

Original Article

An endoscopic endonasal prelacrimal recess-maxillary sinus approach for giant schwannoma of the pterygopalatine fossa in children

Zhihua Yin¹, Guangke Wang¹, Hongjian Liu¹, Bing Zhou²

¹Department of Otolaryngology, The People's Hospital of Henan Province, Zhengzhou 450003, China; ²Department of Otolaryngology Head and Neck Surgery, Beijing Tongren Hospital, Capital Medical University, Beijing 100730, China

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Abstract: Primary tumors of the pterygopalatine fossa in children are rare, and most are schwannomas. These types of tumors usually present with vague and nonspecific radiologic characteristics and it's difficult to diagnose preoperatively. The radiologic characteristics of different kinds of tumors were discussed and analyzed. Magnetic resonance images combined with CT angiography can provide very important information to confirm tumor characteristics before operation, especially enhanced MRI. The pterygopalatine fossa is so deep that it's challenging to reach through traditional operative approaches, and typical approaches may also cause great trauma. Endoscopic endonasal micro-invasion approaches have gradually been adopted for lesions of the pterygopalatine fossa, including those through the pterygoid process, the middle meatus, the middle meatus combined with a Caldwell-Luc incision, and the prelacrimal recess-maxillary sinus. A prelacrimal recess-maxillary sinus approach has the advantages of being short and direct, causing less trauma, offering safety and face protection, reserving the nasal structure and function, being easy to repair and review, and allowing for a second operation.

Keywords: Prelacrimal recess-maxillary sinus approach, schwannoma, children

Introduction

Primary tumors of the pterygopalatine fossa are rare, and most of them are fibroangiomas and neurogenic tumors. The majority of neurogenic tumors are schwannomas, which often originate from the ganglion pterygopalatinum and Schwann cells of the maxillary nerve. Schwannomas of the pterygopalatine fossa have a slightly higher tendency to be found in males and during the 3rd to the 4th decade of life. Neurilemmomas are benign and slow growing tumors. Typical clinical symptoms usually are not found in early stages until pressure symptoms appear. Patients usually present with swelling of the cheek, protopsis, numbness of the face, epistaxis, nasal obstruction, and diplopia. The following case highlights an effective surgical excision for schwannoma of the pterygopalatine fossa and reviews its radiologic characteristics and operative approaches.

Patient information

History and Examination. In May 2013, a 10-year-old boy presented with a two-year history of diminution of vision and a six-month history of protopsis in the left eye. He was now blind in his left eye. A computed tomography (CT) scan obtained at an outside hospital revealed a large mass in the left maxillary sinus and the pterygopalatine fossa, which prompted a transfer to our department for further evaluation. Upon physical examination, the child had protopsis of the left eye, normal findings in the nasal cavity, and normal eye movement and light reflexes; there were no symptoms of facial numbness, epistaxis, nasal obstruction, fever, headache, or loss of olfactory sensation.

Radiologic characteristic

CT angiogram (**Figure 1**) and magnetic resonance imaging (MRI) (**Figures 2-4**) were obtain-

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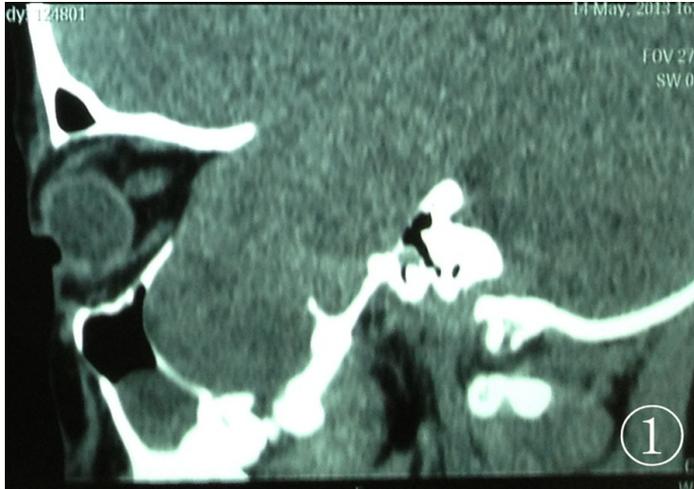


Figure 1. CT scan showing extension of the tumor into the pterygopalatine fossa.

