Journal of Korean Medical Science
Vol. 10, No. 1, 51-56 February 1995

Chondromyxoid Fibroma of the Femur : A Case Report with Intra-cortical Location

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Chondromyxoid fibroma(CMF) is the least common benign cartilaginous tumor, comprising less than 0.5 to 1% of all skeletal neoplasms. This subject was a 16-year-old female with a three-year history of pain involving the distal femoral metaphysis. This case showed an unusual feature : it was intra-cortical in location. Radiologic differential diagnosis included metaphyseal fibrous defect, periosteal chondroma, simple or aneurysmal bone cyst, and cortical abscess. On operation, the lesion filled the intracortical defect with whitish myxoid soft tissue, bulging into the adjacent soft tissue. Microscopically, it showed typical features of chondromyxoid fibroma composed of mainly myxoid nodules and peripheral fibrous elements with focal chondroid differentiation.

Key Words : *Chondromyxoid fibroma, Intra-cortical.*

INTRODUCTION

Chondromyxoid fibroma(CMF) is the least common benign tumor of cartilage derivation (Rahimi et al., 1971 ; Kreicbergs et al., 1985 ; Schajowicz, 1987 ; Zillmer and Dorfman, 1989 ; Moser, 1990 ; Wilson et al., 1991) that was first described in 1948 by Jaffe and Lichtenstein(Jaffe and Lichtenstein, 1948). Approximately 400 cases of CMF have been reported in the literature (Rahimi et al., 1971 ; Andrew et al., 1982 ; Adler, 1985 ; Kreicbergs et al., 1985 ; Zillmer and Dorfman, 1989 ; Moser, 1990). It usually occurs at the metaphyseal region of a long bone.

We recently experienced a case of CMF involving the distal femoral metaphysis in a 16-year-old female. This case showed an unusual, intra-cortical location. These features were found in only nine cases in a review of the English-language medical literature(Schajowicz, 1987 ; Wilson et al., 1991). This unusual location prevents us from making the diagnosis of chondromyxoid fibroma. We discussed the difficulty in the radiologic differential diagnosis.

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CASE REPORT

This subject was a 16-year-old female with a three-year history of pain on her left knee. It was aggravated by hard exercise and running. There was no difficulty in walking and no history of any trauma to the limb. Physical examination revealed mild tenderness over the posterior side of the left distal femur. Knee joint movement was normal. Laboratory examination was unremarkable, except for mild elevation of erythrocyte sedimentation rate and C-reactive protein.

