Scintigraphic Findings of Fibrous Dysplasia

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= 국문초록 =

섬유성 골 이형성증의 골 신티그라피 소견

한일병원 내과

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진단방사선과학교실

강 홍 식

골 신티 그라피는 섬유성 골 이형성증의 초기 병소와 다골성 형 섬유성 골 이형성증의 진단에 예민한 검사 방법이나, 양성 또는 악성 골 병변과의 감별진단에 어려움이 있었다. 이에 저자들은 조직학적으로 확진된 섬유성 골 이형성증 환자 17명에서 (단골성 형 : 12명, 다골성 형 : 5명) ^{99m}Tc-MDP (20 mCi 또는 7,400 MBq) 주사후 2~4시간에 전면, 후면 전신상과 부분상을 얻은 30부위 병소를 X선 사진과 비교분석 하였으며 CT, MRI 영상과 병리조직 소견을 얻었다. 병소 침습 부위별로는 대퇴골-10예, 경골-4, 요골-2, 늑골-2, 척추골-2, 후두골-2, 장골-2, 두정골-1, 하악골-1, 접형골-1, 견갑골-1, 쇄골-1, 척골-1예 였으며 5명의 다골성 형에서는 : 우 대퇴골과 우 비골 : 양측 말단 대퇴골 : 좌 경골과 좌 대퇴골 : 우 요골과 우 장골 : 양측 대퇴골, 양측 경골, 접형골, 두정골, 척추골, 늑골과 장골이었다. 골 신티 그라피 소견상 30예중 28예에서(28/30, 93.3%) 방사능 섭취 증가를 보였으며 2예에서(2/30, 6.7%)는 정상 방사능 섭취 소견을 보였으나 각각은 X선상 불투명 유리상과 골 흡수 병변을 보였다. 하악골의 골 3상 스캔상 병소에 현저한 혈류 증가가 관찰되었다. X선 소견상 30예중 11예에서(11/30, 36.7%) 불투명 유리상의 병변을, 골 변형을 동반한 1예를 포함한 18예에서는 (18/30, 60.0%) 골흡수 병변을 보였으며, 1예의 늑골 병변은 정상소견을 보였다(1/30, 3.3%). 이

상에서 골 신티 그라피 만으로 섬유성 골 이형성증을 진단 하는데는 주의를 요하나 골 대사의 동적 측면인 혈류와 골 재형성 양상 특히, 초기 병변과 단골성 형 침습에서 다골성 형을 진단하는데 필수적이며 결론적으로 골 신티 그라피와 X선 촬영등은 섬유성 골 이형성증 진단에 상호보완적인 검사 방법으로 사료된다.

INTRODUCTION

Fibrous dysplasia is a benign skeletal disorder of unknown etiology, characterized by fibrous replacement of portions of the medullary cavities of a bone or bones[1]. Roentgenographic features of disease are well known[2~5], and although the skeletal lesions of fibrous dysplasia present a variety of roentgen appearances, their features usually are sufficiently disinctive to assure diagnosis. In the long bones, the basic change is replacement of the medullary cavity, which produces lesions varying from those that are completely radiolucent to those of a homogenous, ground glass, increased density, depending on the amount of fibrous or osseous tissue deposited in the medulla. Expansion of bone occurs in the ribs and skull, and also is seen commonly in long bones. Radiolucent abnormalities are more common than dense lesions.

Abnormal fibrous tissue containing trabeculae of poorly calcified bone fills the medullary cavity at single or multiple sites (monostotic vs polyostotic forms)[6]. Eighty five percent of patients with fibrous dysplasia develop pathologic fractures. Bones typically involved include in decreasing order of incidence, the ribs, tibia, femur and maxilla[7~8]. The polyostotic form may be accompanied by skin pigmentation changes and endocrine abnormalities (Albright's Syndrome)[9].

Bone imaging provides a sensitive means of evaluating patients with fibrous dysplasia. A knowledge of the variable bone scan patterns of fibrous dysplasia is essential for their distinction from more common disorders such as Paget's disease, metastatic lesions and fractures. Dentinal dysplasia is a rare inherited disorder that affects both dentitions[10,11]. Their two types are type I, radicular type and type II, coronal type[12,13]. We report the radionuclide manifestation of the disease, and review the value of bone imaging, since bone imaging demonstrates dynamic features.

MATERIALS AND METHODS

A retrospective analysis of 30 lesions was performed on 17 patients with fibrous dysplasia who underwent bone imaging. Of the 17 patients, nine were women and eight were men, 12 monostotic and five polyostotic forms. The age range was 4~51 years old (average=27.6 years). Bone imaging was performed with ^{99m}Tc-MDP.

