

# Surgical Excision of Clivus Chordoma with the Use of Coblator—A Case Report\*

Saloni Shah<sup>1</sup>, Roma Gandhi<sup>1</sup>, Hemang Brahmabhatt<sup>1</sup>, Rajesh Viswakarma<sup>2</sup>

<sup>1</sup>B.J. Medical College, Civil Hospital, Ahmedabad, India

<sup>2</sup>ENT Department, Civil Hospital, Ahmedabad, India

Email: [drsaloni155@gmail.com](mailto:drsaloni155@gmail.com)

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## Abstract

Chordomas are dysembryogenic tumors originating from the notochordal process [1] [2]. They are aggressive tumours with unique diagnostic and management challenges. Primary therapy is complete surgical removal of the tumour as much as possible. The likelihood of recurrence is high in spite of complete surgical resection. A 52-year-old female patient presented with complaints of decreased vision in right eye, nasal bleeding, nasal blockage and difficulties in swallowing. CT scan and nasal biopsy were performed which confirmed the diagnosis of clivus chordoma. The CT scan showed extension into nasopharynx, nasal cavity and oropharynx pushing onto the soft palate. Surgical excision of the mass was performed with coblator by both intraoral and intra nasal approach [3]. On follow-up, nasal endoscopy and CT were done; the patient was relieved of the symptoms and was clinically better.

## Keywords

Chordoma, Clivus, Nasopharynx, Oropharynx, Endoscopic Excision, Coblator

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## 1. Introduction

Chordomas are tumors of notochordal origin that may affect the axial skeleton anywhere from the coccyx to the base of the skull, in either a midline or paramedian position. The cranial and caudal extremes of the spine are most often affected. The notochordal cells are preferentially left behind in the clivus and sacrococcygeal regions when the remainder of the notochord regresses during fetal life. Chordomas are rare, aggressive, slow-growing, invasive, and locally destructive tumors [1] [2].

35% to 40% of these tumors involve the clivus. These are rare tumors with an estimated incidence of 0.51 cases per million and approximately 1% of intracranial tumors [1] [2].

The clivus is the surface of a portion of occipital and sphenoid bones in the base of the skull. It is surrounded by the neurovascular structures of the brainstem, which includes both internal carotid arteries. Tumors of the

\*Guided by Dr. Rajesh Viswakarma (Head of the ENT Department, Civil Hospital, Ahmedabad, India).

clivus can be benign or cancerous; they can be classified as chordomas or chondrosarcoma. Clival chordoma becomes symptomatic by locally invading surrounding cranial nerves and brain stem structures.

Although there is no racial predilection for chordomas, the incidence in males is 2-fold greater than in females (2:1), and the tumors are found primarily in adults, occurring rarely in patients younger than 30 years. The most common sites are the skull base, the sacrum, and the mobile spine [1] [2].

These tumors are difficult to manage because of their critical location and propensity to recur; they have been treated in the past years with combination of surgery and radiotherapy. No single surgical method has emerged as a standard of therapy for resection of these tumors, although numerous surgical approaches to the clivus are described. Surgical goal involves complete removal of tumor as much as possible. The role of adjuvant radiotherapy thereafter is still subject to debate. With the continued development of advanced microsurgical techniques in skull base surgery, more extensive dissections as well as combined approaches to the skull base have been advocated.

## 2. Pathology

Chordomas have 3 histological variants: classic, chondroid, and dedifferentiated. Classic chordomas appear as soft, gray-white, lobulated tumors composed of groups of cells separated by fibrous septa. They have round nuclei and an abundant vacuolated cytoplasm described as physaliferous. They are pathologically identified by their physaliferous features and immunoreactivity for S-100 and epithelial markers such as MUC1 and cytokeratins. Some studies have postulated that the notochord developmental transcription factor, brachyury, could be a novel discriminating biomarker for chordomas [1] [2].

**Clivalchordoma patient presentation:** most common headache, diplopia secondary to VI cranial nerve paresis and visual changes including blurring or sometimes loss of vision. The patient may present with multiple lower cranial nerve palsy symptoms such as facial numbness and asymmetry, dysphagia, hoarseness and speech problems. Finally, large tumors may cause brainstem compression and patients may present with long tract signs and ataxia [4]-[7].

We present a case report of a patient who presented initially with decreased vision in right eye, nasal blockage, nasal bleeding, decreased hearing and difficulty in swallowing and later was diagnosed to have a clival chordoma. We discuss the treatment modalities and present a systematic review of the literature.

## 3. Case Presentation

A 52-year-old Caucasian woman, living alone and previously in good health, was admitted on 25/6/13 at CHA with complaints of decreased vision in right eye and nasal blockage, one month before she was operated for nasal polyp in Bhuj general hospital. After the operation the patient again developed the complaints of nasal blockage and nasal bleeding associated with decreased vision and difficulty in swallowing.