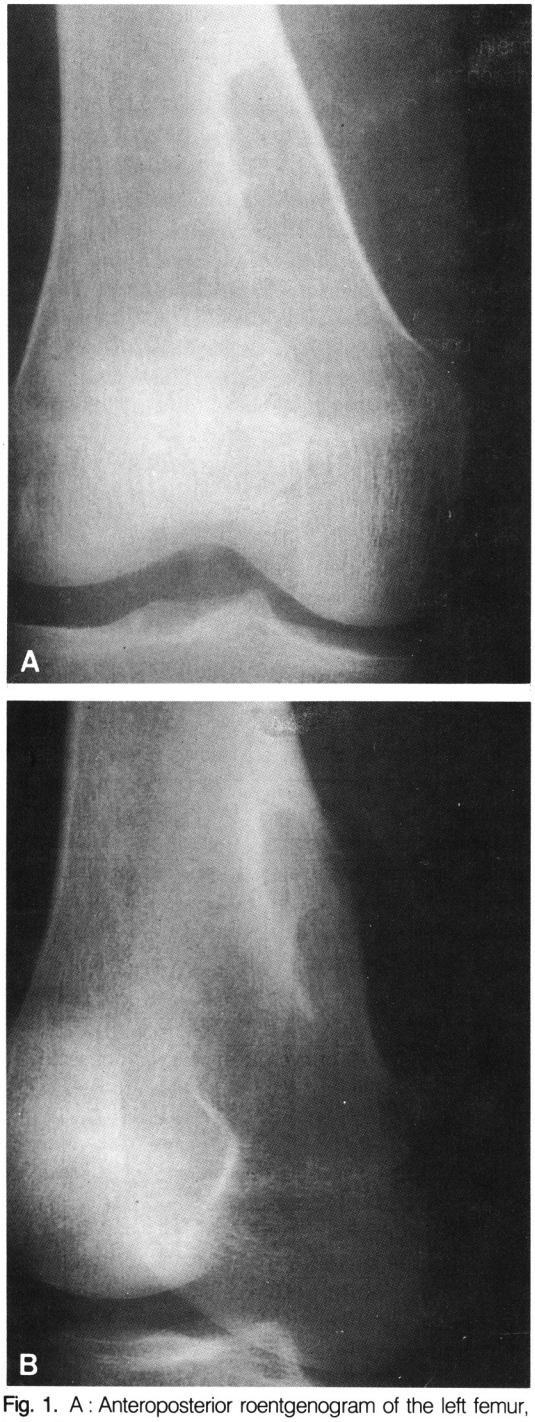

Fig. 1. A : Anteroposterior roentgenogram of the left femur, including the area of the knee, shows an elongated, radiolucent, intra-cortical lesion at the metaphyseal area of the distal femur. B : Left lateral view reveals a surrounding sclerotic rim and non-mineralized matrix.

A plain roentgenogram (Fig. 1) showed an elongated, radiolucent, and well-defined intra-cortical lesion at the metaphyseal area of the distal femur, surrounded by a sclerotic rim. There is no evidence of tumor matrix formation in this plain X-ray.

T1-weighted axial (Fig. 2A) and coronal magnetic resonance images showed a lobulated, low signal lesion with slightly lower signal foci at the posteromedial portion of the distal metadiaphyseal femur. The lesion was located within the cortex without medullary involvement. The inner wall of the cortex showed a moderately thick, sclerotic rim, and the outer wall was thin, and partially disrupted. There was no evidence of significant soft tissue extension. It measured 3×1×1.5 cm. T2-weighted magnetic resonance images (Fig. 2B and 3)

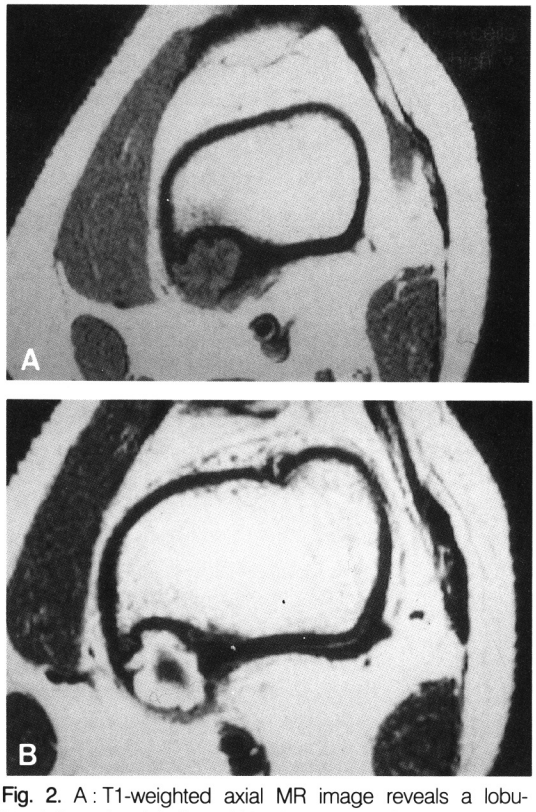

Fig. 2. A : T1-weighted axial MR image reveals a lobulated, low signal lesion within the cortex without medullary involvement. The inner wall of the cortex shows a thick, sclerotic rim and the outer wall is thin, partially disrupted. B : T2-weighted axial MR image reveals bright signal change with remaining foci of low signal.

