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Technical Notes & Surgical Techniques

Endoscopic endonasal transclival resection of a “pontine chordoma”: Technical case report

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ABSTRACT

Clival chordomas represent very challenging neoplasms, due to their tendency to infiltrate bone, frequent recurrence, involvement of important neurovascular structures, and deep location. Lesions involving the brainstem remain difficult to access through conventional skull-base approaches which are often associated with significant morbidities. We present a successful purely endoscopic endonasal transclival resection of a rare chordoma located in the ventromedial pons. Recent advances in endoscopic technology make endoscopic endonasal approach (EEA) a possible alternative to the treatment of ventral brainstem lesions.

1. Introduction

Chordomas are rare malignant bone tumors arising from transformed remnants of notochord. These tumors usually occur in the axis skeleton, with an almost equal distribution in the skull base (32%), mobile spine (32.8%) and sacrum (29.2%) [1,2]. Generally, clival chordomas constitute about 0.15% of all primary intracranial tumors [3]. Although characterized by slow growth and low grade histology, chordoma has a poor prognosis because of its locally aggressive behavior, high recurrence rate, and potential for metastasis. There are few reports of intracranial intradural chordomas [4,5], however, clival chordomas primarily occupy the extradural compartment [3,6]. In some cases, transdural extension of clival chordomas can result in the invasion to the brainstem and surround neurovascular structures, which creates high surgical risk [7].

The optimal approach for removal of brainstem lesions provides the shortest distance from the surface to the lesion with best visualization and minimum neural damage [8]. In recent years, endoscopic endonasal approach (EEA) is increasingly used for skull base surgery, and transnasal resection of ventrally situated brainstem lesions has been proposed [9,10]. We present an asymptomatic chordoma in the ventral pons managed through an endoscopic endonasal transclival approach. The specific tumor growth pattern and absence of clinical symptom led to the extremely rare manifestation of the chordoma. To the best of our knowledge, this is the first reported successful utilization of such an approach to resect a “pontine chordoma”.

2. Case report

2.1. Presentation and examination

An asymptomatic 60-year-old man was referred to our department for the pontine lesion found on magnetic resonance (MR) imaging. MR images revealed a spot like central enhancing 2.4 × 2.7-cm round-like lesion in the ventromedial pons with a large exophytic component (Fig. 1A–C). The lesion was partially encasing the basilar artery, and displacing it towards the left. Nasopharyngeal 3D-CT demonstrated a rugged bone defect at the dorsal clivus and the dorsum sellae (Fig. 1D–F).

Given the further lesion growth with resultant mass effect on the pons and the patient's strong desire for surgery, the decision was made to offer surgical resection. Depending on the tumor location in the ventral pons, we considered approaching the lesion using an endoscopic endonasal transclival approach.

2.2. Operation

After oral endotracheal anesthesia, the patient was placed in the supine position with upper part of the body slightly elevated. The patient's head was fixed in a Mayfield head clamp and rotated 15° towards the surgeon. Facial features were registered to the stereotactic neuronavigation system (StealthStation S7, Medtronic). We used a 2-surgeon, 4-handed bilateral nostril approach for the surgery. The endoscope (0°, 18 cm × 4 mm, Karl Storz) was inserted through the right nostril, and

