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Low Grade Chondrosarcoma Presenting as Progressive Valgus Limb Deformity in a Growing Period

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A femoral bone tumor causing a valgus deformity by affecting the growth plate was found. Long intramedullary diaphyseal tumor was separated by septum at the metapysis. Low grade chondrosarcoma was confirmed diagnosed by pathologists. Progressive limb deformity can be a sign of bone tumor in growing period.

Key words: valgus limb deformity, low grade chondrosarcoma, growing period

The peak incidence age of chondrosarcoma is late adulthood, and only few cases of chondrosarcoma have been reported in children and adolescents.[1-4] These tumors, when they occur in young people, often appear in unusual sites and may have a more ominous prognosis.[2,4,5] It is subdivided into a variety of ways, including by histological grade, by whether it is primary or secondary, and by whether it is peripheral or central; among these, the single most prognostic sub-classification is the histological grade. Sometimes, the low grade chondrosarcoma is misdiagnosed as a benign cartilaginous bone tumor including enchondroma or periosteal chondroma at the time of preoperative biopsy. To decrease of false negative diagnosis of low grade chondrosarcoma, the clino-radiologic findings such as pain, endosteal scalloping and peritumoral edema are important as well as histologic demonstration. Progressive limb deformity can be a sign of bone tumor in growing children by influencing adjacent epiphyseal plate. We experienced the long meta-diaphyseal intramedullary bony lesion accompanying short intracortical separated lesion by trabecular bone of femur in a 15-year old man who was incidentally detected as a valgus deformity of the same side lower limb. The tumor may have caused premature closure of small lateral area at the distal femur physis and resulted in asymmetric growth of the distal femur.

Case Report

A 15-year-old man was referred for evaluation of a valgus deformity (Fig. 1A) and a lesion of the long meta-diaphysial intramedullary bone tumor of the left femur. He had pain around the knee

Figure 1. A 15-year-old man has progressive valgus deformity of left lower limb. (A) Photograph shows valgus deformed lower limb. (B) Radiograph demonstrate 27 degree valgus deformed femur at the metadiaphysis area.

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for 6 months as a dull ache especially during exercise, but he had no trauma history. His body weight was 98 kg and height was 178 cm. Physical examination revealed a slight tenderness on the lateral aspect of the distal thigh that was not clear of its swelling or palpable mass. The plain radiographs revealed a cortical ballooning of an elliptical shape at 6.5 cm above of the knee joint line that had distinctly distinguished with medullary canal by sclerotic border but faintly with surround lateral muscles (Fig. 1B). The valgus angular deformity of the femur at the meta-diaphysis was 27 degrees. The magnetic resonance imaging (MRI) showed intramedullary lesion that was measured 18 cm and intracortical lesion measured 8 cm in longitudinally through distal meta-diaphysis of the left femur (Fig. 2A). These two distinguish lesions showed same signal intensity. Computed tomography (CT) showed obviously calcific stippling in the long intramedullary lesion and distinguished intracortical lesion as well (Fig. 2B & C). Reconstruction CT (Fig. 2B & C-inferior) and coronal MRI (Fig. 3) showed mountain peak shape of the lateral growth plate to the distal tail of the intracortical lesion.

According to clino-radiologic finding, preoperative diagnosis was considering rather a low grade chondrosarcoma than an enchondroma although patient's age is young. An incisional biopsy was performed through direct approach to the lateral cortical ballooned area and pathology was resulted in benign chondroid lesion. The surgery was performed with 6-cm segmental resection of the angular deformed femur. Other lesion was treated by curettage, intramedullary reaming and coablation of argon laser. The reconstruction was performed 1.5 cm over lengthened segmental allograft, non-vascularized fibular autograft which was harvested from the same lower leg and loaded to the intramedullary area of the femur and allograft. The stabilization was obtained by anatomical plate osteosynthesis.

Figure 2. (A) T1- weighted coronal images show the meta-diaphysial intramedullary and intracortical lesion. (B) Coronal computed tomography obtained after open biopsy showing valgus curved femur may caused close connecting growth plate of the intracortical lesion (arrow). (C) Axial computed tomography sections showing the intramedullary calcific stippling (superior), distinguished intracortical osteolytic lesion (middle), and elliptical tailed of lateral intracortical lesion toward growth plate (inferior).