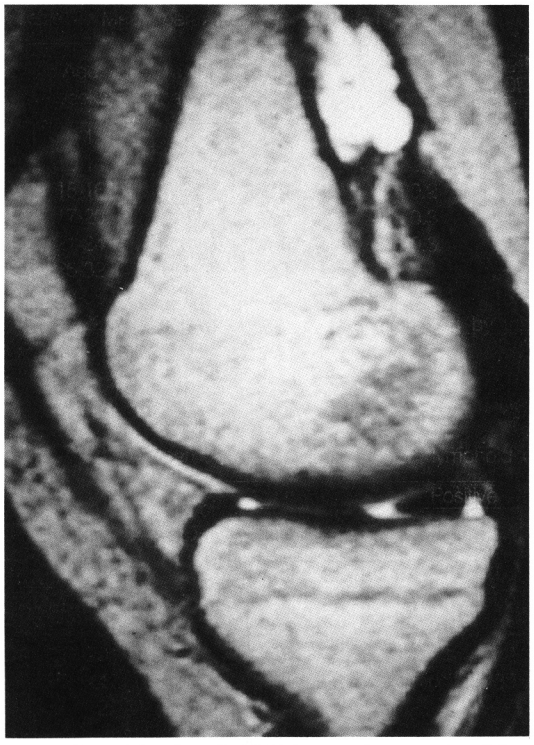

Fig. 3. T2-weighted sagittal MR image also reveals a lobulated, bright signal lesion within the sclerotic cortex.

showed bright signal change with scanty remaining foci of low signal. The radiologic differential diagnoses were metaphyseal fibrous defect, periosteal chondroma, simple or aneurysmal bone cyst, and infection.

On operation, the lesion was 1.5×4 cm in size. It filled the intracortical defect with whitish myxoid soft tissue at the posteromedial side of the distal femur. It was bulging into the adjacent soft tissue. Curettage was done.

PATHOLOGIC FINDINGS

Histological examination revealed well-demarcated nodules surrounded by periosteal fibrous tissue. Tumor nodules were composed of bluish myxoid tissue with intervening septae of proliferating collagenous fibrous tissue(Fig. 4). Tumor cells in the myxoid area were variable in size and shape such as spindle-, stellate-, and round-shaped. A few atypical, bizarre nuclei were noted. However, mitotic

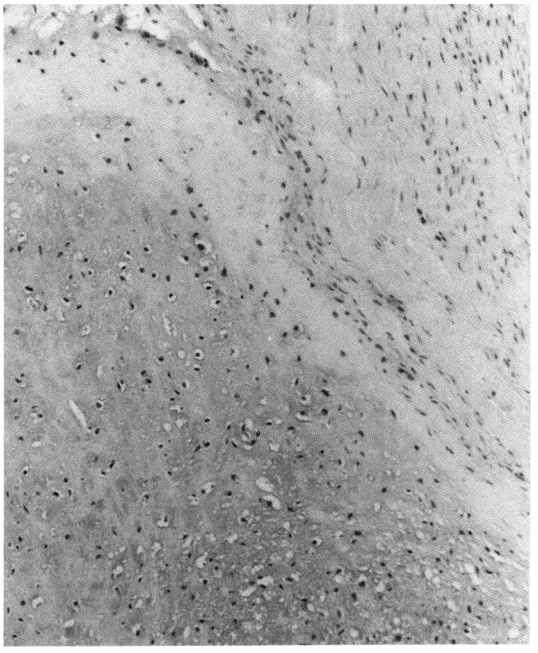

Fig. 4. A low power photomicrograph shows the classical appearance of the chondromyxoid fibroma with myxoid nodules and intervening septae of fibrous tissue(H&E, × 100).

