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Chondrosarcoma of the maxilla: a case report and review of current literature

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ABSTRACT

Chondrosarcoma is a malignant mesenchymal tumour that is the second most common bone tumour after osteosarcoma. Its diagnosis is among the most difficult ones in tumour pathology. Here, we report a very unique case of chondrosarcoma in maxillary sinus.

A 45-year-old woman presented with a tumour on the left side of the maxillary sinus. CT and MRI imaging showed an abnormal mass destructing surrounding tissues. The final diagnosis of chondrosarcoma could not be made for a long time due to lack of correlation between clinical and microscopic examinations. The patient underwent left subtotal maxillectomy via Weber-Ferguson incision, bilateral ethmoidectomy, sphenoidectomy, and right upper turbinectomy, and excision of nasal septum, left frontal sinus, and left exenteration of orbit.

Histological changes in the small biopsy specimen may be not sufficient for definitive diagnosis. Our case shows that radiography combined with histopathology is necessary to make the final diagnosis. The presented case revealed that chondrosarcoma can be a heterogeneous tumour. Collecting tissue samples from different locations is essential for improving diagnosis and reducing diagnostic error. Combining clinical data even with uncertain microscopic examination may be a solution in borderline and complicated cases.

Key words: chondrosarcoma, maxilla, sinus, review

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Introduction

Chondrosarcoma is a malignant mesenchymal tumour that is the second most common bone tumour after osteosarcoma. It usually occurs in iliac bone, ribs, and long bones. Chondrosarcoma most commonly affects adults, with no sex predilection [1]. Most chondrosarcomas arise *de novo*; however, they are common in patients with osteochondromas, Maffucci syndrome, Ollier's, and Paget's disease [2–4]. The genetic mechanism in chondrosarcoma involves amplification of the *c-myc* and *fos/jun* proto-oncogenes. Genetic aberrations found in low-grade conventional chondrosarcomas include amplification of 12q13 and loss of 9p21. The 12q13 region harbours MDM2, a negative regulator of p53. The 9p21 region harbours two cell cycle regulators: *CDKN21/p16/INK4A* and *INK4A-p14*. High-grade

chondrosarcomas are characterised by the loss of *INK4A/p16* expression, suggesting a role for chondrosarcoma progression [5–7]. There are three grades of the malignancy, depending on cellularity, atypia, mitotic activity, nuclear size, and surrounding matrix composition [8]. The rate of metastasis depends on the grade of the tumour. There is 10% risk of metastasis for grade 1 and 70% for grade 3. Chondrosarcoma tends to metastasise to the lungs [8]. The overall five-year survival rate is about 90% for grade 1, 81% for grade 2, and 43% for grade 3 [9]. The diagnosis of chondrosarcoma is among the most difficult in tumour pathology. Histological changes in the small biopsy specimen may be insufficient for definitive diagnosis. In such cases, CT and MRI are required. 12% of all chondrosarcomas are located in the head and neck region and occur mostly in adults between the third and sixth decade of

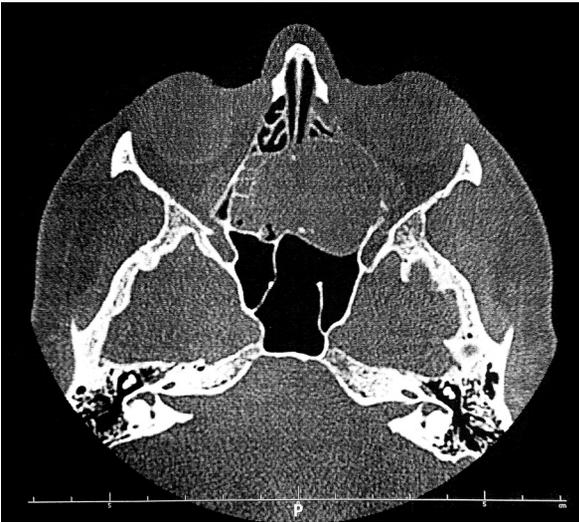


Figure 1. CT image. Abnormal mass measuring 59 × 41 mm on the left side of the maxillary sinus, which caused destruction of nasal septum reaching the medial wall of the right maxillary sinus

life. Patients may present nonspecific symptoms, which vary greatly depending on the location of the tumour [8, 10, 11]. Primary sites of the head and the neck include larynx, thyroid cartilage, and arytenoids [12]. However, chondrosarcomas can occur in all other sites of the craniofacial compartment in which cartilage is found, such as mandible, paranasal sinuses, nasopharynx, base of the skull, and maxilla [13, 14]. The treatment of choice is wide surgical resection of all the involved structures with negative margins and preservation of function if possible. Radiotherapy may be used for prevention of local recurrence. The crucial factors determining the prognosis of chondrosarcoma of the maxilla are the anatomic localisation, size, extension, and histological grade of the tumour [15]. The five-year survival rate for chondrosarcomas of the jaw and facial bones is reported to be 67.6%. However, life-long follow-up is essential, because chondrosarcomas show a wide variation in time of recurrence and metastasis [16].

