of the patient four months after surgery.



**Figure 6:** Pathological manifestations of the tumor. HE staining showed that the tumor had invaded the skin, skin appendages, striated muscle, and lymph nodes [1-5]; Immunohistochemical staining showed EMA (+), GFAP (-), PR (+), SSTR2(+) [6-9].

The classic first-line treatment for all meningiomas is surgery. However, a wait-and-see strategy is acceptable when the clinical situation permits, but requires regular observation of tumor changes [17]. Symptomatic treatments including oral or intravenous steroids help temporarily relieve symptoms by reducing peripheral edema [18]. Complete tumor resection is not always achieved intraoperatively due to tumor location and invasion of surrounding structures and brain parenchyma. Radiation therapy has become the first-line option for skull base lesions encasing vascular nerve structures such as the optic nerve sheath or cavernous sinus. If surgery is not feasible, radiation therapy may be provided alone [19]. Grade I meningiomas are usually treated with surgery or radiosurgery only, with adjuvant radiation therapy used to treat tumor remnants [20]. The 5-year recurrence rates of grade II and III meningiomas are as high as 30-40% and 50-80%, respectively [10,14]; adjuvant radiation therapy to the tumor area is also recommended along with gross resection [21]. side effects of radiotherapy and radiosurgery are usually mild but there is also evidence that radiation increases the risk of malignant transformation [22].

Long-term follow-up studies have shown that even in so-called completely resected tumors, up to 60% of tumors may recur after 15 years [10]. Various pharmacological treatment regimens have been tried, but none have been proven to be effective. Attempts at targeted therapy (anti-angiogenic molecules such as sunitinib or bevacizumab, immunotherapy or FAK inhibitors) depending on the molecular characteristics of the tumor may lead to new therapeutic directions [23,24].

## Conclusion

Meningioma is the most common neurological tumor, and its treatment options and prognosis vary greatly according to different pathological features, and complete surgical resection can reduce the probability of its recurrence. A personalized treatment plan based on the patient's clinical presentation, imaging, and pathological features should be recommended.

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