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Case Report

Digital resection and reconstruction of TMJ synovial chondrosarcoma involving the skull base: report of a case

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Abstract: Synovial chondrosarcoma (SCS) is a very rare malignant cartilaginous tumor. To the best of our knowledge, only three reported studies presented the involvement of the temporomandibular joint (TMJ). Hereby, we present a case of surgical management of a SCS of the TMJ, arising from SC and involving the skull base. The surgical procedure includes digital design, resection guided by digital templates, as well as immediate reconstruction with free iliac bone graft (IBG) and pedicled sternoclavicular joint (SCJ). At 1-year follow-up, the TMJ function and form were improved with no sign of local recurrence or metastasis to bone or other joints. However, its distant metastasis to lung was observed.

Keywords: Temporomandibular joint, synovial condrosarcoma, digital template, iliac bone graft, sternoclavicular joint

Introduction

Synovial condrosarcoma (SCS) is a very rare cartilaginous malignancy arising de novo or secondary to synovial chondromatosis (SC) [1]. It frequently affects the large joints, such as the knee, hip and ankle1. To our knowledge, only three reports have been published to describe SCS of the temporomandibular joint (TMJ), but none of them presented the involvement of the skull base [2-4].

Here, a rare case with SCS of the TMJ, arising from SC and involving the skull base is presented. The surgical procedure includes the resection and reconstruction at one stage. We focus on its surgical management: (1) computer-assisted design was made before surgery; (2) the resection was guided by a digital template; (3) the reconstructions of the skull base and the TMJ condyle were done by harvesting a free iliac bone graft (IBG) and transferring a pedicled sternoclavicular joint (SCJ).

Report of a case

Clinical presentation

A 56-year old woman was referred to our department with a history of pain in the right preauricular region for 1 year. The patient had also noticed a slight swelling in that region over 3 years. There was no previous history of facial trauma or any event contributed to such symptoms. On physical examination, a swelling in the right preauricular region was observed with a slight pain on palpation. Maximal interincisor mouth opening (MIO) was 27 mm, with a deviation of the mandible to the right side. Crepitation was presented in the right TMJ. The occlusion of the patient was stable.

Radiologic examination

The computed tomographic (CT) scan showed a lesion (measuring 45 mm×36 mm×32 mm) arising from the right TMJ and extending to the infratemporal fossa (ITF) as well as the middle-

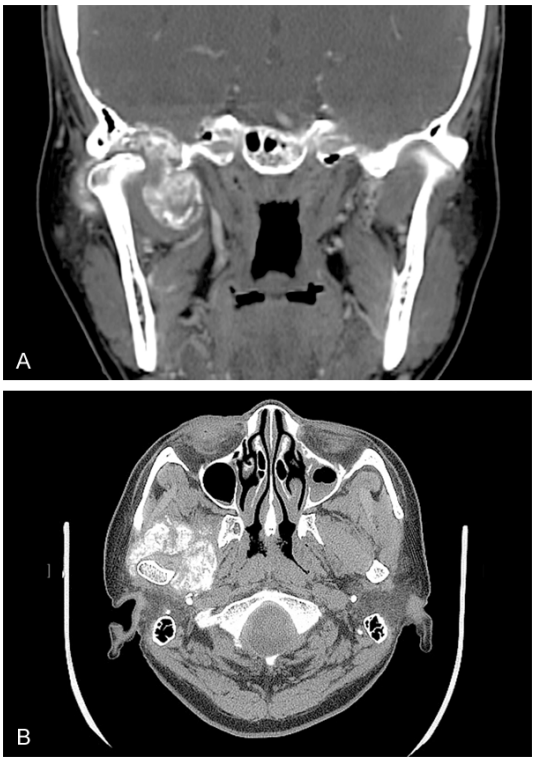

Figure 1. CT showed the lesion extended to the intracranial fossa through the defect of the skull base with dura intact.

cranial fossa. The contour of the lesion was relatively clear, and high-density calcified signals were found inside. The erosion of the right skull base (perforation is 20 mm×12 mm) was also observed (**Figure 1**). Magnetic resonance imaging (MRI) demonstrsted the extension of the lesion to the subdural space, but dura was intact. It was highly suspicious for SC, but chondrosarcoma could not be excluded.

