Analysis of Clinical Features and Outcomes of Skull Base Chordoma in Different Age-Groups

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OBJECTIVE: **The objective of the current study was to compare the clinical features and outcomes between younger and older patients with skull base chordoma (SBC).**

METHODS: **In this retrospective study, patients with SBC who underwent surgical treatment between February 2005 and December 2014 were included. Clinical features were recorded, including the signal intensity ratio of tumor to brain stem in T1 (R_{T1}), T2 (R_{T2}), and enhanced T1 (R_{EN}) sequences in primary patients with complete preoperative magnetic resonance images. The clinical features and outcomes were compared between younger (≤24 years) and older patients (≥25 years).**

RESULTS: **In the present study, 238 patients were included. Younger patients experience more aggressive resection than do older patients ($P = 0.045$), and the SBCs of younger patients tended to be located in the occipitocervical region compared with older patients ($P = 0.007$). R_{EN} value in the younger group was lower than in the older group ($P = 0.014$), and the value of R_{T2} was higher in younger patients than in older patients ($P = 0.015$). The risk of progression was higher in older patients compared with younger patients ($P = 0.030$); the risk of having a poor neurologic status in older patients was higher than in younger patients ($P = 0.044$).**

CONCLUSIONS: **In younger patients, there were more SBCs located in the occipitocervical regions, and younger patients tended to undergo more aggressive resection. The tumor signal intensity of younger patients with SBC was higher in T2 images but lower in enhanced T1 images. A younger age was a favorable factor for a longer progression-free survival and a good neurologic status at follow-up.**

INTRODUCTION

Chordoma is a slow-growing neoplasm that originates from embryonic notochordal remnants.[1,2] The morbidity of chordoma is 0.08–0.089 per 100,000 people, with a higher incidence in males than in females (0.01–0.016 per 100,000 vs. 0.06–0.066 per 100,000, respectively).[3-5] Chordoma grows with bone invasion and mainly occurs in the axial bones of humans. Chordomas are often located in the skull base and comprise as much as 32%–42% of all chordomas.[3,5,6] Skull base chordomas (SBCs) are usually located in or around the clivus, sphenoid bone, or petrous bone, and their growth often encompasses important nerves and blood vessels, thus, aggressive resection of SBCs is difficult and often leads to serious complications.[7] Chemotherapy is of little value in chordoma, and some

Key words
- Age
- Clinical feature
- Outcome
- Skull base chordoma

Abbreviations and Acronyms
CI: Confidence interval
K-M: Kaplan-Meier
KPS: Karnofsky Performance Status
MR: Magnetic resonance
OC: Occipitocervical
OS: Overall survival
PFS: Progression-free survival
R_{EN}: Signal intensity ratio of tumor to brain in enhanced T1 sequence
R_{T1}: Signal intensity ratio of tumor to brain in T1 sequence
R_{T2}: Signal intensity ratio of tumor to brain in T2 sequence
SBC: Skull base chordoma

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studies[8-10] have indicated that radiotherapy for chordoma is far from effective. The current standard therapy for chordomas is still complete resection plus postoperative radiotherapy.[11-13] With the development of new techniques in recent years, including the endoscope, intraoperative navigation, and neurophysiologic monitoring, more aggressive resection of an SBC can be attained.[14] In addition, the application of proton beams, carbon ions, and modulations in stereotactic techniques for radiotherapy have helped to deliver higher doses to the tumor region.[15] Nevertheless, high mortality and recurrence rates continue to affect neurosurgeons.[14]

There are many factors that affect the outcomes of patients with SBC, including race, gender, and tumor location, degree of erosion to surrounding tissues, resection grade, histopathologic type, and therapeutic method.[6,8,10,16-20] Several recent studies indicated that the outcomes are different among patients with SBC in different age-groups.[3,6,9,12,16,19,21,22] We found that the clinical characteristics and outcomes associated with SBCs were age dependent (**Figure 1**). The hypothesis of this study was that age can be a useful variable for studying SBC. Thus, the aim of the current study was to explore the relationship between age and SBC.

METHODS

Overview

This study was approved by the ethics committees of Beijing Tiantan Hospital and Capital Medical University. Informed consent was acquired from all patients at the beginning of this retrospective study. The authors report no financial benefit related to this project. All of the authors who participated in the data collection and data analysis were blinded to the results of similar studies.

Patients

In the present study, we included patients with SBC who underwent surgical treatment in our hospital between February 2005 and December 2014. For patients who underwent multiple surgeries, we used their primary surgery and the corresponding clinical records for the analysis. We used strict exclusion criteria that included the following: 1) patients whose clinical records were absent; 2) patients with ambiguous histopathologic diagnosis, (eg, unable to distinguish the tumor as chordoma or chondrosarcoma); 3) patients who had other serious diseases that may affect the outcome.

