Case Report

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Primary maxillofacial chordoma: a rare case report and literature review

Yu Cui[1], Xiang-yan Cui[1], Tingting Yu[1], Zhan-peng Zhu[2,*] and Xin Wang[1,*]

Abstract

Primary maxillofacial chordoma is extremely rare. We herein report a very rare case of a recurrent maxillofacial chordate tumor that was diagnosed in a 56-year-old woman who underwent three tumor resections. After surgical treatment, the patient healed well with an Eastern Cooperative Oncology Group score of 1. She was discharged to a local hospital for adjuvant radiotherapy. Close follow-up was ongoing at the time of this writing. Radical surgery and adjuvant radiotherapy remain the main treatment strategies for chordoma. Postoperative radiotherapy is particularly important. Our experience is to administer a total dose of 50 Gy to a clearly delineated target. If appropriate comprehensive treatment is available, distant metastasis of primary chordoma is rare, and neck dissection is therefore not generally recommended. Neck lymph node dissection is generally not recommended.

Keywords

Chordoma, maxillofacial, resection, radiotherapy, recurrence, case report

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Introduction

Chordoma is a rare type of sarcoma that occurs along the axial skeleton from the base of the skull to the sacrum and coccyx (tailbone).[1,2] Head and neck chordomas are commonly found in the nasopharynx, and most originate from the nasal cavity. Chordomas in the maxillary sinus are uncommon,[3] whereas primary maxillofacial chordomas are extremely rare.[4] We herein present a rare case of a primary maxillofacial

[1]Department of Otolaryngology Head and Neck Surgery, the First Hospital of Jilin University, Changchun, P. R. China
[2]Department of Neurosurgery, the First Hospital of Jilin University, Changchun, P. R. China

*These authors contributed equally to this work.

Corresponding author:
Xin Wang, Department of Otolaryngology Head and Neck Surgery, the First Hospital of Jilin University, 71 Xinmin Street, Chaoyang District, Changchun, Jilin 130021, P. R. China.
Email: 15804300593@163.com

chordoma that was diagnosed in a 56-year-old woman who underwent three tumor resections. We also discuss the examination findings and treatment plan and present a literature review of maxillofacial chordoma.

Case report

A 56-year-old woman was admitted to our hospital because of a 6-month history of facial swelling and a bulge in the left maxillofacial area. The patient had undergone mass resection in the left maxillofacial region 17 years previously. Two years before the current presentation, mass resection was performed in the same location. This was followed by skin grafting from the femoral area. Pathological examination of the specimen confirmed a maxillofacial chordoma. No additional treatment was performed after the second resection.

At the present admission, the bulge was located on the left zygomatic arch below the last skin flap with a diameter of about 4.0 to 4.5 cm. The mass was swollen and hard with no tenderness. The mass did not affect mouth movement or swallowing. The patient's binocular vision and eye movement were normal. Ultrasound examination showed a hypoechoic mass with uneven echogenicity underneath the left cheek and an acceptably clear border. A punctate blood flow signal was detected. The border between the mass and bone was unclear. Routine preoperative enhanced computed tomography showed that the maxillofacial lesion involved the left maxillary sinus, oropharyngeal and maxillofacial soft tissue, and bone (Figure 1).

As described above, the patient had previously undergone tumor resection twice without further treatment. As requested by the patient, the resection margin was set along the skin flap. After complete tumor resection (Figure 2), the zygomatic arch and the anterior wall of the maxillary sinus were found to be defective. After suction of the sinus cavity, we found that the mucous membrane on the wall of the remaining maxillary sinus was smooth. Necrotic tissue was found and removed from the infratemporal fossa and internal pterygopalatine fossa using a curette. The skin flap was sutured at the end of the surgery. The postoperative pathological examination confirmed a chordoma. Microscopically, fibrous tissue was