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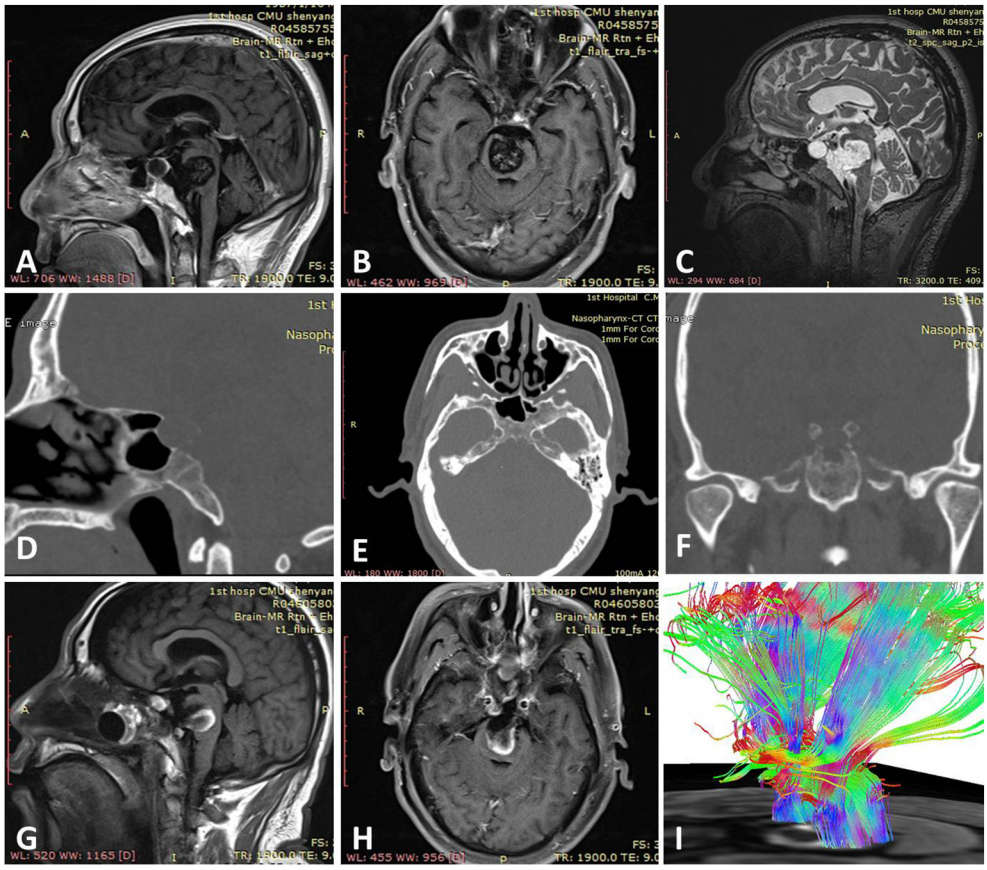

Fig. 1. A–C: Preoperative sagittal (A and C) and axial (B) MR images demonstrating a 2.4 × 2.7-cm ventromedial pontine lesion. D–F: Preoperative sagittal (A), axial (B), and coronal (C) CT images demonstrating rugged bony defect of the dorsal clivus and the dorsum sellae. G–H: Postoperative sagittal (A) and axial (B) MR images demonstrating a near-total tumor resection via endoscopic endonasal transclival approach. I: Postoperative DTI demonstrating preservation of the CSTs that were displaced posteriorly by the lesion.

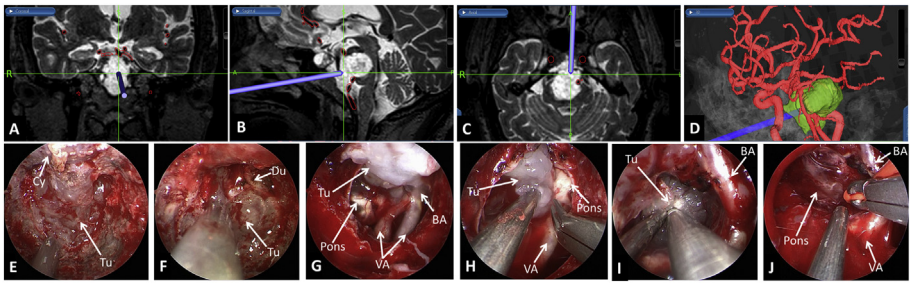

Fig. 2. A–D: Use of stereotactic navigation guidance system with MR images. Excellent accuracy achieved to localize the precise site of the intrapontine lesion. E–J: Endoscopic endonasal view of surgical field. Cy, cyst; Tu, tumor; Du, dura; BA, basilar artery; VA, vertebral artery.

the inferior 1/3 partial middle turbinectomy was performed to provide adequate space. After identification of the opening to the sphenoid sinus, a right nasoseptal flap was dissected and stored in the choana inferiorly. The posterior nasal septum was removed and the left middle turbinate was displaced laterally. The anterior wall and the floor of the sphenoid sinus were removed with a Kerrison rongeur and a high-speed electric endonasal drill (cutting and diamond, 7000 rpm) to expose the clival region. With the bilateral carotid arteries identified by stereotactic navigation, the clivus was removed along the midline. The off-white gelatinous and myxoid lesion was visualized as the clivus thinned out, and samples were sent for pathological examination (Fig. 2E). Intraoperative frozen section suggested chordoma. According to the extent of the lesion, the clivus and the dorsum sellae got further drilled. This was limited laterally by paraclival carotid arteries. A combination of plasma surgery system with a radiofrequency needle electrode and Surgicel absorbable hemostat (Ethicon, INC.) were required to achieve hemostasis of the right cavernous sinus. With the extradural lesions removed through normal suction, a round dura defect was found (Fig. 2F, G), through which tumor encasement of the basilar artery and vertebrobasilar junction was directly visualized. Gentle suction and sharp dissection were used to separate the tumor tissue from the arteries and ventral surface of the pons (Fig. 2H). Care was taken to remove the intrapontine lesion using dissectors, ring curettes, and suction cutter in a piecemeal fashion (Fig. 2I). Once the tumor was resected, hemostasis was achieved using Surgiflo haemostatic matrix (Ethicon, INC.) (Fig. 2J).

