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Clear-cell chondrosarcoma of the maxilla

Report of a case

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Clear-cell chondrosarcoma is a variant of chondrosarcoma which is characterized by a typical histomorphology and a very slow rate of growth. A case is presented in which the tumor was located in the maxilla.

Clear-cell chondrosarcoma is a recently recognized type of chondrosarcoma which is characterized by the presence of benign giant cells, clear cells, and areas of pleomorphic cells in a cartilaginous matrix.[1, 2] In the past, tumors with a similar histomorphology have been reported as malignant chondroblastoma,[3] chondroblastic sarcoma,[4, 5] and atypical chondroblastic tumor of low-grade malignancy.[6]

It is important to distinguish this tumor from conventional chondrosarcoma because clear-cell chondrosarcoma has a slow-growing nature and patients can be cured by radical local excision.[1, 2]

We present a case of clear-cell chondrosarcoma originating in the maxilla and discuss various aspects of this tumor.

CASE REPORT

In February, 1970, a 50-year-old white woman entered the Department of Maxillofacial Surgery of the Utrecht University Hospital for treatment of a swelling in the left maxilla. The swelling had been present for 3 years and had enlarged slowly during that time.

Inspection of the oral cavity revealed a firm, elastic, painless swelling of the maxilla in the region of the upper left canine. The tumor was bulging into the buccal area. The overlying mucosa was not adherent to the tumor, and there was no ulceration. Radiographs showed a multilocular radiolucency in the left maxillary alveolar process, lying between the roots of the lateral incisor and canine and also partly surrounding them (Fig. 1). The periodontal ligament space of these teeth was not visible in some places but root resorption could not be detected. Both teeth were vital, as could be established by testing with faradic current.

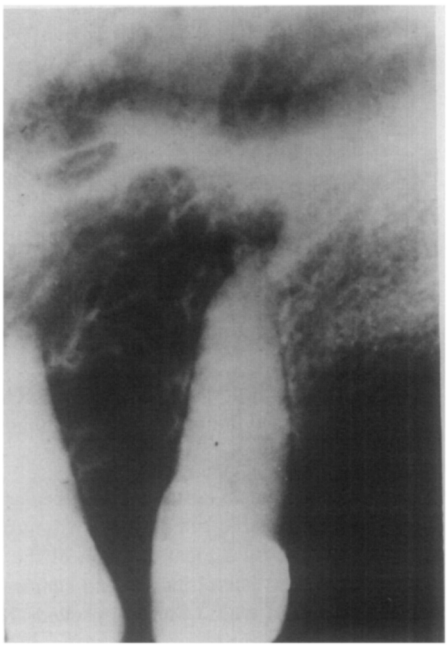

Fig. 1. Radiograph shows multilocular radiolucency in left maxillary alveolar process.

It was decided to remove the lesion by an excisional biopsy. At operation, after removal of the mucoperiosteum, a

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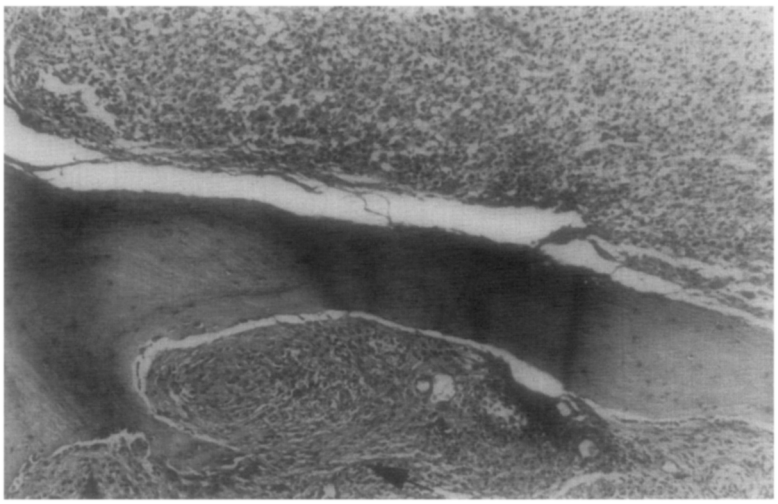

Fig. 2. Photomicrograph demonstrates large compact fields of monomorphic clear cells infiltrating between cancellous bone. In some places, the tumor cells show a spindle-cell morphology *(arrow)*. (Hematoxylin and eosin stain. Magnification, ×40.)

