CASE REPORT

Secondary chondrosarcoma in the mandibular condyle

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We report the case of a woman who had secondary chondrosarcoma in an osteochondroma in the left mandibular condyle, presenting as a large lobulated bone density mass in the left condyle process, to illustrate the clinical characteristics and imaging findings.

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Introduction

Chondrosarcoma is the third most common primary malignant neoplasm of bone. Two-thirds arise *de novo* and are referred to as primary chondrosarcomas.^{1,2} Conversely, chondrosarcomas superimposed on pre-existing benign cartilaginous neoplasms, such as enchondroma or osteochondroma, are referred to as secondary chondrosarcomas. The most common skeletal site is the bones of the pelvis and shoulder girdle; the mandible is a rather infrequent site of occurrence. In this report, we describe an unusual case of a secondary chondrosarcoma arising from mandibular condyle osteochondroma in a 34-year-old Chinese woman presenting with a large lobulated bone density mass in the pterygomaxillary space.

Case report

Clinical history and physical examination findings

A 34-year-old Chinese woman was referred to our hospital in February 2008 for maxillofacial consultation. In March 2003, we curetted the enlarged left mandibular condyle process and a pathological diagnosis of osteochondroma was made. 5 years later, the patient was re-admitted with progressive limitation of mouth opening and facial asymmetry. The patient was generally healthy but an extraoral examination revealed facial

asymmetry with a hard, non-tender swelling of the left face. The overlying skin was intact with no anaesthesia and maximum interincisal mouth opening of 3 mm. She had no history of radiotherapy and was taking no medications. The multiple osteochondromas and family history were not found.

Radiographic findings

Axial CT scans (Figure 1) using a LightSpeed-16 (GE Medical Systems, Milwaukee, WI) were taken with 5 mm contiguous slices with a pitch of 1 mm, a 1 s scan time and reconstruction using the bone algorithm, along with MR images (Figure 2) (1.5 T MRI units, Signa Excite, GE Healthcare, Milwaukee, WI) which had a spin-echo T_1 -weighted sequence (repetition time (TR) 660 ms, echo time (TE) 9.9 ms) and T_2 -weighted sequence (TR 4200 ms, TE 94.6 ms and fat suppression at an effective section thickness of 5 mm). They showed an $8 \times 6 \times 5$ cm mass underlying the left condyle process and mandibular ramus and occupying the left pterygomaxillary space. The various degrees of maturity and calcification in the areas of enchondral mineralization caused a heterogeneous appearance on CT and MR images. Areas of calcified cartilage showed high attenuation on CT scans, but were very heterogeneous on T_2 -weighted MR images (low-signal intensity with all pulse sequences in densely calcified areas vs higher signal intensity in areas with more prominent components of non-mineralized cartilage). Non-mineralized hyaline cartilage cap was thin (1 cm) and had a more fluid characteristic at CT and MR imaging, reflecting its high water content. It was more difficult to detect on the soft-tissue window CT scan than with MR imaging.

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Figure 1 (a) Axial CT scan (soft-tissue window) and (b) multiplanar reformatted coronal CT image show a large lobulated bone density mass in the left pterygomaxillary space. The mass arises from the left mandible. It has an irregularly calcified surface and the organized centre is denser (black asterisks) than the periphery (white asterisks). The thin areas of soft tissue show lower attenuation than muscle around the bone density mass (black arrows). The cranial base was invaded (white arrow)

The mass had extrinsic erosion of the medial aspect of the left mandibular condyle and ramus. The carotid vasculature was posteriorly displaced by the mass. The left pterygoid muscles and lateral pharyngeal wall were displaced and the cranial skeletal base was remodelled.

Histological findings

The patient had not received pre-surgical radiotherapy. Surgery was performed under general anaesthesia. Wide

local resection of the chondrosarcoma included removal of condyle process and mandibular ramus. There were few residues near the foramen ovale. Multiple sections were taken from the surgical specimen, which was grey-violet and hard-elastic. Microscopically, the tumour was cartilaginous, and the tumour cells resembled normal chondrocytes and lay in lacunar spaces embedded within hyaline cartilage matrix (Figure 3). The individual lobule of tumour was seen permeating the marrow spaces and engulfing cancellous bone