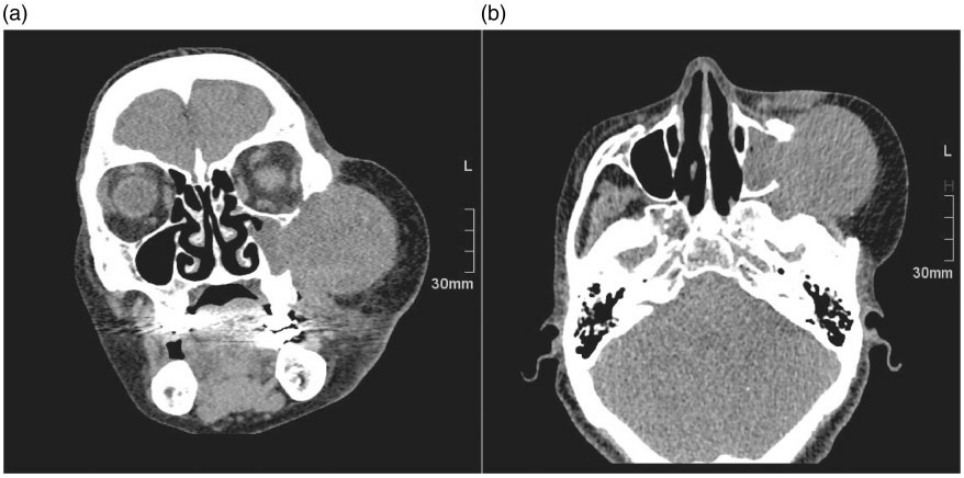

Figure 1. Preoperative computed tomography scan showing the mass on the left maxillary sinus wall. The computed tomography number of the maxillofacial mass was 20 to 40 HU.

interwoven with mucinous tumor tissue. The tumor cells were cuboid, circular, or polygonal in shape with a clear nucleus and low nucleus-to-cytoplasm ratio. The immunohistochemical examination results were as follows: Ki-67 (+10%), pan-cytokeratin (+), vimentin (+), epithelial membrane antigen (partially +), S-100 (partially +), cytokeratin 20 (−), and cytokeratin 7 (−) (Figure 3). The surgical wound healed well with an Eastern Cooperative Oncology Group score of 1. For economic and geographical reasons, the patient was discharged to a local hospital for adjuvant radiotherapy. Telephone follow-up was ongoing at the time of this writing, and no signs of tumor recurrence had been found.

This study was approved by the ethics committee of the First Hospital of Jilin University. Written informed consent was obtained from the patient.

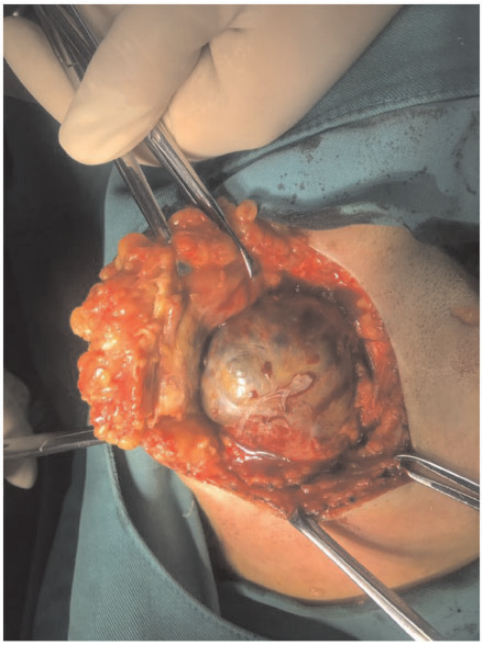

Figure 2. Tumor resection. The tumor had a capsule and was completely resected.

Discussion

A chordoma is a tumor with low-grade malignancy. More than 50% of cases are found in the sacrococcygeal region, 30% are found in the base of the skull, and few are found in other vertebrae.[1,2] A chordoma that occurs in the skull base should be differentiated from a pituitary tumor, meningioma, craniopharyngioma, nasopharyngeal carcinoma, and chondrosarcoma. A chordoma in the sacrococcygeal region should be differentiated from a giant cell tumor, teratoma, neurogenic tumor, chondrosarcoma, and osteosarcoma.