Fat tissue and autologous fascia lata harvested from patient's right anterior lateral thigh were inserted extradurally to reconstruct the defect. Then the nasoseptal flap was elevated and overlaid to complete the reconstruction. The flap was buttressed with fat tissue and absorbable hemostat and held in place with fibrin sealant kit (Guangzhou Bioseal Biotech CO.). 15 cm^3 water was used to fill the balloon from a Foley catheter to hold the flap against the skull base. Nasal packing was placed bilaterally to ensure hemostasis.

2.3. Pathological findings

The tumor pathology was consistent with chordoma.

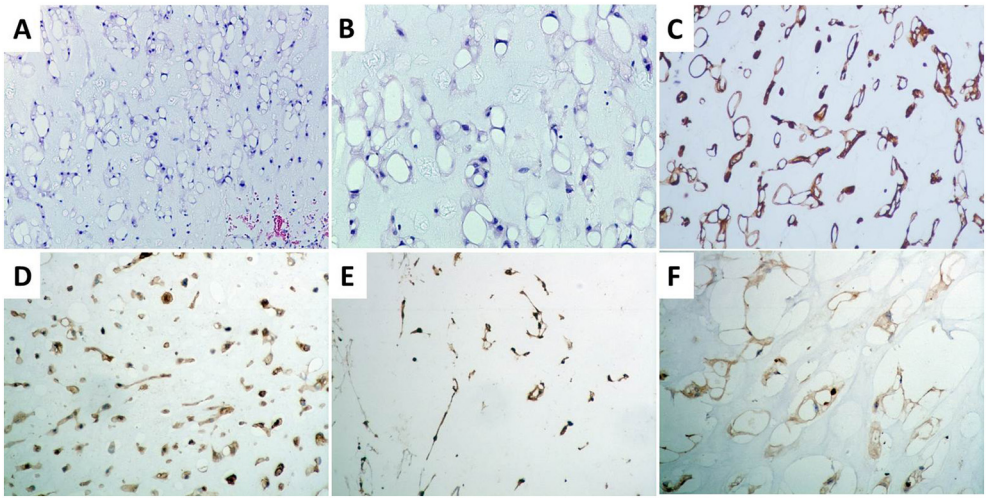

Fig. 3. A–B: Hematoxylin and eosin stain (A, magnification ×100; B, magnification ×200) demonstrating typical findings of a chordoma with markedly vacuolated cells with abundant cytoplasm and small round or oval nuclei embedded within a rich myxoid matrix. C–E: Immunohistochemistry revealed a positive reaction to Cytokeratin (C, magnification ×200), EMA (D, magnification ×200), S-100 protein (E, magnification ×200). F: Ki-67 labeling index (F, magnification ×200) was estimated < 1%.

Microscopically, tumor tissue consisted of cords and nests of markedly vacuolated cells with abundant cytoplasm and small round or oval nuclei embedded within a rich myxoid matrix. Immunohistochemistry demonstrated a positive reaction to Cytokeratin, Epithelial Membrane Antigen (EMA), and S-100 protein. The Ki-67 labeling index was estimated < 1% (Fig. 3A–F).

2.4. Postoperative course

The surgical time was 290 min and the blood loss was 250 ml. The patient was extubated 4 h after the operation and admitted in the neurosurgical intensive care unit for neurologic monitoring for 24 h. No major complication was showed on the early postoperative CT scan. Immediately after the procedure, motor examination revealed 3/5 strength in the left upper and lower extremities. The Foley balloon was removed on 8th postoperative day and no cerebrospinal fluid (CSF) leak was appreciated. MR imaging and diffusion tensor imaging (DTI) performed 10 days after the surgery demonstrated a near-total tumor resection and preservation of the corticospinal tracts (CSTs) (Fig. 1G–I). On postoperative day 14, he was discharged home and his left arm and leg strength had improved to 4+/5 strength. Repeat MR imaging performed 3 months after the surgery revealed expected postoperative changes. At the 6-month follow-up, the patient presented in good condition and was able to finish the activities of daily living independently.