Case report

A 45-year-old woman presented to the ER with complaints of epistaxis and left nasal obstruction. CT and MRI imaging showed an abnormal mass measuring 59 × 41 mm on the left side of the maxillary sinus, which caused destruction of the nasal septum reaching the medial wall of the right maxillary sinus (Fig. 1). All nasal conchae were infiltrated and damaged. Residual fluid was found in many ethmoidal cells. Mucosa of the left sphenoid sinus was thickened. During the first

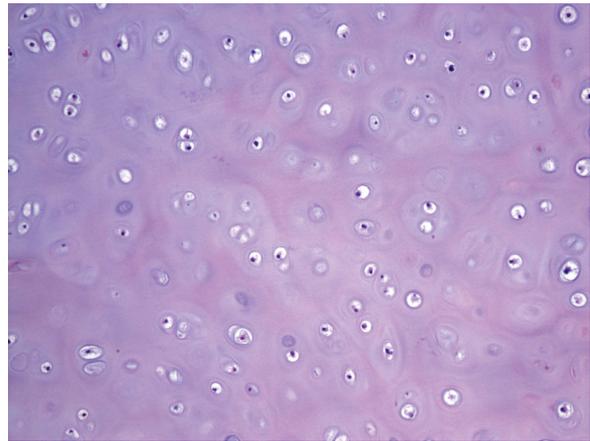


Figure 2. Microscopic image of chondroma. Primary objective magnification 10 ×. Tumour composed of mature hyaline cartilage in which cells resemble normal chondrocytes and produce cartilaginous matrix

examination, there was a suspicion of left alveolar ridge infiltration. The mass adhered to the medial wall of the left eye socket and infiltrated the left medial and inferior straight muscles. Pterygopalatine fossa and base of middle cranial fossa were not affected. Lymph nodes were not enlarged. The histopathological examination revealed mesenchymal hamartoma.

One month later the patient was hospitalised in the Otolaryngology Clinic, where a clinical examination was performed again. The right nasal tract was unobstructed with no pathological discharge. The left nasal tract was obstructed by a hard mass. No pathological changes in oral cavity, pharynx, larynx, and auricles were observed. The next tissue sample was obtained for histopathological examination. After study of HE slides, the diagnosis was inconclusive, and additional procedures were implemented. In immunohistochemical studies, the tumour cells demonstrated positive immunostaining for vimentin and S-100. p53 was positive in 20% and Ki-67 in 5% of cells. Taking into consideration the microscopic pictures and low Ki-67 expression, the diagnosis of chondroma (Fig. 2) was suggested. Due to lack of correlation between clinical and microscopic examinations, more tissue samples were obtained. After multidisciplinary discussion among laryngologists and pathologists with analysis of clinical data, new microscopic slide studies, and radiological images the final diagnosis of chondrosarcoma (Fig. 3) T4aN0M0, stage IVA was made.

The patient underwent left subtotal maxillectomy via Weber-Ferguson incision, bilateral ethmoidectomy, sphenoidectomy and right upper turbinectomy, excision of nasal septum, left frontal sinus, and left exenteration of left orbit. The examination of the whole surgical specimen confirmed the aforementioned diagnosis. The