Digital design of the resection

The data of the CT scan (slice thickness was 0.625 mm; GE, USA) were imported to Proplan 1.3 software (Materialize Co, Leuven, Belgium) for three-dimensional (3D) reconstruction [5, 6], and the lesion was segmented out (**Figure 2**). The posterior part of the lesion involved the petrous bone, with 6 mm to the external auditory canal (EAC) and 5 mm to the internal carotid. The medial part of the lesion extended to the spinous foramen and surrounded the middle meningeal artery, with 5 mm to the jugular vein. The posteromedial part of the lesion involved the bilaminar zone of the disc with 3 mm to the maxillary arteriovenous.

Osteotomy lines were designed on the 3D reconstruction. Two digital osteotomy templates were then manufactured to mark the borders of the lesion (**Figure 2**).

Resection guided by digital templates

Under the general anesthesia, a preauricular temporal approach and a submandibular approach were used to expose the temporal bone, the right zygomatic arch, the joint capsule and the ramus. The trunk of the facial nerve was separated and preserved. The procedure of the resection was as follows (**Figure 2A**): (1) the condyle was osteotomied at the level of the sigmoid notch. (2) the zygomatic arch osteotomy was implemented guided by the digital osteotomy template (**Figure 2B**, **2C**), and the anterior part of the zygomatic arch (15 mm) was pulled downward with the masseter muscle attached. (3) a 3 cm×4 cm temporal craniotomy was performed above the superior border of the tumor marked by the digital template; By carefully separating the dura to the level of oval foramen and retracting the temporal lobe, the lesion was found encapsulated without any attachment to the dura. (4) the anterior and posterior osteotomies were implemented based on the design. (5) the mass was resected as a whole by releasing its anterior, medial and posterior connections.

Immediate reconstruction

Frozen biopsy demonstrated: (1) the lesion was mostly suspicious for SC, but SCS could not be excluded; (2) no tumor cells found in the surrounding soft tissue. Thus, immediate reconstruction was performed as follows: (1) the temporal bone was repositioned and fixed with titanium plates; (2) the free IBG was used to repair the skull base and glenoid fossa defect [7] (**Figure 3A**); (3) the zygomatic arch was repositioned and fixed with titanium plates; (4) the pedicled deep temporal fascial fat flap (DTFFF) was transferred to be an interposition between the “new fossa” and “new condyle” [8]; (5) the pedicled SCJ was harvested to reconstruct the condyle [9] (**Figure 3B**).

Pathological examination and follow-up

The specimen was about 47 mm×38 mm×35 mm. The histological examination showed synovial and cartilage hyperplasia with local

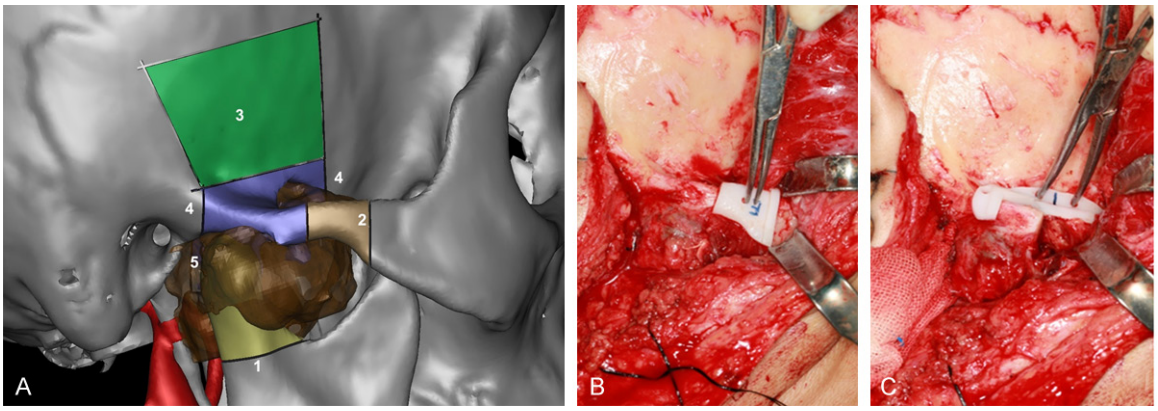

Figure 2. The surgical procedure. A. The resection procedure: 1, condyle osteotomy. 2, temporary zygomatic arch osteotomy. 3, temporary craniotomy. 4, anterior and posterior osteotomy. 5, resection of the tissue between the TMJ capsule and the EAC. B, C. The zygomatic arch osteotomy guided by the digital templates.