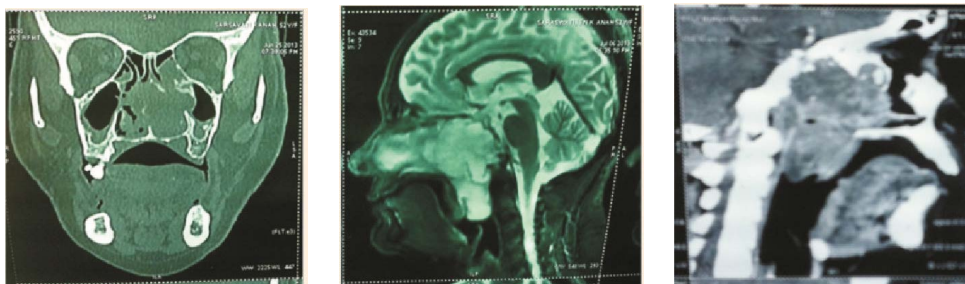
On examination nasal mass was present in right nasal cavity with right side soft palate bulge. Cranial nerve examination was normal.

A computed tomography (CT) scan of paranasal sinuses showed large (55 × 43 × 76 mm) ill defined inhomogeneous enhancing soft tissue density mass filling posterior nasal cavity causing erosion of clivus, sphenoid sinus, ethmoid sinus and medial wall of left orbit, inferiorly extending in to oropharyngeal space.

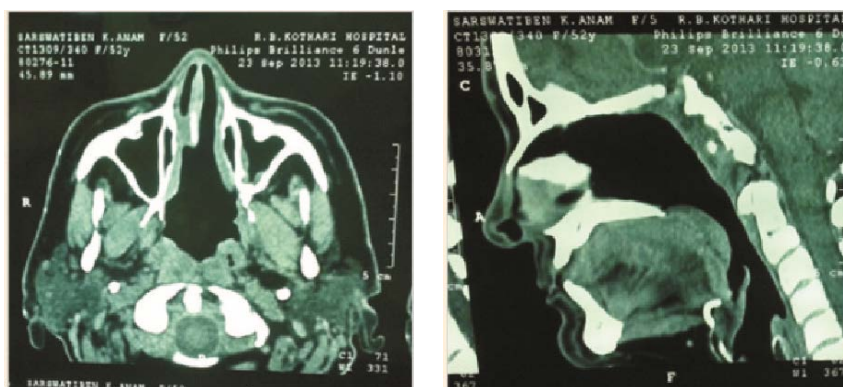
Nasal endoscopy and biopsy was taken on 1/7/13 and sent for HPE which confirmed diagnosis of a chordoma in the clival region.

An endonasal endoscopic excision of the nasal mass was done with the help of coblator on 11/7/13.

### PRE OPERATIVE IMAGES



## POST OPERATIVE IMAGES



Post op the patient was followed. Patient was clinically better with complete relief of symptoms. Post op CT PNS was done on 23/9/13 which showed about 16.6 × 14.4 mm size abnormal enhancing lesion noted at left infratemporal fossa left lateral wall of nasopharynx? Residual mass lesion.

No adjunctive radiotherapy or any other therapy was used for this patient.

#### 4. Discussion

Endoscopic approach to clival chordomas comes with less morbidity to the patient as compared to conventional trans cranial approaches.

Clival chordomas can be managed by a variety of conventional surgical approaches: transcranial, transsphenoidal, transoropharyngeal and maxillary osteotomy approaches. Transcranial approaches involve brain retraction and have increased risks of cerebral edema and hematoma, apart from carotid, basilar artery and optic nerve trauma. These complications can be greatly reduced with anterior (transnasal, transoral and transfacial) approaches. Currently, endoscopic surgery has opened a new avenue in the management of clival chordomas, not only as a direct surgical access but also by providing an excellent visualization of the clivus and surrounding structures, especially the anterior dura and the basilar artery [3] [8].

Radiation therapy can reduce the risk of recurrence after surgery and prolong survival. For patients who are not candidates for surgery, radiation therapy is sometimes used as the primary treatment. Chordomas are resistant to radiation, meaning that very high doses of radiation are required to control these tumors. Because of their proximity to vital anatomy such as the brain and spinal cord, which cannot tolerate high doses of radiation, specialized forms of radiation are used to focus radiation on the tumor while avoiding surrounding tissue [8] [9].

#### 5. Conclusion

Endonasal endoscopic surgery provided safe and reliable tumor resection for a lower clival lesion. We believe that this minimally invasive procedure should be considered as an alternative to traditional surgical treatment.

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