3. Discussion

Clival chordomas are predominantly midline lesions exhibiting various growth patterns in all directions. Aggressive extradural and intradural involvement has been well described. Brainstem invasion is an extremely rare manifestation of clival chordoma with few cases reported in the literature [11,12]. With limited bone infiltration, the case presented transdural extension and grew into the brainstem. Ideal treatment paradigm for clival chordoma consists of maximally safe aggressive resection and adjuvant radiotherapy [1]. Previous study described that cranial chordomas invade the bone but they simply displace surrounding soft tissues [6]. The histological research by Oikawa et al. emphasized the fact that chordomas invade the submucous layer and not the dura and vital neurovascular structures even in the advanced stage [13], making it possible to remove the lesion with minimal risk of a persistent neurologic deficits. However, resection for such tumor is associated with high rates of mortality, including cranial nerve palsy, brain stem infarction, and basilar artery injury. For this reason individualized surgical approach considering the possible complications should be applied for such ventral brainstem lesion.

Various microsurgical approaches and safe entry zones to the brainstem have been well described in the literature, including anterolateral, lateral, and dorsal approaches [14–17]. Traditional open approaches are suboptimal for providing direct access to ventral brainstem surface [8,18,19]. In cases of ventromedial brainstem lesions, external approaches carry the risk of non-negligible morbidities because of the necessity of passing through a greater width of normal brainstem tissue to access the lesion. Large modern series show up to a 30% risk of motor deficits after resection of pontomedullary junction cavernomas when approached laterally [15]. A recent study by Essayed et al. described clinical feasibility, potential indications and limitations of EEA for ventral brainstem lesions [20]. Their report provided the first cadaveric anatomical study towards the application of this route for intraaxial brainstem surgery, and concluded that EEA is best suited for midline exophytic pontine lesions or the nonexophytic lesions strictly anterior to CSTs. Previously there are 7 cases of ventral brainstem cavernous malformations, a recurrent pediatric pontine ependymoma, and a pontine glioma being treated via EEA [9,10,21–27]. The promising surgical results of the cavernous malformations showed that gross-total resection rate reached 85% (6/7) and clinical condition was improved or the same in 100%. As a result of inevitable relapse, the prognosis of the 2 other cases of high-grade glioma and WHO Grade III anaplastic ependymoma remained poor. These 2 reports demonstrated the EEA a viable and advantageous option for the treatment of ventral intrinsic brainstem lesions in select patients.

The shortest route to the lesion described here was undoubtedly through a direct ventral approach. Additionally, the exophytic lesion presented a natural corridor to gain entry into the brainstem and remove the lesion from within. We believe the endoscopic endonasal transclival approach provides adequate visualization of the entire lesion, minimizes retraction of neurovascular structures, and offers a good working angle to facilitate hemostasis and complete resection. At the present time, advances in endoscopic techniques and equipment enable surgeons to reach and remove many skull base lesions from the anterior cranial fossa floor to the C2 vertebrae. Some more lateral areas including jugular foramen, cerebellopontine angle, and middle cranial fossa can be accessed with the aid of angled endoscopies and instruments. The approach proposed here should only be performed by neurosurgeons with considerable skill and expertise in endoscopic endonasal surgery. The surgeon has to minimize surgical trauma to brainstem tracts or nuclei using the microsurgical dissection technique, and deal with intraoperative bleeding from the basilar plexus or the premontane dura mater with the aid of bipolar forceps or hemostatic agents. In our case, bleeding from the basilar plexus is not so severe because it is partially blocked by intradural extension of the chordoma.

In addition, the biggest concern using an endoscopic skull base

approach for intradural lesions is the risk of CSF leak. However, just 2 cases that were reported previously developed the CSF leaks and were successfully treated with revision of the nasoseptal flap in a second surgery [10,27]. Over the years the risk of postoperative CSF leak has decreased significantly with refinements in surgical techniques such as gasket seal closure and vascularized pedicle nasoseptal flap multi-layered reconstruction technique. In this regard, the use of routine postoperative lumbar drainage after endoscopic skull base repair is controversial. In our experience, we mainly perform postoperative lumbar drainage to selected cases for the treatment of previous reconstruction failure, and in this case no CSF leak was documented.

4. Conclusions

We have described a rare case of asymptomatic “pontine chordoma” resected using endoscopic techniques. Compared with lateral surgical approaches, the EEA provides most direct access to the ventral brainstem surface. This corridor should be considered as a possible alternative for ventrally located brainstem lesions, and further clinical studies in very selective cases will be necessary to validate this approach. This kind of surgery requires extensive experience with endoscopic skull base skills and the specific instruments used in endoscopy.

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Conflict of interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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