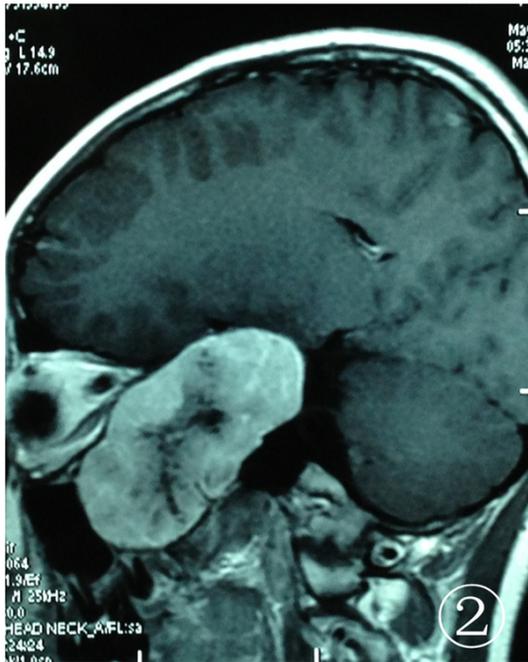


Figure 2. Sagittal section of an MRI scan revealing the tumor occupying the pterygopalatine fossa.

ed to further evaluate the lesion and its vascular supply. The CT angiogram showed a giant irregular mass in the left pterygopalatine fossa with a smooth edge. Nearby sclerotin was pressed and absorbed. The infratemporal fossa, left cavernous sinus, and optic canal were involved with the shifting contents in the orbit and proptosis. The mass had an iso-signal intensity on MR T1 WI and heterogeneous hyper-signal intensity on MR T2 WI. After con-

trast administration, the lesions showed inhomogeneous enhancement and a sharp edge with the surrounding tissues. These studies disclosed erosion of the tumor through the pterygopalatine fossa into the left maxillary sinus and infratemporal fossa. The lesion was diagnosed as a schwannoma derived from the pterygopalatine fossa.

Operation process

An endoscopic endonasal prelacri-mal recess-maxillary sinus approach was adopted. The mucous membranes near the adhering portion of the inferior nasal concha on the lateral wall of the nasal cavity were cut open and peeled under the subperi-

osteum. The adhering edge of the inferior nasal concha and the bony nasolacrimal duct were exposed. After severing the adhering edge of the inferior nasal concha and the bony lateral wall of the nasal cavity, a mucosal nasolacrimal duct-inferior nasal concha flap was formed, and the maxillary sinus was exposed.

Nasal endoscopy revealed a bulge of posterolateral wall of the maxillary sinus. The local mucous membrane and the bony posterolateral wall of the maxillary sinus were peeled, and then the pterygopalatine fossa was exposed. The tumor was peeled layer-by-layer, debulking it gradually, until the tumor was resected completely. The basilar part of the tumor was located near the cavernous sinus. Adequate hemostasis was obtained, and then the operation cavity was cleaned. The mucosal flap of the nasolacrimal duct-inferior nasal turbinate was replaced, and an inferior antrostomy was performed. An aqueous capsule was placed into the operation cavity to achieve hemostasis by compression.

Postoperative condition

The patient had no complications of nerve injury = or cerebrospinal fluid rhinorrhea after operation. However, his eyesight had not improved. Imaging on the third day following surgery (**Figure 5**) confirmed total tumor resection without any residuals. Histopathological examination revealed that the tumor cells were somewhat spindle shaped and arranged in clusters, which confirmed the lesion as a schwannoma.

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Figure 3. Coronal section of an MRI scan displaying the tumor extension into the floor of the orbit.