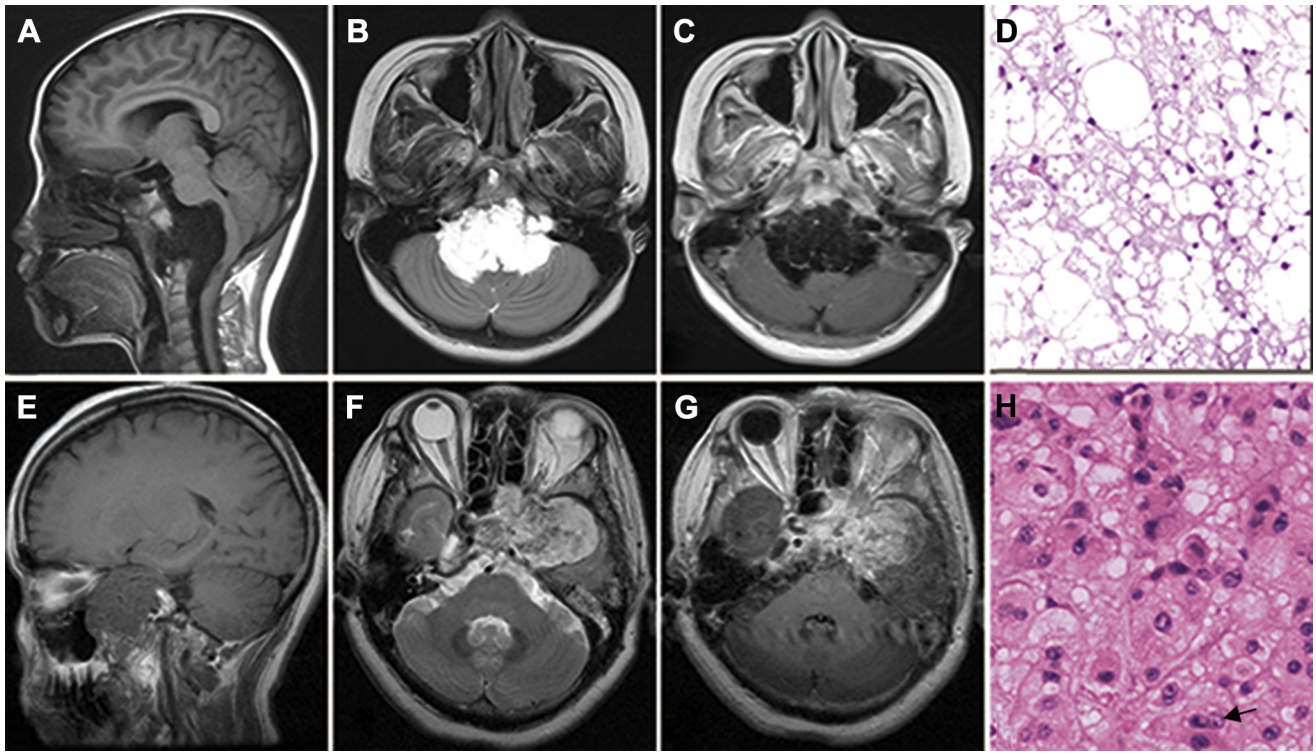

Figure 1. Clinical images of patients. (**A**–**D**) Images of patient 1, an 11-year-old girl. (**A**) Sagittal T1-weighted magnetic resonance (MR) image shows her tumor located in the occipitocervical region; the neoplasm is relatively homogeneous and hypointense compared with the brainstem. (**B**) Axial T2-weighted MR image shows the neoplasm with homogeneous high signal intensity. (**C**) Enhanced T1-weighted MR image shows no enhancement in the neoplasm. (**D**) Hematoxylin-eosin staining pathology slides of the tumor; tumor cells are rich in vacuoles, and no necrosis or nuclear division can be seen. (**E**–**H**) Images of patient 2, a 57-year-old woman. (**E**) Sagittal T1-weighted MR image shows her tumor located in the parasella region; the neoplasm is heterogeneous and relatively iso- to hyperintense compared with the brain. (**F**) Axial T2-weighted MR image shows a heterogeneous mass; the signal intensity was relatively lower than patient 1. (**G**) Enhanced T1-weighted MR image shows moderate enhancement in the neoplasm. (**H**) Hematoxylin-eosin staining pathology slides of the tumor; there are fewer vacuoles in the tumor cell than in patient 1, and nuclear division (*arrowhead*) can be seen in many tumor cells.