Anterior and posterior whole body images were obtained 2~4 hours after intravenous administration of a ^{99m}Tc bone imaging agent (20 mCi or 7400 MBq), and the spot images were added when necessary. Siemens LFOV (Siemens Gamma Sonic Inc.) or ON 410 (Ohio Nuclear) scintilation cameras equipped with general-purpose, parallel-hole collimators were used. All bone scans were reviewed by two observers, and the grades of radioisotope accumulation were classified into two categories: +=normal, and ++=increased over normal. These imaging findings were compared with the findings of X-ray films and additional CT or MRI were reviewed.

RESULTS

Twelve patients with monostotic fibrous dysplasia and five patients with polyostotic fibrous dysplasia had bone imaging performed. After then thirty

Table 1. Comparative Findings of Scintigram and Roentgenogram

Roentgenogram/ Scintigram uptake	Number			
	Ground-glass appearance	Cyst-like changes	Deformity	Normal
(++) Increased	11 (36.7%)	15 (50.0%)	1 (3.3%)	1 (3.3%)
(+) Normal		2 (6.7%)		

(++) : Increased over normal uptake. (+) : Normal uptake.

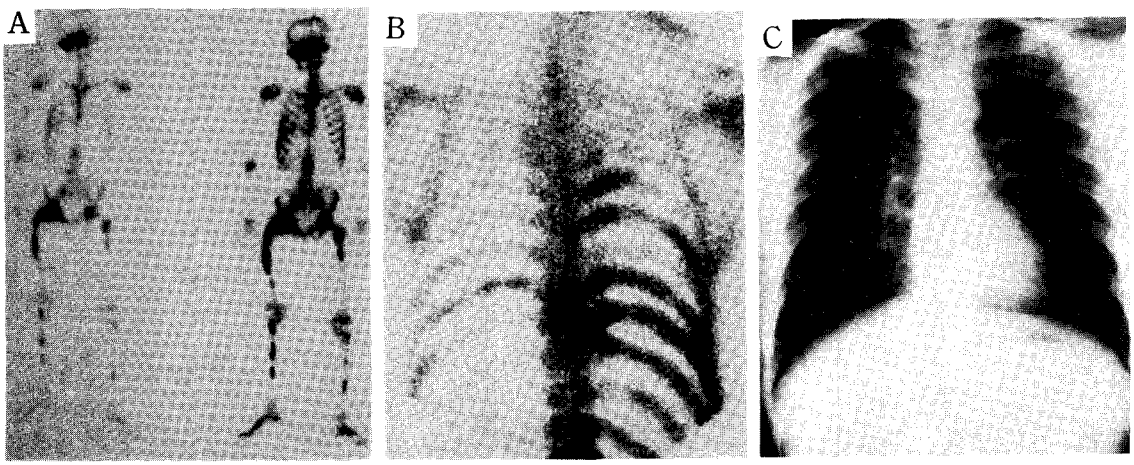

Fig. 1. Polyostotic fibrous dysplasia. 20-year-old man.
(A) The scintigram showed areas of increased uptake of radioisotope in the sphenoid, occipital, spine, ribs, ileum, both femur and both tibia.
(B) The scintigram showed multiple increased uptake of the ribs.
(C) The chest PA roentgenogram showed no definite lesion of the ribs.

lesions of fibrous dysplasia were analyzed. The sites of involvement were femur-10 cases, tibia-4, radius-2, rib-2, spine-2, occipital-2, ileum-2, parietal bone-1, mandible-1, sphenoid-1, scapula-1, clavicle-1 and fibula-1 case. In the polyostotic forms, the sites of involvement were right femur and right fibula; both distal femur; left tibia and left femur; right radius and right ileum; and both femur, both tibia, sphenoid, occipital, spine, rib and ileum.

Table 1 demonstrates the grade of ^{99m}Tc MDP uptake in 30 lesions of fibrous dysplasia, which were roentgenologically divided into four patterns: ground-glass appearance in 11 lesions, cyst-like changes in 17 lesions, bone deformity in 1 lesion, and normal finding in 1 lesion. Bone imaging with ^{99m}Tc MDP revealed an increased uptake of radioisotope in the 28 out of 30 lesions (93.3%) of fibrous dysplasia. Two of 30 lesions showed ground-glass appearance and osteolytic changes respectively. A 'three phase' bone scan in the case of mandible showed increased blood flow to the lesion site.