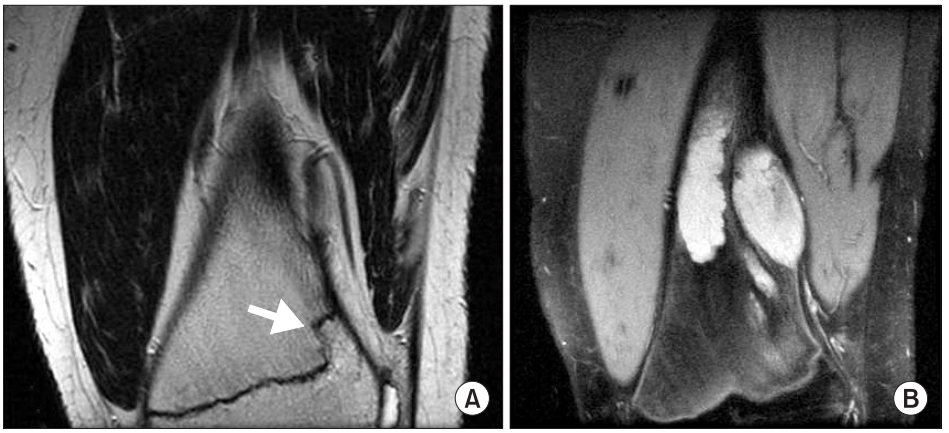

Figure 3. (A) and (B) Coronal MRI showed mountain peak shape of the lateral growth plate to the distal tail of the intracortical lesion.

The gross finding of coronal cut specimen showed thin and eroded inner border of cortex (Fig. 4A). The intramedullary and intracortical separated lesion divided by thin bony septum which has lamellar in pattern and contained Haversian systems as usually seen in cortical bone (Fig. 4A & B). The specialized musculoskeletal tumor pathologist concluded grade I chondrosarcoma by invasive cartilaginous lesion to the surround trabecular bones (Fig. 4C & D). The patient did not have any adjuvant therapy for the tumor. His height showed 2.5 cm growth in one year after the surgery and therefore, the limb length discrepancy was measured to be 1 cm. We tried to minimize the limb length discrepancy with a simple method, and performed contralateral distal femur and proximal tibia transient epiphysiodesis with staples (Fig. 5A).

Six-year follow up radiology including plain X-ray (Fig. 5B), positron emission tomography (PET), bone scan, and MRI showed balanced normal limb axis, no local tumor recurrence and no distant metastasis. The left leg was measured 2 cm short at last follow up, but he could walk normally using shoe insole.

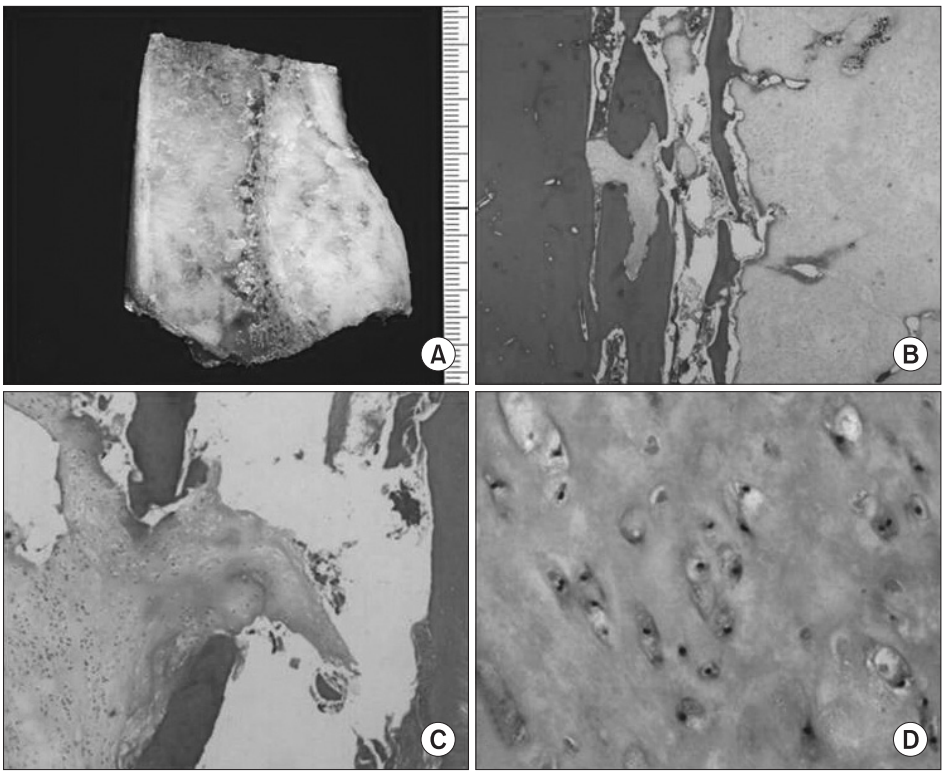

Figure 4. (A) The photograph of coronal cut specimen reveal the distinguished intramedullary and intracortical lesion. (B) Photomicrograph of intramedullary and (C) intracortical lesion showing a low grade chondrosarcoma with intrusion of surrounding mature lamellar bone, (Hematoxylin and eosin, ×100). (D) The obvious atypia of chondrocytes was not revealed on the high-magnification photomicrograph (Hematoxylin and eosin, ×400).