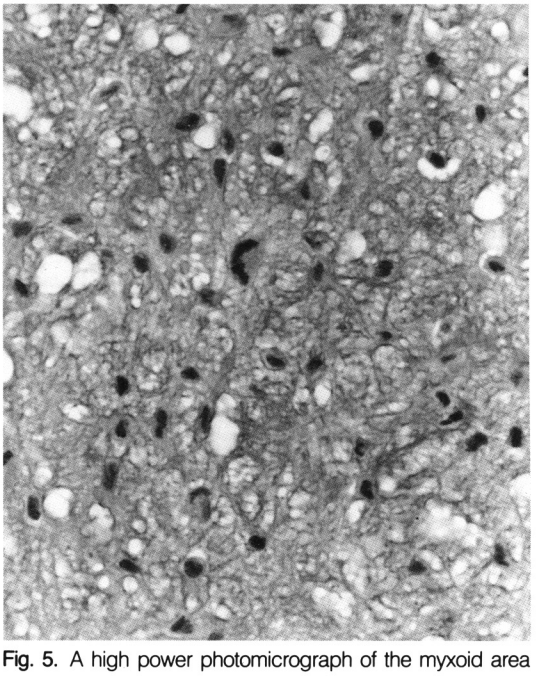

Fig. 5. A high power photomicrograph of the myxoid area shows spindle or stellate tumor cells with some bizarre nuclei(H&E, ×400).

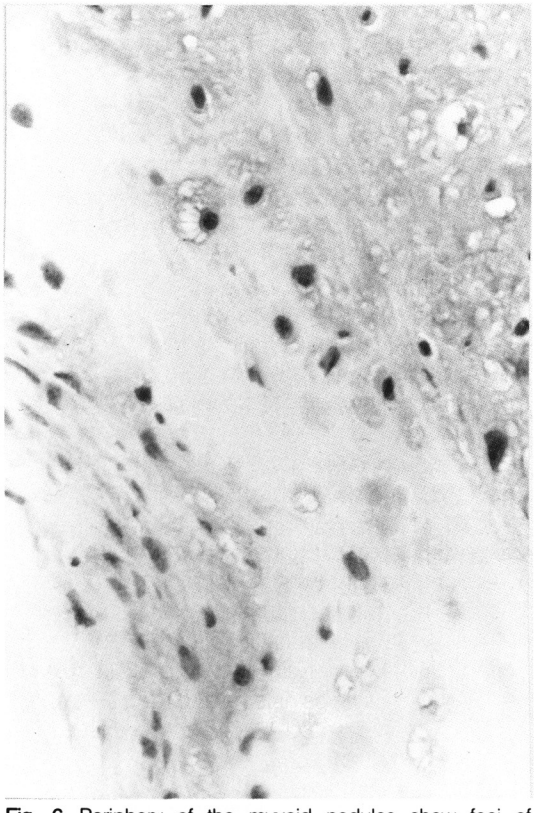

Fig. 6. Periphery of the myxoid nodules show foci of chondroid differentiation with rounded chondrocytes(H&E, ×400).

figures were not found(Fig. 5). Fibrous areas peripheral to the myxoid nodules revealed small foci of faintly bluish chondroid tissue. Rounded chondrocytes within lacunar spaces were scattered in the chondroid areas(Fig. 6). Small, amorphous foci of calcification were noted in myxoid and chondroid areas. Multinucleated giant cells of osteoclastic type were not noted.

DISCUSSION

Chondromyxoid fibroma is a distinctly uncommon tumor, and in most large series it has comprised fewer than 1% of the documented bone tumors(Rahimi et al., 1971 ; Adler, 1985 ; Kreicbergs et al., 1985 ; Schajowicz, 1987 ; Zillmer and Dorfman, 1989 ; Moser, 1990 ; Wilson et al., 1991). Despite its name, chondromyxoid fibroma is by nature a tumor of chondroid cells, rather than fibroblasts. Its similarity to chondroblastoma and the frequent involvement of both the epiphysis and the metaphysis suggests the hypothesis that chondromyxoid fibroma originates from the physeal cartilage plate(Jaffe and Lichtenstein, 1948 ; Ushigome et al., 1982).

This case showed an unusual feature : it was intra-cortical in location. The intra-cortical locations were found in only nine cases in a review of the English-language medical literature(Schajowicz, 1987 ; Wilson et al., 1991). In this location the differential diagnosis included metaphyseal fibrous defect, periosteal chondroma, simple or aneurysmal bone cyst, and cortical abscess. These unusual radiologic features of this tumor prevent us from making a diagnosis of chondromyxoid fibroma. Radiographs of this case were reviewed by John W. Beabout, Mayo Clinic, Rochester, MN. He considered this to be a purely intracortical lesion with features of chondromyxoid fibroma.