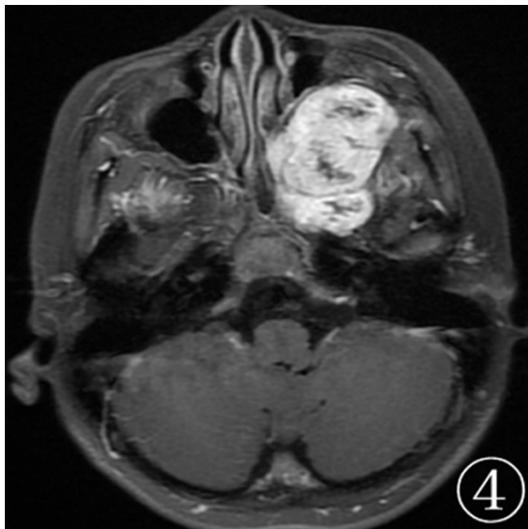


Figure 4. Axial section of an MRI scan showing bowing of the posterior wall of the maxillary sinus.

The child was discharged home; follow-up visits demonstrated a normal eye ball and a well-healed nasal mucosa.

Discussion

Radiologic characteristics of tumors in the pterygopalatine fossa

Tumors in the pterygopalatine fossa are rare and lack typical radiologic characteristics. It's difficult to diagnose these types of tumors be-



Figure 5. Postoperative MRI scan indicating total tumor resection without any residual.

fore surgery. Primary tumors include schwannomas, angiofibromas, etc., while secondary lesion can be metastatic carcinoma, adenoid cystic carcinoma, meningioma, etc. Confirming the tumor's characteristics preoperatively can provide great assistance to planning the surgery, determining the scope of the surgery, choosing an approach, and evaluating the prognosis. MRI combined with a CT angiogram can offer vital information for confirming the tumor's character before operation, especially enhanced MRI [1, 2].

CT angiograms of schwannomas show expansion of the pterygopalatine fossa, pressed and absorbed sclerotin, a clear border, homogeneous density, inhomogeneous enhancement after contrast administration and, rarely, calcification. The invasion can be located anywhere between the mass and the peripheral space. The mass showed iso-signal intensity on MR T1 WI and heterogeneous hyper-signal intensity on MR T2 WI. After contrast administration, the lesions exhibited inhomogeneous enhancement. Therefore, MRI has great diagnostic significance.

Angiofibromas have an irregular shape on CT scans. The mass showed hyper-signal intensity on both MR T1 WI and T2 WI with a local

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empty signal. Adenocystic carcinoma also has an irregular border and shape on CT scanning. Sclerotic damage can usually also be found. It has a hypo-signal intensity on MR T1 WI and a heterogeneous hyper-signal intensity on MR T2 WI. Meningiomas can have a hardened edge appressing to the middle cranial fossa. They exhibit iso- or hypo-signal intensity on MR T1 WI and iso- or hyper-signal intensity on MR T2 WI. Enhancement scanning showed a moderate to severe degree of homogeneous enhancement.

The child had typical radiologic characteristics of schwannomas. The diagnosis was formed as a benign lesion in the pterygopalatine fossa preoperatively, which was confirmed by histopathological examination. We prudently chose an appropriate operative approach after comprehensively evaluating the lesion.

Selection of operative approaches of tumors in the pterygopalatine fossa

The pterygopalatine fossa is so deep that it's difficult to reach through common operative approaches. It's challenging for surgeons to thoroughly resect tumors while also preserving organ function as much as possible. Classic approaches involve anterior and lateral approaches. Most lateral approaches are made through the infratemporal fossa to expose the anatomical structure of the pterygopalatine fossa and the infratemporal fossa. These approaches allow access to the jugular vein, internal carotid, and any nearby lesions, but they also carry a high risk of facial paralysis, hearing loss, and occlusion disturbance. Anterior approaches include maxillary, middle face, and nose-side incisions [3, 4].

