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Clinical commentary

Osteosarcoma of the skull base: An analysis of 19 cases and literature review

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ABSTRACT

Background: Osteosarcoma of the skull base is rarely observed; most published studies comprise case reports. The clinical features and optimal treatments have not been clearly established. The purpose of this article is to present 19 cases of skull base osteosarcoma and review the literature to analyse the clinical features and treatment of skull base osteosarcoma.
Methods: The clinical data of 19 patients with skull base osteosarcoma from January 2005 to December 2016 were retrospectively analysed; pertinent English literature from 1976 to 2016 was reviewed.
Results: Six female and 13 male patients were included. The ages ranged from 11 to 55 years (mean 34 years). Gross-total resection of the tumour was achieved in 13 cases, and nearly total resection was achieved in 6 cases. Five cases were treated with surgery alone, whereas 14 cases received comprehensive treatment. The follow-up period ranged from 3 to 132 months (mean 33 months) with 17 patients who underwent follow-up. The median survival durations of the patients who underwent surgery alone and who received comprehensive treatment were 18 and 50 months, respectively. The literature results were similar to the current findings. Overall, the 5-year survival rates of the patients in our series and in the literature were 30.5% and 37.8%, respectively.
Conclusions: Skull base osteosarcoma had a low complete resection rate, a high recurrence rate and a poor prognosis because of the complex anatomy and vital structures involved. Radical surgery with comprehensive treatment is most appropriate for this disease.

1. Introduction

Osteosarcoma is the most common malignant bone tumour; it primarily affects the long bones in children and adolescents. The incidence of osteosarcoma is approximately 1:100,000 per year [1], and it can be classified as long bone osteosarcoma or head and neck osteosarcoma by location. A total of 6–10% of all osteosarcoma cases have presented in the head and neck, usually in the mandible and less frequently in the maxilla [2,3]. Osteosarcomas that arise from the skull base or the head and neck with skull base invasion (skull base osteosarcoma) are very rare. Approximately 120 cases have been confirmed in the English literature from 1976 to 2016 [4–15]. Chen YM and his colleagues reported 64 cases of maxilla and skull base osteosarcoma [16], and Shellenberger TD and his colleagues reported 96 cases of sinonasal tract, ear and skull base osteosarcoma [17]; however, they did not present the exact number of cases of skull base osteosarcoma or specifically address the characteristics of skull base osteosarcoma. There is no systematic review to clarify the clinical features and treatment of skull base osteosarcoma. The clinical features of skull base osteosarcoma remain unclear, and there is a lack of consensus regarding the optimal therapeutic strategy. In this study, we present the largest series, to our knowledge, of osteosarcoma of the skull base in a single centre and review the literature with a focus on the clinical features and comprehensive treatments.

2. Materials and methods

2.1. Patients

Nineteen patients with osteosarcoma that originated in the skull base or head and neck osteosarcoma that involved the skull

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base who were treated at the Cancer Hospital, National Cancer Centre of China, Chinese Academy of Medical Sciences, Peking Union Medical College from January 2005 to December 2016 were included in this study. All patients were surgically treated, and the diagnosis of osteosarcoma was verified by histological examination. The medical data were retrospectively reviewed with regard to age, sex, tumour site, clinical symptoms and signs, surgical procedures, radiological features, treatment, extent of resection, recurrence, and follow-up results.

2.2. Neuroimaging studies

All patients underwent skull base computed tomography (CT) scans and magnetic resonance imaging (MRI) with and without enhancement preoperatively. Enhanced MRI was performed 1–3 months after surgery to evaluate the extent of tumour removal, followed by every 3–6 months to detect recurrence. To more clearly describe the size and scope of the tumour, we defined different compartments of the skull base and head and neck, which included the anterior cranial fossa, middle cranial fossa, posterior cranial fossa, maxillary sinus, sphenoid sinus, ethmoidal sinus, orbit, infratemporal fossa and pterygopalatine fossa; tumours that involved the anterior cranial fossa, middle cranial fossa and the clivus were defined as skull base osteosarcoma.

2.3. Management

2.3.1. Surgical procedures

A preoperative biopsy was performed when possible, and a multidisciplinary preoperative evaluation was conducted to determine whether induced chemotherapy and/or radiotherapy was needed according to the biopsy results, as well as to plan the surgical strategy. The tumour was removed by a neurosurgical team or a multidisciplinary team composed of neurosurgeons and head and neck surgeons. Different surgical approaches were adopted for tumour removal according to the location and extension; for example, the extended anterior skull base epidural approach was used for anterior skull base tumours; a transnasal approach was implemented for clival and sellar region tumours; the maxillary swing approach was utilised for giant tumours, which involved the central skull base and/or pterygopalatine fossa; and an infratemporal approach was implemented for tumours located in the infratemporal fossa and middle skull base.