Schajowicz(1987) reported three cases of chondromyxoid fibroma with predominant cortical involvement in a review of 50 cases filed at the Latin American Registry of Bone Pathology. All three tumors were located in the upper metaphyseal region of the tibia, to be probably either intra-or subcortical in origin. They were unusually small at the time of the initial examination. The radiologic features were misleading, favoring diagnoses such as fibrous dysplasia, metaphyseal fibrous defect, or intracortical abscess.

Wilson et al.(1991) reported two tumors (5% of 38 total cases), with their centers close to the cortical margin and classified as "cortically centered".

Chondromyxoid fibroma usually occurs at the metaphyseal region of a long bone. Lesions occuring in the lower extremities account for about 75% of cases, and the proximal tibial metaphysis is a particularly frequent site(Murphy and Price, 1971 ; Rahimi et al., 1971 ; Beggs and Stoker, 1982 ; Gherlinzoni et al., 1983 ; Zillmer and Dorfman, 1989 ; Moser, 1990 ; Mitchell et al., 1992). This case was a 16-year-old female patient with a three-year history of pain involving the distal femoral metaphysis.

The radiographic appearance is variable and depends upon the precise anatomic site of localization(Murphy and Price, 1971 ; Rahimi et al., 1971 ; Beggs and Stoker, 1982 ; Blair et al., 1984 ; Moser and Madewell, 1987 ; Zillmer and Dorfman, 1989 ; Moser, 1990 ; Mitchell et al., 1992). Characteristically lesions of long bone appear as eccentric, sharply

outlined, radiolucent round or oval areas that often cause expansion of part of the cortex. The inner border tends to be scalloped and well delineated by a thin sclerotic rim toward the bone cavity. If not round, the tumors usually have their long axis parallel to the bone(Murphy and Price, 1971 ; Moser and Madewell, 1987 ; Schajowicz, 1987 ; Moser, 1990 ; Wilson et al., 1991). Expansion and erosion of the overlying cortex are common, and cortical breakthrough or periosteal new bone formation may occur rarely(Mitchell et al., 1992). Some of the lesions causing cortical expansion were found to contain areas of aneurysmal bone cyst formation(Zillmer and Dorfman, 1989). The typical ring-or arc-like calcifications seen in many chondromatous lesions are seldom encountered(Mitchell et al., 1992). Chondromyxoid fibroma may often mimic more common tumors, such as giant cell tumor, nonossifying fibroma, aneurysmal bone cyst, and simple bone cyst-(Murphy and Price, 1971 ; Moser, 1990 ; Wilson et al., 1991).

With its superior soft tissue contrast resolution, magnetic resonance(MR) is useful in further characterizing the tumor and in documenting the extent of soft tissue involvement(Aisen et al., 1986 ; Moser and Madewell, 1987 ; Mitchell et al., 1992). The signal characteristics of chondromyxoid fibroma vary with the proportion of chondroid, myxoid, and fibrous tissue within the lesion. Most chondromyxoid fibroma is a predominantly solid tumor with minimal cystic or hemorrhagic components. Typical signal characteristics of tumor were identified in our case, with relatively long T1 and T2 values, resulting in hypointensity on T1 weighted images and hyperintensity on T2 weighted images(Mitchell et al., 1992).

Chondromyxoid fibroma has quite a variety of histologic features, the general pattern being that of a mixture of fibrous, myxoid and cartilagenous elements, sometimes mingled with scattered spicules of new osteoid. It is composed of several nodules, with rounded or irregular areas of myxoid or chondroid tissue, the edges of which show a zone of increased cellularity(Murphy and Price, 1971 ; Rahimi et al., 1971 ; Zillmer and Dorfman, 1989 ; Moser, 1990). The cells within the center of the lobules are stellate to spindle-shaped and usually sparse, in contrast to the characteristic higher cell density towards the periphery(Kreicbergs et al., 1985). Areas of chondroblastic differentiation or multinucleated giant cells may also be present at the periphery(Zillmer and Dorfman, 1989). Neither genuine hyaline cartilage nor truly fibromatous areas are not seen-(Moser, 1990). Tumor cells were at times bizarre, pleomorphic, and binucleate, but rarely contained mitoses. Histologic sections of our case showed typical features of chondromyxoid fibroma composed of mainly myxoid nodules and peripheral fibrous elements with focal chondroid differentiation.