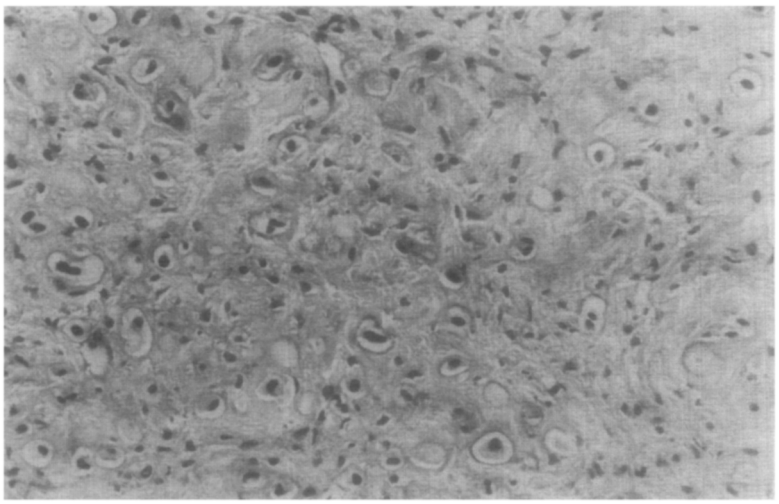

Fig. 3. Photomicrograph demonstrates cells lying in a chondroid matrix and showing hyperchromatism, pleomorphism, and double nuclei in lacunae. (Hematoxylin and eosin stain. Magnification, ×100.)

defect was noted in the buccal cortex of the alveolar process through which a gray, friable tumor mass was protruding. The tumor was removed by a local excision of the alveolar process, including the left lateral incisor and canine, and the bony defect was enlarged with a bur and packed. The postoperative course was uneventful.

Histopathologic examination of the specimen showed a lesion composed of clear monomorphic cells with a small central nucleus and distinct cytoplasmic boundaries. These cells were infiltrating into the marrow spaces of the preexisting cancellous bone (Fig. 2). Some of the clear cells had a faintly acidophilic cytoplasm and transitions could be traced between the clear cells and areas in which there were cells lying in a chondroid matrix and showing hyperchromatism, pleomorphism, and multiple nuclei in lacunae (Figs. 3 and 4). There were also transitions between the clear cells and fields of spindle-shaped mesenchymal cells (Fig. 5). Multinucleated giant cells were scattered among the spindle cells and surrounded bony spicules in this area. Within the compact fields of clear cells, multinucleated giant cells were not present. In some places, the chondroid matrix contained calcified material, partly as fine sheets between the tumor cells and partly as amorphous clumps in areas in which the chondroid matrix was more prominent.

The greater part of the tumor was composed of monomorphic clear cells with a uniform nucleus in which there were areas of pleomorphic cartilaginous cells and also areas of spindle-shaped mesenchymal cells. Because of this histomor-

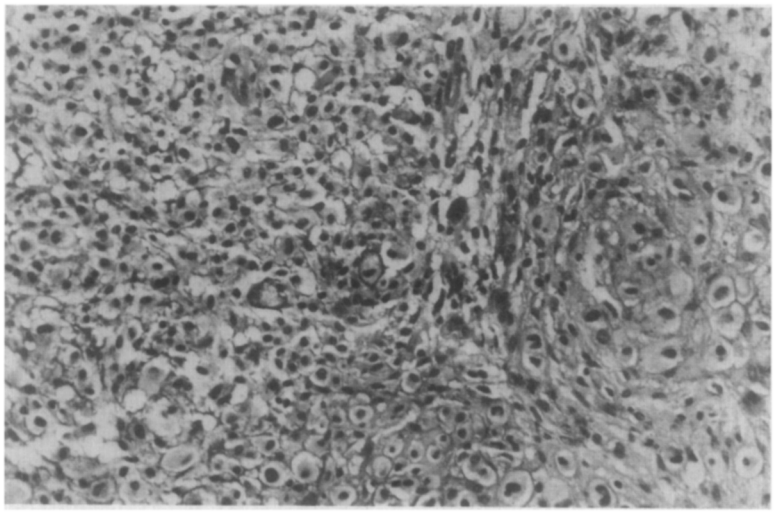

Fig. 4. Photomicrograph demonstrates transition between monomorphic clear cells (left side) and pleomorphic cartilaginous cells (right side). (Hematoxylin and eosin stain. Magnification, ×100.)

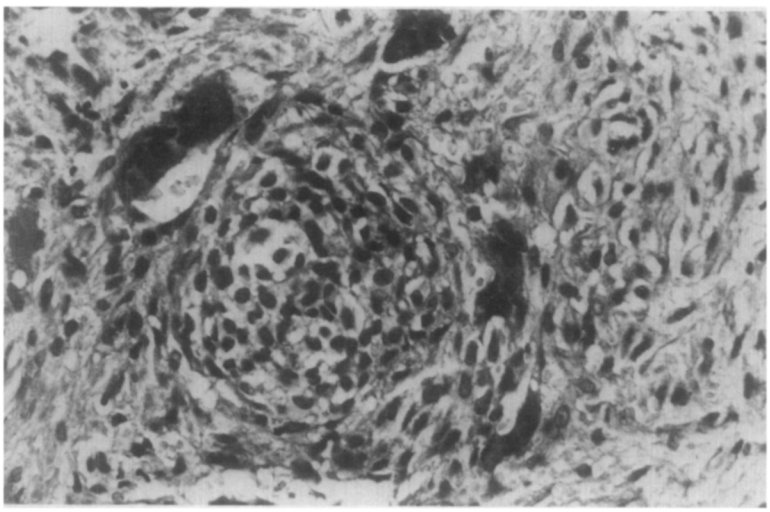

Fig. 5. Photomicrograph demonstrates transition between nest of clear cells and surrounding spindle-shaped mesenchymal cells. A few multinucleated giant cells are also present in this area. (Hematoxylin and eosin stain. Magnification, ×150.)

