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ABSTRACT

Chordomas are rare locally aggressive midline tumors usually seen involving clivus and craniovertebral junction. Safe maximal resection is the mainstay of treatment. We report a case of a young girl with chordoma where a transoral approach was used with limited resection of hard palate and vomer for optimal exposure and maximal safe resection. Good exposure for radical decompression and reduced postoperative morbidity can be achieved using individualized modifications in transoral approach for chordomas involving craniovertebral junction.

Keywords: Chordoma, Craniovertebral junction, Transoral approach.


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Conflict of interest: None

INTRODUCTION

Chordomas are rare, slow-growing malignant bony tumors that arise from remnants of the notochord and account for approximately 1% of intracranial tumors.1 Approximately 35 to 40% of these tumors occur in the skull base, where they typically involve the clivus.2 Even though they show benign histological behavior, the prognosis is poor due to their deep location, the presence of nearby important structures, and their propensity to invade locally. Also, the very high recurrence rates make it a challenge for the surgeons to achieve total resection.3,4 They are conventionally managed by transoral approaches with palatal splitting.5 If the tumor is extending superiorly, transfacial approach with or without maxillotomy is often needed, resulting in increased morbidity. Most of the time, a combination of operations with various surgical approaches is required. We describe a novel modification for avoiding palatal splitting and thus reducing the morbidities associated with it.

CASE REPORT

A 11-year-old girl presented with neck pain, radiating right upper limb pain, progressive weakness of both upper limbs, nasal twang in speech along with difficulty in mixing bolus of food in the mouth. Neurological examination revealed bilateral hypoglossal involvement, wasting of intrinsic muscles of hand, bilateral upper limb weakness, brisk reflexes, and right C5-8 hypoesthesia. Magnetic resonance imaging (MRI) revealed T1 hypointense, T2 hyperintense, heterogeneously enhancing solid clival lesion measuring 4.0 × 4.5 × 5.0 cm (anteroposterior × transverse × craniocaudal) compressing on the medulla and upper cervical spinal cord (Fig. 1). These features were suggestive of chordoma.1

We decided to perform a combination of surgical approaches in a staged manner. In view of her rapid worsening due to cord compression, she initially underwent a posterior approach C1, C2 laminectomy, and decompression of lesion. She showed immediate postoperative clinical improvement in her right upper limb power. Histopathological examination confirmed the diagnosis of chordoma.

A second surgery, the definitive anterior approach, had to be planned. Traditional transoral approach would not have provided the adequate exposure required. An added maxillotomy would do so but with a high risk of significant postoperative morbidity, such as swallowing difficulty and change in voice. Hence, a modified transoral approach was planned, with tailored resection of posterior hard palate and vomer to provide adequate exposure, which would also reduce the risk of postoperative morbidity (Fig. 2).
Surgical Technique: Tailored Resection of Hard Palate and Vomer

Under general anesthesia, with the patient in supine position, mouth was kept open with retractors. Palatal mucosa was incised and pushed laterally. Posterior hard palate and vomer were excised piecemeal using rongeurs and drill to get a panoramic view of superior aspect of the posterior pharyngeal wall (Fig. 2). Posterior pharyngeal wall was incised. On either side of the apical ligament, a grayish white mass was seen. The tumor was removed piecemeal. It was grayish white, firm in consistency, and moderately vascular. The tumor was seen extending posterior to odontoid process. The tip of the odontoid process was drilled out to reach the posterior component. Piecemeal resection of the tumor was done rostrally up to clivus, posteriorly till the dural tube was seen and inferiorly till the body of C2 vertebra (Fig. 3). Gross total resection of the lesion was achieved. Dura mater was intact.

Postoperative imaging revealed no residual lesion (Fig. 4). There was resolution of lower cranial nerve symptoms and improvement in the right upper limb power. She did not receive any radiotherapy.

Follow-Up

The patient had progression in the size of the tumor after 16 months and underwent posterior approach and decompression. Thirty months after the transoral procedure, she underwent another reexploration—posterior decompression—for progressive myelopathy and recurrence. She is on regular follow-up for over 5 years now, asymptomatic, and is going to school.

DISCUSSION

Chordomas are relatively rare, slow growing, locally aggressive tumors, originating from embryonic remnants of the notochord.1 These lesions are deep and in midline and most commonly seen in the sacrum and skull base/
Modified Transoral Transpalatal Approach with Tailored Resection of Hard Palate and Vomer for Clival Chordoma

Figs 2A to D: Transoral modification: schematic and intraoperative images. (A, B) Mucosal incision and flap elevation. (C) Planned area of hard palate removal. (D) Highlighted area showing the tailored posterior vomer and hard palate removed.

Figs 3A to C: Intraoperative images. (A) Incision of posterior pharyngeal wall. (B) Piecemeal excision of the grayish white, firm tumor. (C) Dura and the dens after tumor excision.
clivus region. They often reach very large sizes. They often require multiple surgical procedures; even then, multiple local recurrences are common.3,4,6 Current evidence indicates that initial radical surgical removal supplemented by proton beam radiotherapy is the best mode of management of these lesions.7

The chordoma in our patient involved the clivus, extended anteriorly into perivertebral space and inferiorly up to the C2 vertebral body. Initially, posterior approach for tumor resection was preferred, as it would relieve rapidly worsening medullary and cord compression symptoms. There was still a significant residual mass remaining. To tackle it, a modified transoral approach was used with limited resection of hard palate and vomer. This provided adequate surgical field to reach the entirety of the tumor and resect it. Thus, we were able to restrict the postoperative morbidity which might have been incurred in cases of more radical approaches.

We would like to emphasize that surgical procedures for chordomas should be planned on a case-to-case basis and minor modifications of procedures will not only
provide adequate surgical field for tumor excision but also spare the patient of debilitating postoperative morbidities.

REFERENCES