A chordoma can develop at any age, but it is most common in middle-aged men. It grows slowly and can infiltrate the

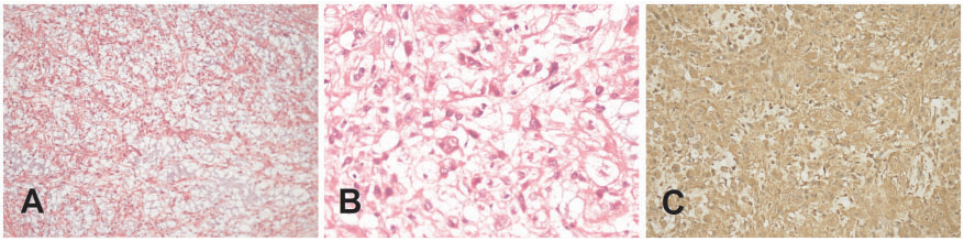

Figure 3. Histopathological examination findings. (a) Fibrous tissue was interwoven with mucinous tumor tissue [×100, hematoxylin and eosin (H&E) staining]. (b) The tumor cells were cuboid, circular, or polygonal in shape with a clear nucleus and low nucleus-to-cytoplasm ratio (×400, H&E staining). (c) Immunohistochemical examination revealed visible brown particles in the tumor cytoplasm and positivity for vimentin.

surrounding bone structure and soft tissue. Primary chordomas in the base of the skull can spread to the cranial cavity, nasopharynx, and sinus. Maxillary sinus chordomas are uncommon,[3] and maxillofacial chordomas are extremely rare.[4] In the present case, we found that the zygomatic arch and the anterior wall of the maxillary sinus were defective. After suction of the sinus cavity, we found that the mucous membrane on the wall of the remaining maxillary sinus was smooth. The tumor was located underneath the skin flap from the previous resection. Although no sign of tumor growth was present along the wall of the maxillary sinus, this finding indicated that the primary insult was the left maxillofacial region and that no infiltration from other structures had occurred.

The diagnosis of chordoma relies on the pathological examination findings. Some studies have suggested that immunohistochemistry combined with magnetic resonance imaging can significantly reduce the rate of misdiagnosis.[5] Chordomas express both epithelial and mesenchymal characteristics. Histopathology and immunohistochemistry can assist in the differential diagnosis of chordomas. The pathological classification of chordomas can be divided into three subtypes: conventional, chondroid, and dedifferentiated. Chordoma-specific brachyury immunostaining examination is valuable for the diagnosis of chordoma. Chordoma-specific brachyury immunostaining was not performed in the present case because of the limited pathological conditions in our hospital. However, the clinical course combined with the microscopic findings indicated that this patient had the conventional subtype of chordoma. Her tumor was positive for pan-cytokeratin and S-100, which was supportive of the diagnosis. Negativity for cytokeratins 20 and 7 can exclude tumor metastasis from the gastrointestinal system.

Radical surgery and adjuvant radiotherapy remain the main treatment strategies for chordoma. However, most chordomas are located in the bone tissue, and it is difficult to perform frozen section analysis of the margin. Additionally, chordomas are often located in close proximity to organs or tissues at risk. The resection margin should be tight to avoid damage to neighboring important tissues or structures, such as blood vessels. Thus, postoperative radiotherapy is particularly important. Chemotherapy plays a role in the prevention of tumor recurrence.[6] Our experience is to administer a total dose of 50 Gy to a clearly delineated target. In the present case, the patient received postoperative radiotherapy at another local hospital, and the prescription dose of radiotherapy was unclear. However, the current chordoma exhibited *in situ* recurrence. If appropriate comprehensive treatment is available, distant metastasis of primary chordoma is rare, and neck dissection is therefore not generally recommended. Neck lymph node dissection is generally not recommended.

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Declaration of conflicting interest

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ORCID iD

Yu Cui https://orcid.org/0000-0001-9107-3579

Tingting Yu https://orcid.org/0000-0001-9438-1016

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