



Endoscopic Endonasal Removal of a Large Petrous-Apex Epidermoid Tumor With Clival Involvement and Intradural Invasion

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ABSTRACT

Introduction: Epidermoid tumors comprise 1% of intracranial tumors. Although reported, intraosseous epidermoid tumors are even more rare. Cystic lesions of the petrous apex are uncommon and surgically challenging; the most rare pathology is presumed to be epidermoid.

Case Presentation: This is a case of a 61-year-old woman with a large skull-base tumor extending inferiorly from the C1-C2 articulation and superiorly to the tuberculum sellae. The lesion replaced the left-sided petrous apex, and the lateral extension of the tumor reached the styloid process. The tumor displayed intradural invasion medial to the internal auditory meatus, producing an intra-axial mass at the level of the upper pons. The patient presented with a 4-month history of headache and total unilateral deafness. Under image guidance, an endoscopic endonasal approach was used to totally resect the tumor. By following the tumor's dural defect, the intra-axial part of the tumor was safely resected, and the dural defect was successfully repaired.

Conclusions: Image-guided endoscopic endonasal surgery is a versatile approach that can safely and easily address a large epidermoid tumor in this challenging region, obviating the need for demanding and sophisticated transcranial surgery.

► Implication for health policy/practice/research/medical education:

In addition to rarity of this pathology, it is worth to explain to neurosurgeons and ENT- surgeons the versatility of this minimally invasive approach to resect such a vast extended tumor in the petro-apex region.

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1. Introduction

Intracranial epidermoid cysts have an outer capsule comprised of connective tissue, which surrounds a layer of keratinized stratified squamous epithelium. They are relatively uncommon benign tumor-like lesions with oc-

currence rates of 0.2%-1.8% among primary intracranial tumors (1-3). Most epidermoid cysts are relatively large intradural tumors located in the posterior fossa at the cerebellopontine angle (CPA) with an insidious growth pattern. Extradural epidermoid cysts are more rare, but show the same slow growth pattern, allowing them to reach a considerable size before diagnosis due to neurological deficits (1, 4). Epidermoid tumors can be found in any part of the skull, but most commonly occur in the CPA, parasellar region, and in the petrous bone, which can be congenital or acquired from chronic ear infec-

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tions (5-8). Some reports have identified these cysts in uncommon locations, such as supratentorium and intradiploic, which represent less than 0.25% of all primary intracranial tumors (5-8). In addition, in 2006, Fassett *et al.* reported a case of a clival epidermoid cyst (9). Regardless of their location, most of these epidermoid tumors are benign; however, there have been reports of a few cases with malignant transformation at the same site of previous benign ones, such as the CPA (10, 11). Radical surgical excision is the first-line curative treatment for epidermoid cysts; however, removal of the capsule is difficult during surgery due to its adherence to adjacent structures, such as cranial nerves, vascular structures, and the brain-stem pia mater (12, 13). As reported previously, only 50%-80% of intracranial epidermoid tumors can be completely removed (14). In contrast to traditional cranial-base open surgeries that require extensive cranial-base removal and neurovascular manipulation, endoscopic endonasal transsphenoidal surgery offers a direct and minimally invasive approach that prevents iatrogenic brain retraction (3, 15, 16). Here, we describe the ability of the endoscopic endonasal transsphenoidal approach, which is a midline approach, to allow access to a large epidermoid tumor located at the center of the skull base and the craniovertebral junction with considerable superior, inferior, and lateral extensions.

2. Case Presentation

A 61-year-old woman presented with a 4-month history of left-side frontal headache and periorbital pain in addition to facial pain and hearing loss. She had experienced vomiting and vertigo for 1 month before admission. She had a history of chronic obstructive pulmonary disease and coronary artery bypass grafting. Her neurological examination revealed significantly diminished sensation at the left-sided V1, V2, and V3 dermatomes. Pure-tone audiometry showed total deafness of the left ear. A computerized tomography (CT) scan revealed a large petroclival lesion that completely replaced the clivus and left petrous apex (Figure 1A). Magnetic resonance imaging (MRI) showed a mass with a hypointense signal and rim enhancement around its capsule in T1W images and a hyperintense signal in T2W images that extended inferiorly to the C1-C2 articulation and superiorly to the tuberculum sella (maximum superior-inferior diameter of 6 cm). The tumor laterally extended to the styloid pro-

cess (maximum diameter of 5 cm on axial images) and showed obvious intradural invasion medial to the internal auditory meatus, producing an intra-axial mass at the level of the upper pons (maximum anterior-posterior diameter of 4.3 cm) (Figure 1B and C). These image findings confirmed the diagnosis of an epidermoid cyst. In some cases, the signal intensity difference between an epidermoid cysts and cerebrospinal fluid (CSF) is indistinguishable, making the differential diagnosis between an epidermoid cyst and an arachnoid cyst quite difficult using conventional spin echo images. In this case, the use of proton density imaging can be useful. In the proton density view, the epidermoid tumor showed an isointense signal to brain (Figure 1D). Fluid-attenuated inversion recovery (FLAIR) can also be helpful by attenuating the signal of CSF.