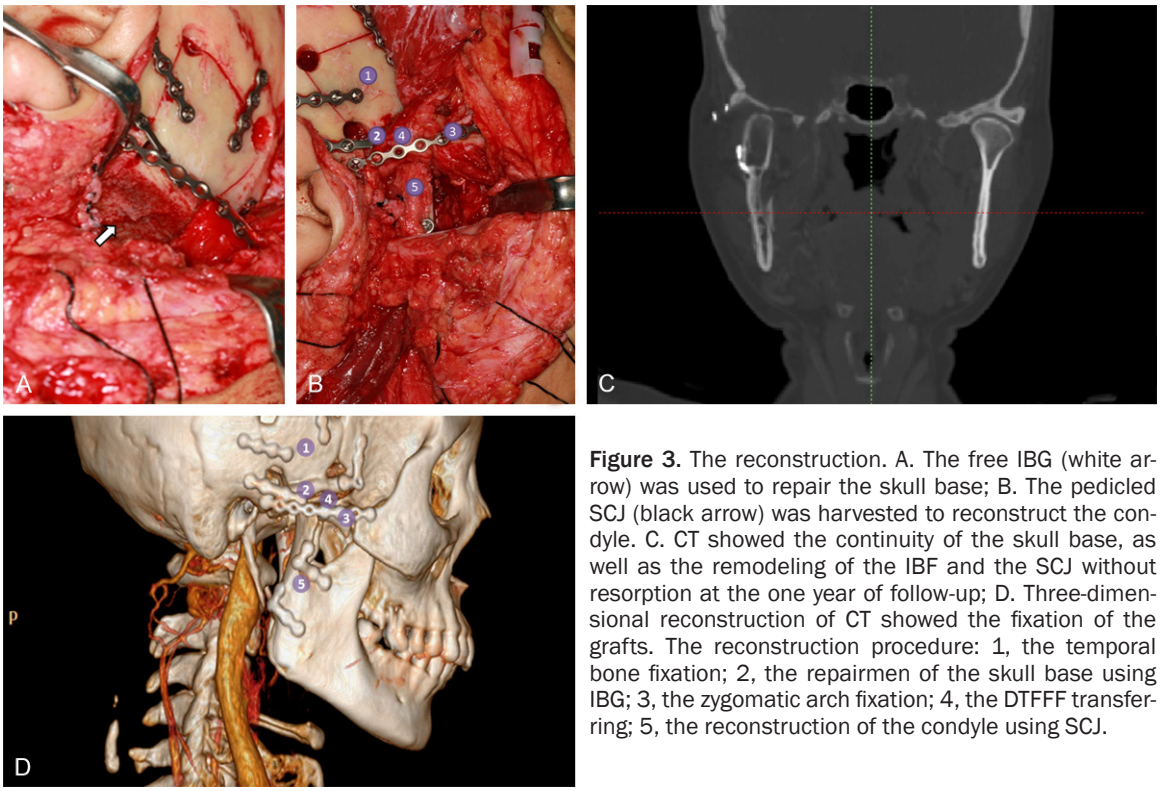

Figure 3. The reconstruction. A. The free IBG (white arrow) was used to repair the skull base; B. The pedicled SCJ (black arrow) was harvested to reconstruct the condyle. C. CT showed the continuity of the skull base, as well as the remodeling of the IBF and the SCJ without resorption at the one year of follow-up; D. Three-dimensional reconstruction of CT showed the fixation of the grafts. The reconstruction procedure: 1, the temporal bone fixation; 2, the repairmen of the skull base using IBG; 3, the zygomatic arch fixation; 4, the DTFFF transferring; 5, the reconstruction of the condyle using SCJ.

absence of clustered malignant cartilage cells. A diagnosis of SCS (grade II of Evan' system [10]) arising from SC was made.

At one year follow-up, the patient recovered with neither facial asymmetry nor paralysis. The MIO was 30 mm, while the occlusion was stable. Clinical examination and imaging showed no evidence of local recurrence as well as the resorption of IBF and SCJ. Furthermore, the bone grafts remodeled to fit the fossa better (**Figure 3C**, **3D**). But metastasis to the lung was found 1 year after the surgery.

