



Clinical Study

Bone invasiveness is associated with prognosis in clivus chordomas



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ABSTRACT

Clivus chordomas present a great challenge for neurosurgeons, and the prognosis is poor. To investigate bone invasiveness characteristics in regard to the prognosis of clivus chordomas, a retrospective study of 19 patients with primary clivus chordoma was performed. Clinical data, MRI, CT scans and follow-up data were examined, and the bone invasiveness of the chordoma was classified into two growth patterns (endophytic and exophytic) which were analyzed with regard to prognosis. The overall survival rate was 78.9% with radical surgery and adjuvant radiation, with a mean follow-up of 44.5 months. There were 12 patients in the endophytic group and seven in the exophytic group, and the exophytic group exhibited a higher recurrence rate than the endophytic group ($p = 0.006$). Chordomas with an exophytic growth pattern were more likely to recur than those with an endophytic growth pattern, and the surgical approach can be tailored according to each growth pattern.

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1. Introduction

Chordoma is a rare tumor derived from the remnants of the notochord with an estimated incidence of 0.08 per million in Western nations [1] and 0.04 per million in Taiwan Han Chinese populations [2]. They usually occur in the axis, and 49.4% occur in the clivus regions [3]. The treatment for clivus chordomas is challenging, and total removal is ideal [4–8]. However, the slow-growing, yet infiltrative, characteristics of this relatively low malignancy tumor makes complete removal difficult, particularly because of the surrounding vital neurovascular structures, which contributes to recurrence and poor prognosis [8,9]. One systematic review found 5 year and 10 year survival rates were 70% and 63%, respectively [9].

Modern technologies and improvements in neurosurgery have resulted in several well-documented approaches for treatment, the goals of which are to remove as much tumor as possible, improve quality of life and prolong survival [6,8]. Anterior approaches, lateral approaches, and multi-staged combined approaches are used extensively according to the site of the tumor and its relationship to the clivus [4–8,10–13]. Our previous report [8] demonstrated that radical surgery contributed to a longer survival. However, a case series by Sen et al. [7] reported a survival rate of only 58% with radical resection, and radical resection was

dependent on preoperative tumor volume and the number of anatomical areas involved with tumor.

Our previous report [8] classified these tumors into four states according to their dura mater infiltration. However, dura mater infiltration was not an independent risk factor for prognosis. This classification was insufficient because it did not consider the relationship of the tumor to bone (clivus). Bone invasiveness makes complete surgical removal difficult to achieve, which contributes to tumor recurrence. Rarely, case reports demonstrate that chordoma may arise in the intradural space [14,15]. MRI and CT scans reveal the various relationships between these tumors and nearby bones. Our recent proteomics study showed that the growth pattern of clivus chordomas (endophytic and exophytic types) may be differentiated by transforming growth factor β (TGF β) and phosphatase and tensin homolog deleted on chromosome 10 (PTEN) [16]. The clinical importance of these two growth patterns is not well documented. Therefore, we retrospectively reviewed our own series of primary clivus chordomas to investigate this characteristic.

2. Materials and methods

2.1. Patient population

The senior author treated 19 patients with primary clivus chordoma in the Department of Neurosurgery, Tian Tan Hospital, Capital Medical University, China from February 2004 to June 2005. All cases were retrospectively studied using detailed data.

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Cases without pathological confirmation were excluded. Patient follow-ups were performed via postsurgical office visits and telephone interviews with patients who were unable to visit in person. All patient data were compiled from hospital and office records, imaging studies, and the patients' own records. All patients voluntarily participated in the study under the protection of the Hospital Ethics Committee.

2.2. Classification of growth pattern

The growth pattern of the clivus chordoma was identified according to the bone invasiveness of the tumor on preoperative images, including plain and enhanced head MRI, thin layer skull base CT scanning and three-dimensional reconstruction, as described in a previous report [16]. The chordomas were

classified into two subtypes: endophytic and exophytic, as shown in Figure 1 and 2. The following methods were used: the maximum diameter at eyeball level on T2-weighted axial MRI was defined as the baseline level, and the area of the bilateral carotid cavernous lateral walls connected to the bilateral petrous apex at the baseline was used as the standard region. A tumor was identified as endophytic if at least 50% of the tumor, which can invade the bone in all directions, was located within the standard region at the baseline level or if the clivus bone was transformed into a “bubble” or “dumbbell” (Fig. 1A, 2A,C). Tumors were identified as exophytic if at least 50% of the tumor was located outside the standard region with limited bone invasiveness at baseline levels; these tumors may show a “pedicle-like” attachment to the clivus into intracranial areas on MRI and CT scans (Fig. 1B, 2B,D).