We used an image-guided endoscopic endonasal approach to access this large invasive tumor. At the opening of the sphenoid sinus, we observed bulging of the posterior wall of the sinus and, while opening the mucosa, we encountered a pearly-white tumor. This appearance is characteristic of an epidermoid tumor. The cyst was removed by curette and suction until the dura appeared at the midline. We then followed the tumor to resect all parts of the mass. One-and-a-half centimeter away from the midline, the tumor intradurally extended

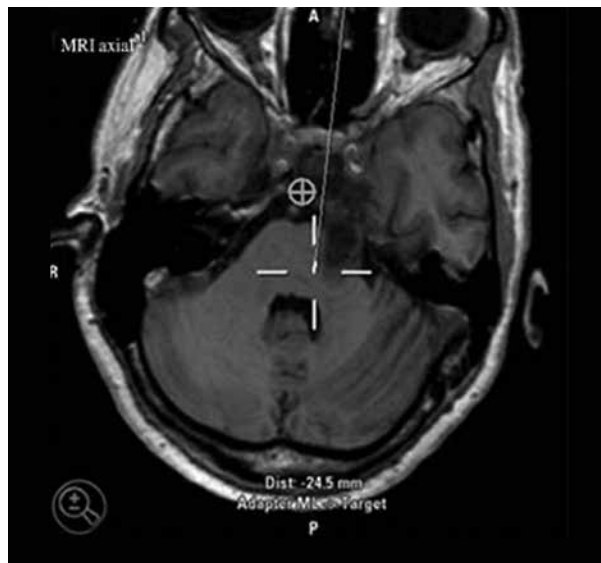
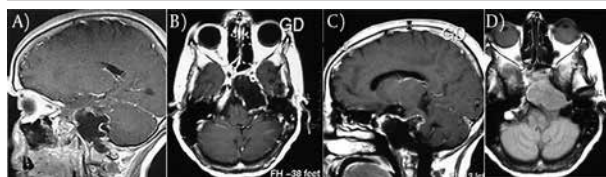


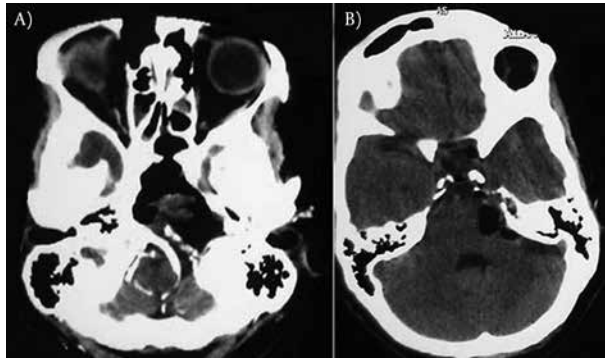
Figure 2. Intraoperative Images on Navigation System Screen

Figure 1. Preoperative Images



A) CT image without contrast; B) Axial MRI; C) Sagittal MRI; D) Proton density image

to the CPA cistern, where it invaded the dura; CSF leaked while the tumor was removed (Figure 2). Following the tumor laterally, the petrous apex compartment of the tumor was resected under image-guidance with the aid of angled lenses. Following the tumor inferiorly, we reached the craniovertebral region and cleared that area of the tumor. Finally, we repaired the dura using fat and fascia lata, and the CSF leak was successfully stopped. CT scan images acquired 3 days postoperatively are shown in Figure 3.

Figure 3. Postoperative CT Scans

A) Air in the CPA and inside the pons replacing the tumor; B) Extent of tumor resection in relation to the bony skull base is evident.

On the sixth postoperative day, the patient became febrile and developed signs of meningeal irritation. There was no evidence of CSF leak on endoscopic exam. A CSF smear and culture were also negative. Following 3 weeks of intravenous antibiotic therapy, the patient had no complications except for fever. She was discharged with the diagnosis of aseptic meningitis and was prescribed 15 mg per day of oral prednisolone, which was tapered over 6 weeks. Thereafter, she had no complications until postoperative month 3, when she died of pneumonia.

