

Title:

Sequential approaches for resection of clival chordoma

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Abstract:

Introduction

Chordomas are extra-axial tumours that originate from the primitive notochord. They are rare diseases that present mostly between the third and the fifth decades of life with an incidence rate of 0.08 per 100,000 in the United States. Approximately 35% of chordomas arise in the clivus of the skull base (clival chordoma). In spite of their slow-growing and non-malignant histopathological appearance, they are potentially life threatening due to their locally destructive and invasive nature. Clival chordomas are located near the sphenoid-occipital junction at the base of the skull, behind the sphenoid sinuses. Involvement of the surrounding cranial nerves accounts for their symptomatology. It is notorious for its high tendency of recurrence following treatment, with a reported recurrence rate of 68%. Therefore, gross total resection in combination with adjuvant radiotherapy is the standard mode of treatment.

Methodology

The authors adopt a sequential approach aiming for gross total removal of the clival chordoma. The first stage of surgery involves a lateral approach, going through a subtemporal window. The lateral wall of the cavernous sinus is split and the temporal lobe is retracted. A combination of transpetrosal, transcavernous plus transtentorial dissection are made to reach the upper, lateral and posterior portion of the tumour. The tumour is then dissected and removed medial to the cavernous segment of internal carotid artery (ICA). Medially, the contralateral cavernous sinus is reached. Posteriorly, the basilar artery and the lower clivus are exposed and clear of tumour. Small remnant of tumour is deliberately left in the anterior portion of the tumour to avoid inadvertent entry to the sphenoid sinus to prevent cerebrospinal fluid (CSF) leak. After subtotal removal, a pedicle fascial graft, harvested from the temporalis fascia, is rotated and placed at the back wall of the resection cavity.

After 6-8 weeks, second stage of surgery will be performed from an anterior approach, mainly using endoscopic transsphenoidal approach. The sphenoid sinus is exposed. Tumour arising from the sellar and diseased clivus is removed until the thick fascial graft is reached. The fascial graft serves as a protector for CSF leak and assume a complete clival removal. Laterally, part of the clival bone infiltrated by the tumour is removed by high-speed diamond drilling until normal cancellous bone comes into view. That is classified as gross total removal. If feasible, adjuvant radiation therapy is given after the surgery.

Results

From Jan 2007 to July 2014, a total of 10 patients, 6 male and 4 female, with clival chordoma (n=8) or chondrosarcoma (n=2) were treated in our hospital. Tumour sizes ranged from 3 to 6cm in maximum diameter, occupied the central clival region. Nine out of ten patients the tumour also extended to the cavernous sinus. Six patients presented with cranial nerve palsy, mainly in the form of impaired lateral gaze due to abducent nerve palsy. The rest of the patients presented with headache, gait instability, or swallowing difficulties due to brainstem compression. Five patients were managed by other hospitals and were referred to our institute due to tumour progression. Four out of five patients had received surgery with partial removal. Four received adjuvant irradiated by either conventional RT or Gamma knife.



All patients were operated by a sequential approaches, firstly a lateral subtemporal approach and followed by a second stage transsphenoidal approach in 6-8 weeks. Eight out of ten patients (80%) achieved gross total removal. If possible, all patients were treated with adjuvant wide-field radiation. Four patients were complicated by CSF leak and required re-exploration for dural repair. After the introduction of pedicle fascial graft repair in stage one resection two years ago, no more CSF leak was observed. Follow up duration ranged from 6 months to 7 years and mean follow up duration was 32 months. There was no operative mortality in this cohort. Eight out of 10 patients achieved gross total removal and follow up MRI brain showed no tumour recurrence. One patient died 14 months after surgery due to tumour progression. One patient remained in minimal conscious state four months after the surgery. Concerning cranial nerves deficit, five patients suffered transient oculomotor nerve palsy and all showed significant improvement after a 6-month period. Permanent and transient 6th nerves palsy occurred in 3 patients and 4 patients respectively. Nine out of 10 patients the trochlear nerves were cut during the transcavernous dissection. All patients adapt well to the trochlear nerve palsy. Karnofsky score of these 8 patients ranged from 80-100%.

Conclusion

The sequential surgical approach to clival chordoma, stage one lateral approach follow by stage two anterior approach is an effective way for maximal resection with acceptable morbidity. 80% of the patients achieved gross total removal and showed no tumour progression during follow up.