A CASE OF CHONDROMYXOID FIBROMA SHOWING EXTENSIVE CHONDROBLASTOMATOUS DIFFERENTIATION*

Yaygın Kondroblastomatöz Differansiyasyon Gösteren Kondromikzoid Fibroma Olgusu

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Abstract

Chondromyxoid fibroma is an unusual bone tumor of cartiliginous origin. This neoplasm constitutes approximately 1-2% of all bone tumors. Chondromyxoid fibroma affects long tubular bones especially of the lower limbs. In long bones, the lesion typically involves the metaphysis. Chondroblastoma is important in the spectrum of differential diagnosis of chondromyxoid fibroma. Although chondroblastomas share some histological features with chondromyxoid fibroma, the histological similarity is usually limited.

A radiolucent, well-demarcated tumoral lesion with widespread round oval cells and lobulated, grooving nuclei and multiple focal degenerated areas in upper metaphysial region of right tibia in a-36-year old woman was interpreted as chondromyxoid fibroma because of the lobular grooving pattern and metaphysial location of the tumor. In the present study, the histopathological similarities between chondromyxoid fibroma and chondroblastoma, and the difficulties in differential diagnosis are discussed.

Key Words: *Condromyxoid fibroma, chondroblastomatous differentiation.*

Özet

Kondromikzoid fibroma kıkırdak doku kökenli nadir bir kemik tümörüdür. Tüm kemik tümörlerinin yaklaşık %1-2'sini meydana getirir. Özellikle alt ekstremitelerin uzun kemiklerinde görülür. Uzun kemiklerde tipik olarak metafiz lokalizasyonludur. Kondromikzoid fibromun ayırıcı tanı spektrumunda kondroblastoma önemlidir. Kondroblastomalar kondromikzoid fibroma ile bazı histolojik özellikleri paylaşmalarına rağmen histolojik benzerlik genellikle sınırlıdır.

36 yaşında kadın hastanın sağ tibia metafizinde yerleşimli radiolusen, iyi sınırlı, kahve çekirdeği görünümlü nükleuslara sahip, yuvarlak-oval biçimli hücrelerin yaygın olarak izlendiği, multifokal dejenere alanlara sahip tümöral lezyon, metafiziyel lokalizasyon ve lobüler gelişim paterni nedeni ile kondromikzoid fibroma olarak yorumlandı. Bu çalışmada, kondromikzoid fibroma ve kondroblastoma arasındaki histolojik benzerlikleri ve ayırıcı tanıdaki güçlükleri tartışıyoruz.

Anahtar Kelimeler: *Kondromikzoid fibroma; kondroblastomatöz differansiyasyon.*

Introduction

Chondromyxoid fibroma and chondroblastoma are benign neoplasms of bones originating from chondroid tissue. Both the neoplasms appear about the same age. Chondroblastoma typically shows epiphysis location. Although the two tumors have different histopathological characteristics in prototype cases, intermediate forms may cause difficulties diagnosis.

In this study, we present a case of chondromyxoid fibroma localized in the upper metaphysial region of right tibia in a 36-year-old woman. Soft tissue component of the tumor was also present and histopathologically extensive chondroblastomatous differentiation areas were observed in the sections.

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Case Report

A 36-year-old woman was referred to the Department of Orthopedics and Traumatology due to pain and swelling of the right knee. Radiographic analyses showed a radiolucent, well-demarcated cystic lesion with sclerotic and scalloped margins in the upper metaphysis of the right tibia. Soft tissue component accompanying bone lesion was also observed in radiography. The bone lesion with soft tissue component was resected and curetted surgically.

In the histopathological examination, a lobulated tumoral lesion extensively destructing cortical bone was observed (Figure 1). The tumor was predominantly chondroid with less myxoid matrix. Tumor cells with vesicular nuclei showed nuclear grooves were present in the matrix (Figure 2). Stellate shaped cells were seen in some areas. A few osteoclast-like giant cells were observed in the periphery of the tumor. The differentiation of hypercellular and hypocellular areas could not be clearly defined. While some focal areas were hypercellular, the border of hypercellular and hypocellular areas were not clear. Soft tissue infiltration area by the tumor was only observed in an area (Figure 3). In this infiltration area, there was an increase in the cellularity according to other tumoral areas. In addition, extensive degeneration areas were seen in the tumor. Atypia, necrosis and mitosis were not observed in any area of the tumor.

The case was diagnosed as chondromyxoid fibroma, due to the radiologic characteristics and histopathological features of the tumor.

Discussion

Chondromyxoid fibroma, that may be misdiagnosed as sarcoma because of alterations in its histomorphology, is a very rare bone tumor originating from chondroid tissue (1-5). Some reports state that it accounts for less than 1% of bone tumors (4,5), while other reports state this incidence as 1-2% (2). In these tumors, the presence of soft tissue infiltration areas and cytological atypia cause problems in the differential diagnosis. WHO defines chondromyxoid fibroma as “a benign bone neoplasm characterized by lobules of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material” (2,3). Local recurrences have been reported in some cases of chondromyxoid fibroma (2,3). Although textbooks state that chondromyxoid fibroma usually occurs during the second and third decades, we observed cases of chondromyxoid fibroma in every age group, according to the literature (2). However, in general, this neoplasm is encountered more frequently during childhood and adolescence (1,2). Although the upper metaphysis of the tibia is the typical involvement area, it has been reported in the literature that bones such as frontal bone and mandible are also involved in chondromyxoid fibroma (3,4). Slight male predominance for chondromyxoid fibroma is noted in the literature (2).

