Clear-Cell Chondrosarcoma

A Light- and Electron-Microscopic and Histochemical Study of Two Cases

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Summary. Two cases of clear-cell chondrosarcoma located in the upper end of the right femur of men aged 30 and 40 years are reported. The roentgenologic appearances suggested a chondroblastoma. Both patients are alive, one and four years after surgical removal of the tumor.

Glucosaminoglycans were studied with cationic dyes at different pH, with and without pretreatment with testicular hyaluronidase, and with the Scott technique at the light-microscopic level. Ultrastructurally, the glucosaminoglycans were studied with the high iron diamine and dialyzed iron techniques and glycogen with the PATH-method. Light-microscopically, the tumors were characterized by clear vacuolated cells with distinct cytoplasm boundaries and scattered multinucleated giant cells of osteoclast type. Histochemical studies at the light-microscopic level indicate the presence of chondroitin 4- and 6-sulphate but no keratosulphate. Ultrastructurally, the predominant clear-cells showed features characteristic for chondroblasts. The cytoplasm showed areas lacking organelles and containing a low-density, finely granular matrix. These areas are considered to correspond to the clear cytoplasmic vacuoles seen under the light microscope. Most of the organelles were seen in the perinuclear region. The irregular tumor cells formed delicate protruding cytoplasmic extensions, which delineated intercellular spaces appearing as vacuoles under the light microscope. The benign multinucleated giant cells had an ultrastructural appearance typical of osteoclasts. Histochemical analysis at the electron-microscopic level showed the presence of sulphated glucosaminoglycans in the intercellular matrix and in association with the cytoplasmic membrane. Glycogen and non-sulphated acid glucosaminoglycans were found within the cytoplasm of the clear-cells.

Key words: Chondrosarcoma – Clear-cell chondrosarcoma – Glucosaminoglycans – Histochemistry – Ultrastructure.

Introduction

Sixteen cases of a rare cartilaginous tumor of low grade malignancy, called clear-cell chondrosarcoma, which originally had been considered benign, were
recently described from the Mayo Clinic (Unni et al. 1976). In several instances the chondroid matrix and foci of bone formation had led to an erroneous initial diagnosis of atypical chondroblastoma or osteoblastoma. Roentgenographically, the lesion was usually well defined and indistinguishable from chondroblastoma. However, the clear-cell chondrosarcomas occur in an older age-group than chondroblastomas. The designation clear-cell chondrosarcoma refers to the peculiar histological appearance, that is clear, vacuolated tumor cells with distinct cytoplasmic boundaries. A constant finding in these tumors was scattered, small, osteoclast-like giant cells. Very recently, Le Charpentier et al. (1979) reported five cases of clear-cell chondrosarcoma (one of which was included in the Mayo Clinic series) and briefly described the electron-microscopic appearance in three of them.

This paper describes two cases of clear-cell chondrosarcoma of the upper end of the femur, which were studied histochemically with respect to glucosaminoglycans and carbohydrates: One case was studied electron-microscopically and the histochemical analysis was in this case extended to the ultrastructural level.

Case Reports

Case 1. A thirty-year-old man, who suffered from poliomyelitis as a child, from which he recovered without any sequelae, consulted in April 1979 for pain in his right hip. The pain started one and a half years earlier after trauma to the right hip and gradually increased, so that he limped and had to use a cane. Finally the pain became so severe that he was unable to walk at all. At examination the mobility of the hip joint was normal. Plain radiograms disclosed a well delineated osteolytic area with partly sclerosed distinct borders within the head and neck of the right femur (Fig. 1). Angiography showed some delicate vessels within the osteolytic area and diffuse opacification. There were no signs of arteriovenous shunting.

In June 1979 the area was explored and the lesion curetted. The defect was filled with transplanted bone from the iliac bone. Since then no further surgery has been performed. The patient has been followed with repeated roentgenographic examinations and in September 1979 there is no sign of recurrence, and the transplanted bone has homogenized.

Case 2. A forty-year-old man, who for many years was active as a speedway driver, consulted in 1976 because of severe pain in his left hip. During the time he was active as a speedway driver he had sustained numerous traumata, including a tibial fracture and a hepatic rupture. He had also been operated upon for duodenal ulcer and for cholelithiasis. Before he sought medical care the pain had gradually increased over a period of several months, and he finally limped severely and had constant pain when walking or standing. Plain roentgenograms showed a large osteolytic area within the head and neck of the left femur (Fig. 2). This was well delineated, with partly sclerosed margins and a few delicate bony septa inside the lesion. The roentgenographic appearance was interpreted as a benign bone cyst. After one year, in March 1977, the osteolytic area was explored and the lesion was curetted. The histopathologic anatomic diagnosis was chondrosarcoma and the patient was referred to the Department of Orthopedic Surgery, Sahlgren's Hospital, Göteborg, for radical surgery. In May 1977, a left hemipelvectomy was performed. Since then the patient has been seen at regular intervals, without any signs of recurrence or metastases. At examination in April 1979 the patient is well and walks with a prosthesis without using a cane.

Pathology

Histological, Histochemical and Electron-Microscopic Methods

The surgical specimens were fixed in 4% formaldehyde solution and embedded in paraffin. Five-micron sections were stained according to the van Gieson method and with hematoxylin