ACKNOWLEDGEMENT

We wish to thank Dr. Beabout JW of the Department of Diagnostic Radiology, Mayo Clinic, Rochester, MN, for his thorough review of the radiologic findings.

REFERENCES

Adler CP . *Case report 338. Skeletal Radiol 1985 ; 14 : 305-8.*

Aisen AM, Martel W, Braunstein EM, McMillin KI, Phillips WA, Kling TF . *MRI and CT evaluation of primary bone and soft tissue tumors. Am J Radiol 1986 ; 146 : 749-56.*

Andrew T, Kenwright J, Woods C . *Periosteal chondromyxoid fibroma of the tibia. A case report. Acta Orthop Scand 1982 ; 53 : 467-70.*

Beggs IG, Stoker DJ . *Chondromyxoid fibroma of bone. Clin Radiol 1982 ; 33 : 671-9.*

Blair WF, Robinson RA, Buckwalter JA . *Chondromyxoid fibroma in a carpal bone. Clin Orthop Relat Res 1984 ; 188 : 199-202.*

Gherlinzoni F, Rock M, Picci P . *Chondromyxoid fibroma. The experience at the Instituto Ortopedico Rizzoli. J Bone Joint Surg 1983 ; 65-A : 198-204.*

Jaffe HL, Lichtenstein L . *Chondromyxoid fibroma of bone. A distinctive benign tumor likely to be mistaken especially for chondrosarcoma. Arch Pathol 1948 ; 45 : 541-51.*

Kreicbergs A, Lonnquist PA, Willems J . *Chondromyxoid fibroma. A review of the literature and a report on our own experience. Acta Pathol Microbiol Immunol Scand 1985 ; 93 : 189-97.*

Kyriakos M . *Soft tissue implantation of chondromyxoid fibroma. Am J Surg Pathol 1979 ; 3 : 363-72.*

Mitchell M, Sartoris DJ, Resnick D . *Case report 713. Skeletal Radiol 1992 ; 21 : 252-5.*

Moser RP . *Cartilaginous tumors of the skeleton. Mosby, Philadelphia 1990 ; 114-54.*

Moser RP, Madewell JE . *An approach to primary bone tumors. Rad Clin North Am 1987 ; 25 : 1049-93.*

Murphy NB, Price CH . *The radiological aspects of chondromyxoid fibroma of bone. Clin Radiol 1987 ; 22 : 261-9.*

Rahimi A, Beabout JW, Ivins JC, Dahlin DC . *Chondromyxoid fibroma : A clinicopathologic study of 76*

cases. Cancer 1971 ; 30 : 726-36.

Schajowicz F . *Chondromyxoid fibroma : Report of three cases with predominant cortical involvement. Radiology 1987 ; 164 : 783-6.*

Sehayik S, Rosman MA . *Malignant degeneration of a chondromyxoid fibroma in a child. Canad J Surg 1975 ; 18 : 354-60.*

Uematsu A, Coy JT, Hodges SO, Goodman RP, Brower TD . *Malignant chondromyxoid fibroma of the scapula. South Med J 1977 ; 70 : 1469-71.*

Ushigome S, Takakuwa T, Shinagawa T, Kishida H, Yamazaki M . *Chondromyxoid fibroma of bone. An electron microscopic observation. Acta Pathol Jpn 1982 ; 32 : 113-22.*

Wilson AJ, Kyriakos M, Ackerman LV . *Chondromyxoid fibroma : Radiologic appearance in 38 cases and in a review of the literature. Radiology 1991 ; 179 : 513-8.*

Zillmer DA, Dorfman HD . *Chondromyxoid fibroma of bone : Thirty-six cases with clinicopathologic correlation. Hum Pathol 1989 ; 20 : 952-64.*