2.3.2. Chemotherapy and radiotherapy

Postoperative chemo radiotherapy was administered to prolong the survival time of patients. The radiotherapy dose was 30–64 Gy. The chemotherapy regimens included the combination of gemcitabine and cis-platinum (GP) and the combination of ifosfamide and cis-platinum. If a substantial tumour involved vital structures (such as the cavernous sinus, the optic chiasma or the internal carotid artery), preoperative induced radiotherapy and/or chemotherapy according to the biopsy histopathology should be considered to reduce the tumour size to facilitate surgical resection.

2.4. Follow-up and statistical analysis

Follow-up studies were obtained by conducting outpatient office visits and/or telephone interviews. The follow-up data included the postoperative therapeutic regimens, presence of metastatic and recurrent disease, subsequent treatments, and time and cause of death. The cumulative probability of survival was calculated using the Kaplan-Meier method, and the log-rank test was used to compare survival curves. A P-value less than 0.05 was considered statistically significant. All statistical analyses were performed using SPSS version 16.0 for Windows (Chicago, IL).

2.5. Literature reviews

Digital literature searches without time restrictions were performed in the PubMed databases for relevant English publications until July 2016. The search terms included skull base osteosarcoma and head and neck osteosarcoma. The inclusion criteria included primary skull base osteosarcoma, i.e., that arose from the skull base, and secondary skull base osteosarcoma, i.e., that arose from the head and neck with skull base invasion. The literature contained at least one example of the data including the patient's age, sex, site of tumour, presenting symptom, grade, histologic subtype, treatment modality, surgical approach, recurrence, imaging findings and survival. The exclusion criteria were as follows: (1) review articles, (2) duplicate publication, (3) studies published only as an abstract or in conference proceedings.

3. Results

3.1. Clinical features

The clinical characteristics at the initial presentation of the patients were summarised in Table 1. There were 6 female and 13 male patients in this series. The patient age at presentation ranged from 11 to 55 years, with a mean age of 34 years. The mean duration of symptoms prior to admission was 2.9 months (ranging from 20 days to 12 months). Twelve of 19 patients received initial treatment for tumour (including 2 patients with a diagnosis of osteosarcoma based on biopsy histopathology at other hospitals), and the remaining 7 patients had been treated elsewhere and were transferred to our institute for the treatment of a recurrent tumour. Osteosarcoma originated in the skull base in 9 cases, including the anterior skull base in 3 cases, the middle skull base in 4 cases and the posterior skull base in 2 cases. Head and neck osteosarcoma invaded the skull base in 10 cases, and arose from the mandible in 5 cases and the maxilla in 5 cases. The main clinical manifestations included facial lumps (6 cases), epistaxis (3 cases), headache (2 cases), toothache and loose teeth (3 cases), exophthalmos (2 cases) and other symptoms (3 cases). Skull base osteosarcoma was likely induced by radiation in 3 patients. Two patients (case 7 and 19) had received radiation therapy for nasopharyngeal carcinoma 17 and 25 years earlier and underwent surgical resection of his and her skull base osteosarcoma 7 years and 5 months earlier respectively. Another patient (case 11) had received preoperative radiotherapy (50 Gy) for cementoma of the upper jaw and subsequently underwent maxillary neoplasm resection 16 years ago; a tumour that arose from the original site with the mid-anterior skull base invasion was removed and histologically re-diagnosed as osteosarcoma 5 years ago.

3.2. Imaging findings

According to the radiological examinations, the tumours were located at the anterior skull base in 8 cases, the middle skull base in 4 cases, the posterior skull base in 1 case, the mid-anterior skull base in 3 cases and the mid-posterior skull base in 3 cases. The tumors had both intra- and extra cranial extensions in 6 cases. The tumours occupied one skull base compartment in 8 cases, 2 compartments in 8 cases, and 3 compartments in 3 cases. The largest tumour diameter ranged from 3.7 to 15.0 cm, with an average of 8.1 cm. The CT scan displayed different degrees of bone destruction and ossification in all cases. Tumours exhibited isointensity on T1-weighted images and moderate or hypointensity on T2-weighted images; the signal distribution was heterogeneous, and there was moderate to significant enhancement after contrast injection (Fig. 1).