3. Discussion

This was a case of an unusual extradural epidermoid tumor within the petroclival region with significant superior, lateral, inferior, and posterior extensions and intradural invasion. A variety of conventional cranial-base approaches, including anterior, anterolateral, and posterolateral routes, are often used alone, in combination, or as a staged approach to excise such extensive tumors (2, 17). One of the lateral approaches is the transpetrosal approach. Mandell *et al.* (18) reported a partial labyrinthectomy using the petrous apicectomy approach (PLPA) that provides extended exposure of the petroclival region and markedly increases the incident angle toward the brainstem as compared with retrosigmoid routes. Presently, a transnasal corridor can provide access to the entire ventral skull base extending from the crista Galli to the spinomedullary junction. Fassett *et al.* (9) reported an expansile cystic lesion in the middle of the clivus that was explored through a transnasal transsphenoidal approach. They removed a thin shell of the bone overlying the cystic lesion to enter and remove the lesion through the clivus. Finally, the bone opening was widely fenestrated into the nasopharynx.

The wide extension of the tumor in the present case required a multidirectional route to gain access to the whole tumor. The endonasal approach has been constructed as a series of sagittal and coronal modular approaches that run between the 2 internal carotid arteries or lateral to the internal carotid artery (19). This

approach allows for the treatment of lesions in the sellar and suprasellar regions and provides extensive corridors to the lower skull base and even the upper cervical spine. Kassam and colleagues (19) defined the inferior limit of the endoscopic procedure as a line drawn in the sagittal plane from the caudal region of the nasal bone to the posterior hard palate (k-line). The present case exhibited involvement of the entire skull-base, which could be endoscopically approached through the transnasal route. Another factor enabling us to use an endoscopic endonasal approach was the consistency of the tumor, because most epidermoid tumors can be resected by suction and curette, and they are generally avascular. Aside from its cephalocaudal extension, it also had grown laterally into the temporal bone, causing acoustic problems. We were confident that this kind of extension could be safely addressed endonasally because of its slow growth and non-adherence to the carotid artery.

Previous studies have reported endonasal removal of intradural epidermoid tumors in the clival region (20). In the present case, the tumor was initially extradural, but it invaded the dura and assumed an intra-axial location due to its large size and chronic nature. After the core of the tumor was removed, we reached the remaining thinned clival bone, which had been penetrated by the tumor near the petrous apex. Then, we followed the tumor by a curette and fine-suction aspirator to remove the intra-axial portion as well (Figure 2). The dural repair was performed using fat, muscle, and fascia grafts that were secured with fibrin glue. Optimal management of an epidermoid tumor involves complete removal of the cyst and the surrounding capsule (21-23). Incomplete resection of the tumor can cause a chronic granulomatous reaction, and make complete removal of the residual tumor impossible (5, 15). This reaction could lead to the formation of dense adhesions between the capsule and intracranial vessels (4). In situations where local surgical excision of a lesion is not safe, surgeons should consider marsupialization. Marsupialization involves fenestration of the tumor capsule and opening a wide window on the cyst to let the tumor debris pass through and prevent the lesion from growing, bulging, and subsequently destructing the adjacent structures (9).

In cases where such tumors do not breach the arachnoid mater and there is no evidence of dural invasion, we advocate the use of marsupialization to the nasopharynx for better long-term control of the lesion. Otherwise, classic skull-base repair is mandatory in order to avoid postoperative CSF leak, which can be deadly. Another problem that may occur during the surgery on epidermoid tumors is spillage of the cyst contents into the subarachnoid space through small tears in the capsule, which can induce aseptic meningitis (24). However, a variety of maneuvers have been suggested to reduce the risk of postoperative aseptic and bacterial meningitis (23, 25). Furthermore, repeated washing of the cavity with saline can reduce the risk of aseptic meningitis

(26). The present patient developed aseptic meningitis, which was treated with oral corticosteroid therapy tapered over 6 weeks. The advantages of the endoscopic endonasal approach are that it offers a minimally invasive, anatomically direct trajectory and prevents brain retraction. In addition, using an angled endoscope gives the surgeon a panoramic view, allowing lateral, cephalad, and caudal visualization. Although endonasal transclival approaches for intradural epidermoid tumors of the CPA are controversial, we believe that the endonasal transsphenoidal approach using an angled endoscope is an ideal approach for excision of extradural petroclival tumors, even large, extended ones. However, the possibility of dural invasion of the tumor should be kept in mind in order to avoid CSF fistula formation, aseptic meningitis, prolonged hospitalization, and increased cost.

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Authors' Contribution

Maryam Jalesi, Guive Sharifi and Mohammad Farhadi, carried out the procedures and directly involved in the drafting, giving critical comments and revising of the manuscript. Ali Ahmadvand, Rozita Jafari and Sahar Zahedi collaborated in the gathering the information and were directly involved in the drafting and revising of the manuscript and postoperative care.

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