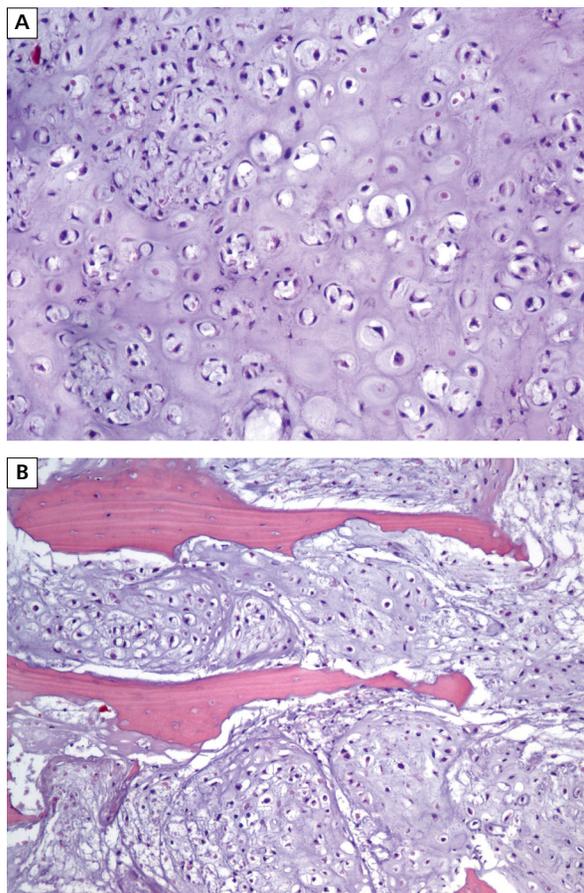


Figure 3. Microscopic image of chondrosarcoma. Primary objective magnification 10 ×. **A** — chondrosarcomas composed of atypical neoplastic chondrocytes with loss of normal architecture and distribution; **B** — invasion of the bone

patient's postoperative period was uneventful and the patient was discharged from the hospital nine days after the operation.

Discussion

Patients with chondrosarcoma of the jaws are reportedly divided equally between males and females. Maxillary tumours are difficult to eradicate and therefore are less amenable to cure. Local recurrence leads to death by direct extension of the tumour into vital structures of the head. These tumours arise from remnants of embryonic cartilage precursors from nasal and septal development in the anterior part of the maxilla and from Meckel cartilage precursors in the posterior aspect of the mandible [1]. The most common sign of chondrosarcoma of the maxilla is painless swelling. Other symptoms are paraesthesia, trismus, and loosening of the teeth [15].

The diagnosis of chondrosarcoma is among the most difficult ones in tumour pathology. This bone tumour

is usually discovered on radiographs. In some cases it can be difficult to differentiate between benign and malignant lesions. Additional techniques such as CT scan and MRI can deliver more information about the nature of the tumour mass. In the majority of cases, biopsy of a tissue sample and histopathological examination brings the final diagnosis of chondrosarcoma.

In our case, there were difficulties in recognising chondrosarcoma in the primarily taken small samples of the tumour with lack of full clinical data sent to the pathologist. In some cases there are problems in distinguishing chondroma from low-grade chondrosarcoma. Chondroma is a tumour composed of mature hyaline cartilage with the highest prevalence in the fifth to seventh decade of life. It usually occurs in the mid-line of the axial skeleton and is generally asymptomatic. It can be called enchondroma when it occurs within the medullary cavity of tubular bones. The cells resemble normal chondrocytes and produce cartilaginous matrix. The histological appearance of enchondroma could be similar to grade-1 chondrosarcoma. In both cases calcification and ossification are common. However, in higher grades, chondrosarcoma displays significant hypercellularity and atypia. Cells of chondrosarcoma are often binucleate with clumps of chromatin within the nucleus. Additionally, islands of cartilage remain separated from the trabecular bone in chondroma, unlike in chondrosarcoma. In the latter, bone is usually infiltrated [17]. Chondrosarcoma is graded from 1 to 3. Grade 1 chondrosarcoma grows relatively slowly and the cells resemble unaffected ones. In grade 2, dysplasia is more prominent. The amount of chondroid matrix is significantly reduced. In grade 3, pleomorphic cells show the highest mitotic activity [1, 8, 11]. There is a high tendency towards both local and distant recurrence [18].

Mesenchymal chondrosarcoma (MC) and chondroblastic osteosarcoma should be included in the differential diagnosis. MC accounts for up to 3–9% of all chondrosarcomas. It usually affects the head and neck region [19]. It is characterised by a biphasic pattern of neoplastic cartilage with associated neoplastic small round cell component and calcification within the chondroid matrix [20]. In our case, the small round cell component was not found. Moreover, MC occurs most often in the second to third decade of life, which is earlier than in our case. Chondroblastic osteosarcoma is one of three subtypes of conventional osteosarcoma, besides osteoblastic and fibroblastic. The chondroblastic type occurs predominantly in the head and neck region. Histologically, the tumour shows atypical chondroid areas composed of pleomorphic cells, large hyperchromatic nuclei, and noticeable nucleoli. The characteristic feature of chondroblastic osteosarcoma is ossification, although this was absent in our case [21].

Our case shows that radiography and histopathology along with detailed clinical data are necessary to make

the final diagnosis. The presented case revealed that chondrosarcoma can be a heterogeneous tumour. Collecting tissue samples from different locations should be an important part of the tumour examination. On the other hand, a pathologist having diagnostic doubts should be able to communicate with a clinician to discuss borderline and complicated cases.

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