Case Report

An intradural skull base chordoma presenting with acute intratumoral hemorrhage

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We present a rare case of skull base chordoma of extraosseous intradural type that presented as acute intratumoral hemorrhage. Surgical removal of the tumor was accomplished using a skull base approach.

Key words: Echordosis physaliphora, intradural chordoma, intratumoral hemorrhage

Introduction

Skull base chordomas are in general extradural tumors and present with slow progressive symptoms over several months. We report a case of skull base chordoma that was intradural in location and presented with acute intratumoral hemorrhage.

Case Report

A 35-year-old woman suffered a sudden episode of headache and right hemibody hyperalgesia. Investigations showed a hemorrhagic tumor extending intradurally on CT and a contiguous relationship between the tumor and the dural wall of the clivus on enhanced T1-weighted sagittal images [Figure 1]. No destructive alteration of the clivus was detected on serial high-resolution CT.

A transcervical approach revealed a gelatinous tumor covered by thickened arachnoid membrane. Internal debulking of the tumor was first accomplished via the space between the 9th and 10th nerve complex and the 7th and 8th nerve complex [Figure 2]. The tumor appeared to grow invasively into the brainstem. Tumor removal revealed dural attachment of the tumor at the clivus. The tumor was totally intradural and the bony structure of the petroclival area was intact. Subtotal removal of the tumor was accomplished and residual tumor tissue remained in relationship to the brainstem and the dural layer of the clivus. Pathological examination of tumor tissue showed a lobular pattern of physalipherous cells with abundantly vacuolated cytoplasm [Figure 3]. Histological examination of the tumor revealed typical features of a chordoma.

Postoperatively the patient was noted to have worsened in consciousness. Postoperative CT scanning revealed the localized hemorrhage in the tumor cavity. Emergency surgery for removal of the hematoma was performed. The second surgery suggested that postoperative hemorrhage was localized in the tumor cavity and was caused by the bleeding from the residual tumor. Three months after surgery, the neurological status of the patient had further improved and she was able to carry on normal activities with some effort. Postoperative MR images, obtained at 3 months after surgery, indicated a small residual tumor [Figure 1B]. At a six-month follow-up scan, growth of the residual tumor was identified. Adjuvant treatment of stereotactic radiosurgery was administered.

Discussion

Patients with skull base chordomas generally present with progressive clinical symptoms. We identified 11 documented cases, from the literature, of skull base chordomas presenting with acute intratumoral or intracranial hemorrhage [Table 1]. Numerous mechanisms have been proposed to explain the etiology of
intratumoral or intracranial hemorrhage in conjunction with brain tumors.\(^1,2\) The growth pattern of skull base chordomas suggests that extraosseous intradural chordomas are different from the classic type. Extraosseous intradural chordomas are hypothesized to originate from the ecchordosis physaliphora.\(^3\) Preoperatively, we did not consider that this hemorrhagic tumor was extraosseous intradural chordoma; therefore, we selected the transcondylar approach. However, for radical resection of a chordoma of this region, extended middle fossa approach described by Goel may have been ideal.\(^4\) Adjunct therapy after subtotal resection of the skull base chordoma may be indispensable. Conventional radiation therapy, stereotactic radiosurgery and proton beam therapy are alternatives.

## References


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