Discussion

SC is a rare benign tumor characterized by the subsynovial cartilage metaplasia [11, 12]. However SCS is rarer than SC in TMJ. In our department, 144 cases of SC were treated

from 2004 to 2014, but only one SCS was found [12]. The malignant transformation of SC to a SCS is also rare 1. In the studies of major joints (hip, knee, et al), the incidence is estimated to be in the range of 1%-6% [1], while the average transformation period is 20 years [1]. Among 3 studies on SCS of the TMJ [2-4], only Coleman's [4] and our report could confirm SCS arising secondary to SC according to the histological examination. The main differential diagnosis of SCS is SC. However, it's difficult to distinguish SCS from SC clinically and radiographically [1]. The final diagnosis should be relied on the pathologic examination with the evidence of malignancy [13].

Radical resection is considered as the treatment of choice for SCS in the three previous reports [2-4]. Compared with the other three, our case was the largest SCS of the TMJ, which occupied the condyle, the whole ITF and extended to the skull base. The challenge of our surgery was removing such huge SCS with complete capsule, as well as the reconstructions of the skull base defect and the condyle.

The surgical approach is difficult for tumors in infratemporal space, which is deeply located and related to rich vascular anastomoses. The typical approaches to this space include the transcondylar, transcoronoid, or transzygomatic approaches [14]. In this case, the resection of the condyle is not enough for surgical exposure, so a temporary zygomatic arch osteotomy was also applied. With masseter muscle attached, a part of the zygomatic arch was pushed downward to provide an adequate surgical field.

With computer technology assisted, the relationship between tumor and important adjacent anatomic structures could be presented clearly, guiding the design of the resection. Resecting the posterior part of the tumor should consider the internal carotid and EAC. The damage to the former one leads to the massive bleeding, while damage to the latter one leads to the EAC defect. The medial resection should take care of the middle meningeal artery and the jugular vein. The former one was involved in the tumor, thus electrocautery and hemostatic materials should be carefully applied. The latter one should be protected in case of excessive bleeding. The postero-medial resection should be careful about the maxillary arteriovenous to prevent excessive bleeding.

The digital osteotomy templates, which could be manufactured according to the digital design to achieve precise resection, were previously reported in resections of osteochondroma [5, 6]. There was no damage to the critical nerves and vessels in the surgery, and no local recurrence after the surgery, which suggests the precision of our digital design and resection.

Tumors involving the TMJ should consider the immediate reconstruction of the condyle to improve TMJ function and form, which is not conducted in the other three case reports. The SCJ has several advantages: (1) it has an interarticular fibrocartilage disc which resembled TMJ [15]; (2) it is usually pedicled with better blood supply; (3) the muscle attached to it could be used to fill the soft tissue defect [9]. Free costochondral graft (CCG) is not suitable in this case because of its poor blood supply in such old woman. And total joint prosthesis is not appropriate because of its inability to fill the large soft tissue defect. Thus, we chose SCJ in our case to prevent the necrosis or resorption of the "new condyle", and fill the large defect as well. The shape of the iliac bone is like both the glenoid fossa and the skull base bone, and the success rate of the transplantation is very high based on our clinical experience. So we harvested the IBG to repair the defect of the skull base. To avoid the resorption of the SCJ and the IBG, as well as the bony fusion between them, we transferred the DTFFF as an interposition. We chose the DTFFF because it usually does not absorb and could help the "new joint" move smoothly [8, 9]. One year after the surgery, the CT showed the remodeling of the SCJ and the IBF without resorption, demonstrating the bone grafting was successful under an adaptive pressure.

Besides the complete surgery, the prognosis including the recurrence and the metastasis of SCS is related to the histological grading of chondrosarcoma (the Evans' grading system [10]). Metastasis to bone and joints of chondrosarcoma is rare, and its rates in grade I, II, III (low, moderate and high grade, respectively) were 0%, 10% and 17% [16]. But the distant metastasis of condrosarcoma to the lung or brain is common [13]. In our case, SCS was in grade I, and neither recurrence nor metastasis

to bone or joints was found one year after the surgery. But metastasis to lung was observed. Thus, ECT or PET-CT should be recommended as a regular examination in patients with SCS.

In conclusion, although very rare, SC of the TMJ could transfer to SCS. With the guide of the digital osteotomy template, the resection for SCS of the TMJ extending to the skull base could be of precision. Immediately harvesting the IBF to repair the skull base, as well as the SCJ to reconstruct the condyle could improve the TMJ function and form.

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Disclosure of conflict of interest

None.

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