Figure 2 (a) Axial T_1 weighted (600/9.9) and (b) coronal T_2 weighted (4240/80.9) images show lobulated heterogeneous signal intensity mass with low signal intensity (white asterisk) with all pulse sequences, corresponding to densely calcified areas, and multiple hyperintense foci (black asterisks), corresponding to hyaline cartilage. A thin band of high signal intensity can be seen at the lesion periphery (white arrows) that could represent a non-mineralized portion of the cartilage cap. The carotid vasculature was posteriorly displaced by the mass

Figure 3 Microscopic image with haematoxylin of malignant cartilage cells in lacunae and surrounding hyaline matrix (short black arrows), indicating Grade 1 chondrosarcoma, that is permeating marrow spaces and engulfing cancellous bone trabeculae (long thin black arrows) ($\times 100$)

trabeculae. Low cellularity, mild nuclear atypia containing hyperchromatic plump nuclei of uniform size were present and the diagnosis of Grade 1 chondrosarcoma was made.

Discussion

Chondrosarcoma is a malignant tumour with pure hyaline cartilage differentiation.³ Secondary chondrosarcoma is a chondrosarcoma arising in a benign precursor, either an osteochondroma or enchondroma.⁴ The risk for chondrosarcoma in solitary osteochondroma has been reported to be 2–7.6%⁵ and that for osteochondromatosis 5–36.3%.⁵ Patients with Ollier disease and Maffucci syndrome have a 25–30% risk of developing chondrosarcoma.^{6–8}

Primary chondrosarcoma is a tumour of adulthood and old age. The peak incidence is in the fifth to seventh decades of life, whereas secondary chondrosarcoma occurs in a younger age group. According to Ahmed *et al*, the mean age at the time of diagnosis was 34.9 years (range, 15–77 years).⁵ Any portion of the skeleton may be involved. However, the pelvic and shoulder girdle bones are more frequently affected. A PubMed search showed that 21 temporomandibular joint (TMJ) chondrosarcoma cases had been reported up to 2009.⁹

In a review of the literature,^{10–12} the lesion site, which is lateral or medial to the mandibular condyle, was important for an early *vs* late diagnosis. Lesion sites may also cause progressive occlusal changes¹³ and deviation and progressive impairment in mandibular motion are always found.

A change in clinical symptoms in a patient with a known osteochondroma or enchondroma precursor heralds the development of chondrosarcoma. Sudden pain and/or an increase in swelling are frequent complaints.

In the surgical case presented here, the final diagnosis was of low-grade chondrosarcoma and, because of its pathological diagnosis of osteochondroma made 5 years before, the lesion was clearly defined as secondary chondrosarcoma. To the best of our knowledge, secondary chondrosarcoma arising in an osteochondroma involving condyle process has never been reported.

Mandibular chondrosarcomas involving the condyle process may be confused with osteochondroma. Osteochondroma is a more common tumour in mandibular condyle process than chondrosarcoma. The major clinical distinctions between osteochondroma and chondrosarcomas of the mandibular condyle process are patient age and rate of growth. Osteochondroma tends to occur in younger patients before the age of 20 years^{14–16} and grows much more slowly than secondary chondrosarcomas. Mirra *et al* reported that patients with secondary chondrosarcomas were, in general, 12 years older than those with pure benign solitary enchondromas.¹⁷ CT and MR images can be valuable in the pre-operative differentiation of osteochondroma and chondrosarcoma. CT and MR images of osteochondroma show a bony mass with a sharply defined periphery, a more lucent but organized centre with cortex and medullary cavity continuous with the bone from which it arises and a thin cartilage cap. Chondrosarcoma shows a soft-tissue mass with an inhomogeneously mineralized centre. There may be destruction of adjacent bone or abnormalities of adjacent soft tissues. MR imaging is the best radiological modality for evaluating the hyaline cartilage cap. Mineralized areas in the cartilage cap remain of low-signal intensity with all MR pulse sequences. The high water content in non-mineralized portions of the cartilage cap have intermediate to low-signal intensity on T_1 -weighted images and very high-signal intensity on T_2 -weighted MR images.^{18,19} Hyaline cartilage cap thickness is an extremely important criterion in determining

osteochondroma malignant transformation. Two series have reported the cartilage cap thickness differences in benign osteochondromas (0.1–3.0 cm; average, 0.6–0.8 cm) and those with secondary chondrosarcoma (1.5–12 cm; average, 5.5–6.0 cm).^{20,21}

At pathological analysis, chondrosarcoma secondary to osteochondroma shows a thick lobulated cartilage cap. Secondary chondrosarcomas are generally low-grade tumours with few distant metastases.

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