phology, we consulted several pathologists. Some of them suggested that the lesion represented an atypical chondroma; others concluded that the lesion was a chondrosarcoma or even an osteosarcoma. A final diagnosis was not reached at that time and therefore it was decided to refrain from further therapy and to provide the patient with very careful follow-up. Since the initial treatment in February, 1970, the patient has remained entirely free from recurrence or metastasis.

The publication of the papers of Unni and his colleagues[1] and Le Charpentier and his colleagues[2] prompted us to review the slides of this patient and to consult Prof. Le Charpentier. He confirmed our presumption that this lesion was another example of the recently recognized clear-cell chondrosarcoma. After 10 years, a final histopathologic diagnosis was reached.

DISCUSSION

Chondrosarcomas rarely arise in the jaws. Next to reports of single cases, only small series[7–12] or compilations[13, 14] have been published. In the past there has been much discussion about the presence of chondroid rests in the maxilla from which a cartilaginous tumor could arise,[15, 16] but today it is realized that cartilaginous differentiation can occur in tissues which normally do not show production of chondroid matrix.[17, 18]

Clear-cell chondrosarcoma has not yet been de-

scribed in the maxillofacial skeleton to the best of our knowledge. It is a tumor which is characterized by the presence of closely packed monomorphic clear cells with a centrally placed uniform nucleus, areas of pleomorphic cells lying in a chondroid matrix, and multinucleated giant cells showing the features of osteoclasts. The clear cells have a distinct cytoplasmic boundary and sometimes their cytoplasm shows a faint eosinophilia. They have been shown to be of chondroid nature by electron microscopy.[2] Dispersed between the tumor cells, fine lines of amorphous calcifications are seen and short seams of osteoid and bone are present in some instances.[1, 2]

When the histopathologic data of our patient are compared with the above-mentioned characteristics, various similar aspects are noted. Clear cells, multinucleated giant cells, and pleomorphic cells lying in a chondroid matrix were present. Seams of osteoid and bone were not seen, but these structures are considered to be a result of stromal reaction and not to be of neoplastic nature.[2] In addition to the already mentioned fine lines of calcified material, [1] we also noted the occurrence of amorphous clumps of calcified material in those areas where there was abundant chondroid matrix. In our opinion, the presence of calcified material in the chondroid matrix should not be considered as a result of dystrophic calcification, but as a phenomenon that is normally occurring in cartilage that is prone to undergo endochondral ossification. The multinucleated giant cells in the tumor are also considered to be of a reactive nature.[1, 2] We observed them only in relation to bony spicules or extracellular material between the tumor cells in those areas where these cells showed a spindle-cell morphology. This conforms to the opinion of Chambers[19] that in tumors producing collagen and extracellular ground substance, giant cells can form following the recognition of abnormal extracellular material produced by the neoplastic cells.

Unni and his colleagues[1] discussed the relation between clear-cell chondrosarcoma and chondroblastoma—a relation that could exist because of the similarity in location and some identical histologic features. It is considered that clear-cell chondrosarcoma could be a chondroblastoma that had undergone a malignant change. Our case, however, does not support this hypothesis. Chondroblastomas have not been reported in the maxilla or nasal skeleton. Only cases in the mandibular condyle have been seen.[20, 21]

The radiographic appearance of clear-cell chondrosarcoma in the skeleton is that of an osteolytic expansive lesion. Bone sclerosis can sometimes be noted at the margins, but in larger tumors the margins become poorly defined.[1, 2] The radiographs of our patient showed a multilocular, poorly defined, osteolytic lesion, an appearance which is consistent with malignant mesenchymal tumors located in the jaw bones.[14] We could not detect a localized thickening of the periodontal ligament space which is considered to be typical for chondrosarcoma[22] and also for osteosarcoma.[23] The lamina dura of the tooth socket had been destroyed by the tumor without producing the effect of local widening of the periodontal ligament space.

The clinical course of clear-cell chondrosarcoma is rather favorable. The tumor is slowly enlarging with a low metastatic potential and symptoms of tumor are generally of long standing before patients seek treatment.[1, 2] Treatment by en bloc resection is advised.[1, 2] Our patient's case illustrates the low-grade malignancy of this particular form of chondrosarcoma. Despite the patient's delay of 3 years and a rather limited treatment, no recurrence or metastasis has been observed up to 10 years after the initial therapy. As Evans and colleagues[9] have shown that most recurrences become evident within 5 years, the longest interval being 9 years, 10 years should be sufficient to assume that the patient has been cured of her tumor.

Sincere thanks are due Prof. Y. Le Charpentier for his advice in the histologic evaluation of the reported case.

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