Diagnostic problems are not usually encountered in typical cases of chodromyxoid fibroma during the histopathological examination. However, typical lobular growth pattern characterized by stellate and spindle shaped cells in chondromyxoid matrix is not encountered in every case. Chondroblastomatous areas could be seen focally in chondromyxoid fibroma (1,2). In our case, tumor cells with histiosit- like oval shaped nuclei were extensively observed in many areas of the tumor. Typical features of chondromyxoid fibroma, hypocellular and hypercellular areas could not be clearly distinguished in the sections (1). In addition, the typical chicken wire pattern of calcification for chondroblastoma was not seen in our case. In our case, extensive degeneration areas were present and matrix of the tumor consisted of hyalen chondroid cartilage. Myxoid areas were focally observed in the tumor. Stellate and spindle shaped cells were clearly noted in the periphery of the tumor and in a limited area. Giant osteoclast-like cells were also observed at the periphery of the tumor. The histopathologic findings of our case were concordant with chondromyxoid fibroma, although the present findings also pointed to chondroblastoma. However, when the metaphysis location of the tumor was also considered, the histopathological diagnosis was reported as concordant with chondromyxoid fibroma. Another feature that caused problems in the histopathologic analysis was a limited tumoral infiltration observed in the surrounding soft tissue. Some authors suggest that a diagnosis of chondromyxoid fibroma was needed to be regarding with precaution in the case of soft tissue infiltration being observed in the tumor (1,2). Mitosis

and cytological atypia were not determined in the soft tissue infiltration area in our case.

The differential diagnosis of chondromyxoid fibroma includes myxoid chondrosarcoma, chondromyxoid fibroma-like or chondroblastic osteosarcomas, fibrous dysplasia, and chondroblastoma on histologic examination (2).

The myxoid chondrosarcomas show the lobular growth pattern, myxoid stroma, and peripheral cellularity such as chondromyxoid fibroma. However, it is known that chondrosarcomas have much more monotonous cellular composition and giant cells at the periphery of the lobules are not seen. In addition, the lobules are rather large and often show abundant myxoid ground substance. Hyaline cartilage is more likely to be seen in chondrosarcomas (2).

Chondroblastic osteosarcomas, which have very large chondromyxoid fibroma-like areas, are difficult to differentiate from chondromyxoid fibroma on a small biopsy. Mitoses and nuclear atypia seen in osteosarcomas are not characteristic features of chondromyxoid fibroma. The diagnostic hallmark of osteosarcoma is the presence of osteoid production, which is totally lacking in chondromyxoid fibroma (2).

Fibrous dysplasia may show extreme myxoid change, however, the accompanying presence of irregular osteoid seams and the radiological appearance help to make the distinction (2).

Chondroblastomas should be also consider in the differential diagnosis. However, unlike chondromyxoid fibroma, chondroblastomas are epiphyseal. The chondroblasts have eosinophilic, polygonal cytoplasm. Calcification is a conspicuous feature of most chondroblastomas. Chondromyxoid fibroma may occasionally be accompanied by some foci which have overlapping features with chondroblastoma (2). The lack of typical diagnostic features in every case, and the propensity for recurrence may suggest a malignant tumor diagnosis. Hence, any lesion labeled as chondromyxoid fibroma is best confirmed and managed in a specialist bone centre (2).

Figure 1: Lobulated growth pattern of tumor (x5, HE).

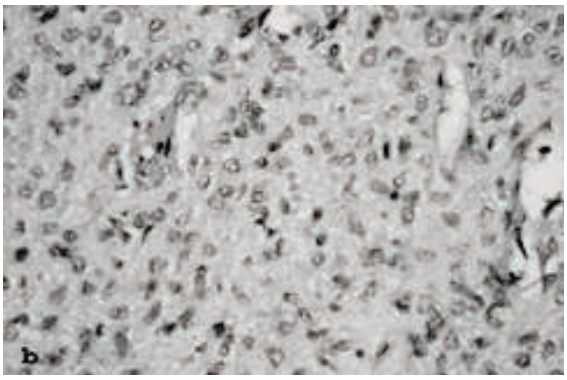

Figure 2: Tumor cells reminiscent to those of chondroblastoma (x30, HE).

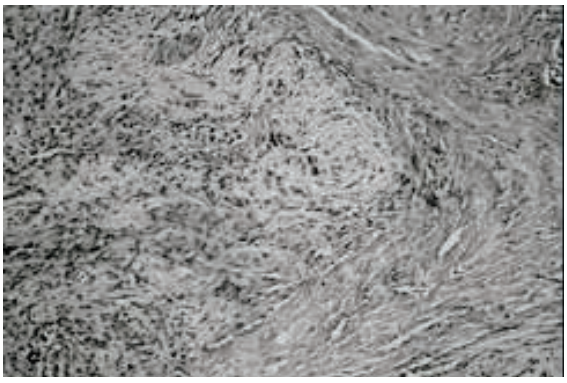

Figure 3: Soft tissue infiltration area in tumor (x10, HE).

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