These approaches may well deal with lesions of the pterygopalatine fossa, but they have a high risk of injury to the face, facial nerve, and infra-orbital nerve. For benign lesions of the pterygopalatine fossa, anterior and lateral approaches offer a wide operative field but also may result in great operative trauma and many sequelae. These typical approaches should not be used in children. Following the recent development of endoscopic technology, endoscopic endonasal micro-invasion approaches have been gradually adopted for lesions of the pterygopalatine fossa, including through the middle meatus, the middle meatus extended Caldwell-Luc inci-

sion, and the prelacrima recess-maxillary sinus [5-10].

This patient was a 10-year-old child and his giant lesion had been evaluated as a benign schwannoma that originated from the pterygopalatine fossa preoperatively. Therefore, for resection, we chose an approach through the prelacrima recess-maxillary sinus. Zhouing et al. reported this approach for the first time [11]. This method has been largely used for lesions of the maxillary sinus, and it is only rarely adopted for pterygopalatine fossa lesions. It has the advantages of being short and direct, causing less trauma, offering safety, protecting the face, obstructing no important structures, being easy to repair and review, and providing the opportunity for a second operation when needed. In this approach, only the inferior nasal concha and the bony lateral wall of the nasal cavity are resected, which partly preserves most of the nasolacrimal duct and turbinate, maintains the structure and function of the nose, and offers decreased morbidity and shorter recovery periods compared with other approaches. As a result, for the benign lesions of the pterygopalatine fossa, we should select micro-invasion approaches whenever possible and especially in children, even if the tumor has to be resected in stages due to haemorrhage.

In contrast, this approach for lesions in the pterygopalatine fossa has the disadvantage of providing limited operative space and being inconvenient for tumor separation and hemostasis. Surgeons who attempt this technique should have sophisticated endoscopic technology and extensive local anatomic knowledge. When the lesion is large enough to involve the infratemporal fossa, involvement with the jugular vein and internal carotid may also pose difficulties. The "double-persons and four hands" operating pattern has been put forward by some scholars. However, this pattern requires long training times and cooperation between the surgeon and the assistant, but lesions on the left side pose much difficulty. Therefore, an endoscopic endonasal prelacrima recess-maxillary sinus approach can not replace the typical approaches completely. The lesion must be comprehensively evaluated preoperatively to select the best approach and indication. In special patients, such as children and those with a benign lesion in the pterygopalatine

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fossa, this approach can be adopted. Omni-range imaging can also be used when necessary or safety and to allow the use of micro-invasion. The cases should be chosen discreetly, such as for malignant lesions, those that involve important vessels and nerves, or lesions that require reconstruction of a wide range of the skull base.

Schwannomas have a complete diolame. In the past, both the diolame and the tumor were fully resected in these cases. However, for lesions in the pterygopalatine fossa, nearby important vessels and nerves make endoscopy difficult, and uncontrolled hemorrhaging often occurs. However, it is safe to resect the lesion within the diolame, as we did in this case, without complication either during or after the operation. Magnetic resonance images following surgery only showed an intensified diolame. The rate of relapse has not been subjected to systematic research or any unified conclusion by reserving the diolame. There have been several cases of resection of schwannomas with the diolame reserved by Zhou Bing et al. No relapse was found on follow-up visits more than six years postoperatively. Nevertheless, the long-time safety of this pattern should be researched and discussed in further studies.

Conclusions

Tumors in the pterygopalatine fossa lack typical radiologic characteristics. MRI combined with CT scanning can provide vital information to confirm the tumor's character preoperatively, especially enhanced MRI.

An endoscopic endonasal prelacrima recess-maxillary sinus approach can be adopted for pediatric patients or those of any age with a benign tumor in the pterygopalatine fossa. This approach provided useful access to this cryptic anatomical region while also ensuring a satisfactory cosmetic result. Comprehensive evaluation prior to surgery is vital. In addition, further discussion of diolame reserving or resecting is needed.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Zhihua Yin, Department of Otolaryngology, The People' Hospital of Henan Province, No. 7 Weiwu Rd, Zhengzhou

450003, Henan, China. Tel: +86-0371-65580014; Fax: +86-0371-65964376; E-mail: Y_zhih@163.com

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