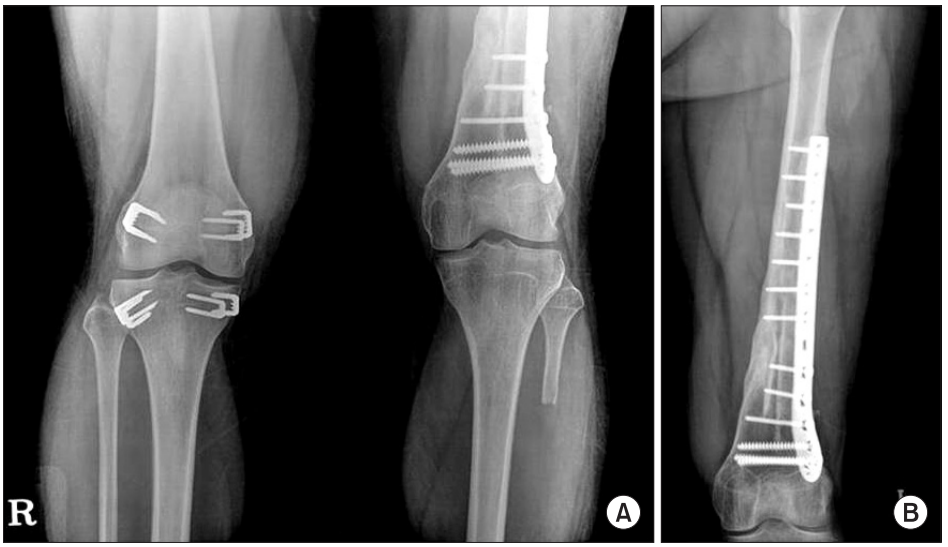

Figure 5. Post operative six-year follow up (A) both knee antero-posterior radiograph shows the epiphysiodesis around right knee and different knee joint level by shortening of left femur. (B) Left femur antero-posterior radiograph demonstrates well incorporation of allograft and intramedullary loaded autogenous fibular graft with correction of deformity.

Discussion

The presence of a large sized central chondrosarcoma and an intracortical chondrosarcoma within the femur incidentally detected by its angular deformity in the adolescent age has not been reported. Two similar cases have been described with titles "concurrent enchondroma and periosteal chondroma of the humerus mimicking chondrosarcoma" in 19-year old woman,[6] and "concurrent periosteal chondroma and enchondroma of the fibula mimicking chondrosarcoma" in 13-year old boy.[7] They performed wide excision because the imaging findings made suspicious for chondrosarcoma although the pathologic results had described benign-looking cartilage tumor in the preoperative open biopsy.[6,7] In the English literature we found only one report of an intracortical chondrosarcoma which was located in the subtrochanteric area of the left femur with radiological revealed 1 cm in diameter and small lytic lesion in 42-year old man.[8] They performed open biopsy by curettage followed by en bloc surgical excision because the biopsy result was reported grade-2 myxoid chondrosarcoma.[8]

In the current case, the pathologic result was changed to a low grade chondrosarcoma after the operation from a preoperative diagnosed benign chondroid tumor. Our multidisciplinary team including pathologist, radiologist and orthopaedic oncologist discussed about the pathologic diagnosis and confirmed as grade I chondrosarcoma rather than enchondroma although it could be showing more aggressiveness in the children. In the literature, the operation in selected patients with a less radiographically aggressive Grade I chondrosarcoma can be safely treated with intralesional curettage without compromising patient outcome.[9]

At first, we worried about using the term of intracortical chondrosarcoma because it might be a periosteal chondrosarcoma with cortical destruction or an intramedullary lesion with septation to cortical erosive ballooning. In the current case, the lesion was no soft extension beyond cortex and periosteum and intracortical confined lesion made by sharply divided of the cortex like elliptical shape. Importantly, histologically chondroid lesion was encircled by cortical lamellar bone.

The degree of malignancy of a chondrosarcoma is determined by several histologic criteria. These include structural characteristic, cytologic findings, and replicate activity. Certain other histologic signs are also indicative of malignancy, such as invasion of trabeculae by tumor tissue, infiltration of bone marrow and Haversian systems and permeation through the cortex. However, these features, which are essential for the discrimination between an enchondroma and a low-grade chondrosarcoma, are not an integral part of an accepted grading system.[10]

We think that distal femur angular deformity which was incidentally detected as valgus deformed limb that may have been caused by growth plate trapping of tumor.

Progressive limb deformity can be a sign of the asymmetrical involvement of physis by adjacent bone tumor in a growing period.

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성장기 저등급 연골육종에 의한 사지의 진행성 외반

강현귀 • 박원서* • 박석연*

국립 암센터 골연부 종양 클리닉, *병리과

성장기 인접 성장판에 영향을 주어 하지의 외반 변형을 보이는 대퇴골 종양이 발견되었다. 골간에 긴 골수내 종양은 성장판 근처의 골간단에서 격막에 의해 나누어졌으며 병리 검사상 저등급 연골육종으로 진단 되었다. 성장기 점진적 하지 변형이 골종양의 성장판 영향에 따른 증상일 수 있다.

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