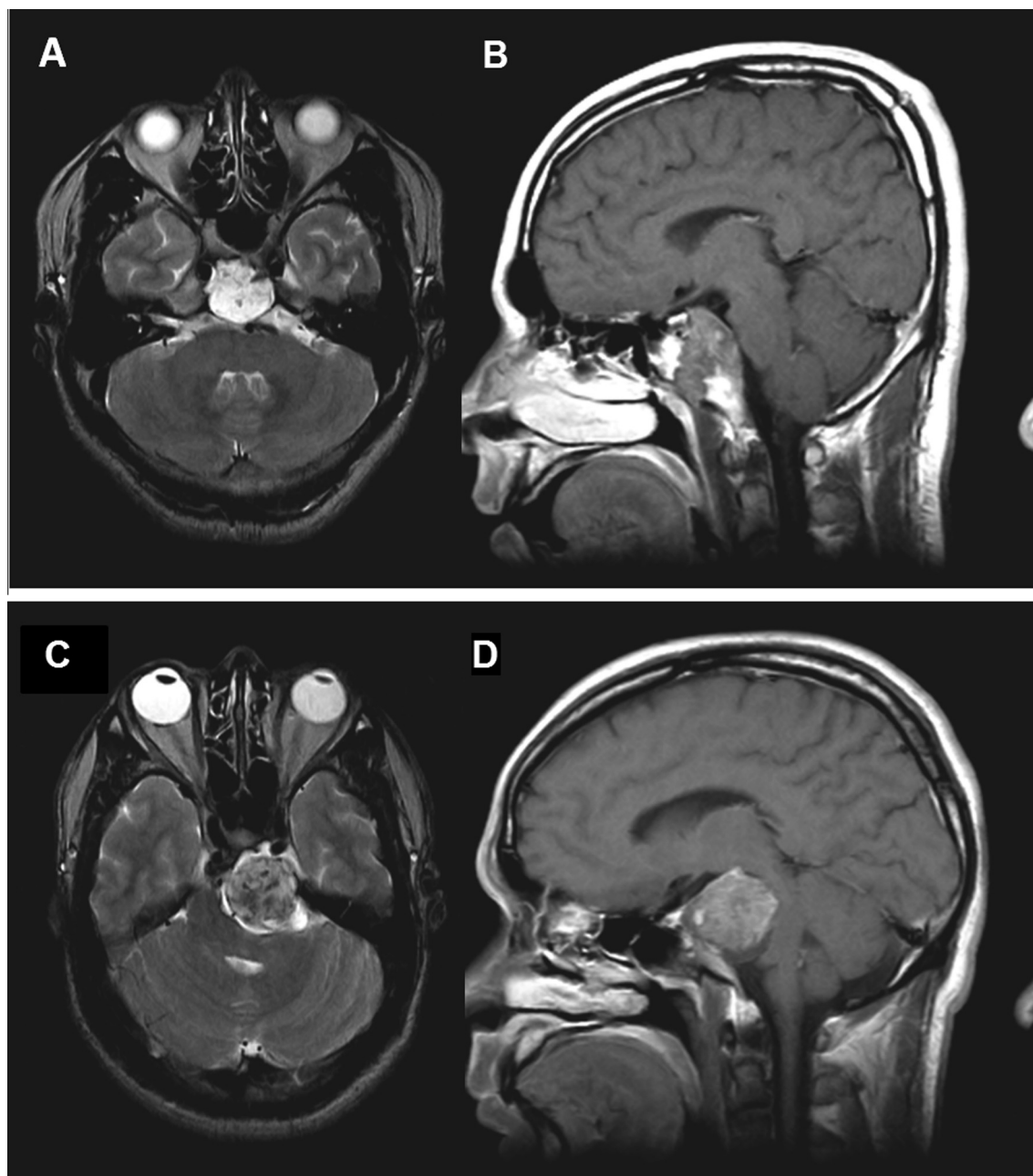


Fig. 1. Growth pattern in clivus chordomas. (A) Axial T2-weighted and (B) sagittal T1-weighted with enhancement MRI of an endophytic clivus chordoma. (C) Axial T2-weighted and (D) sagittal T1-weighted with enhancement MRI of an exophytic clivus chordoma.

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