Roentgenologic findings showed ground-glass appearance in 11 lesions (11/30, 36.7%), osteolytic changes with or without sclerotic rim in 18 lesions (18/30, 60.0%) including deformity in one case and normal in only one case (1/30, 3.3%) Fig. 1 shows the scintigram and roentgenogram of a 20-year-old man with polyostotic fibrous dysplasia. The scintigram showed areas of increased uptake of radioisotope in sphenoid, occipital, spine, ribs, ileum, both

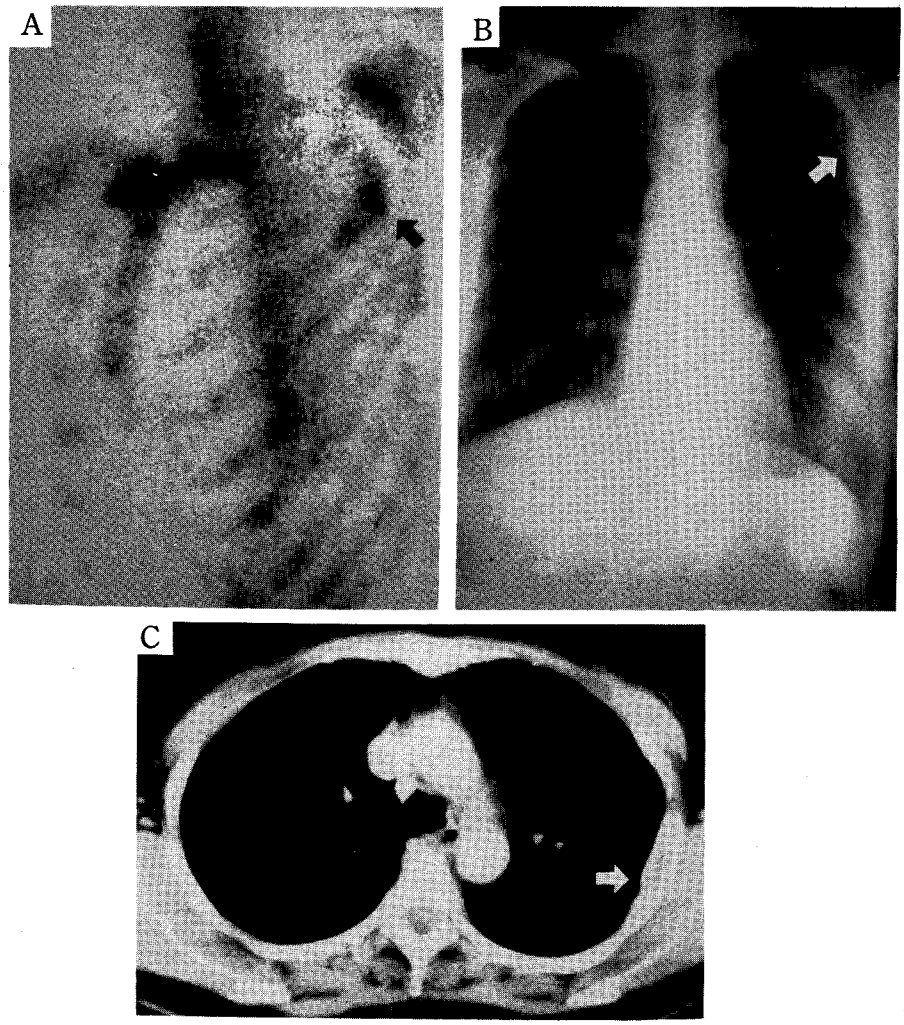

Fig. 2. Fibrous dysplasia, Left fourth rib. 36-year-old woman.
(A) The scintigram showed increased uptake in the left fourth rib.
(B) The chest PA roentgenogram showed osteolytic lesions with ballooning and cortical thinning of the axillary arc of left fouth rib.
(C) The chest CT showed cortical thinning and replacement of bone marrow with soft tissue of the axillary arc of left fourth rib, just below the aortic arch level.

femur and both tibia. The chest scintigram showed multiple increased uptake of ribs but the chest roentgenogram showed no definite lesions of ribs.

Fig. 2 shows the scintigram, roentgenogram and CT of a 36-year-old woman with monostotic fibrous dysplasia of left fourth rib. The scintigram showed increased uptake. The chest roentgenogram showed osteolytic lesions with ballooning and cortical thinning of axillary arc of rib. The chest CT showed cortical thinning and replacement of bone marrow with soft tissue of axillary arc of rib.

Fig. 3 shows the scintigram, roentgenogram and MRI of a 20-year-old man with polyostotic fibrous dysplasia in medial metaphysis of both distal femur. The scintigram showed no increased radionuclide activity. The AP roentgenogram showed radiolucent

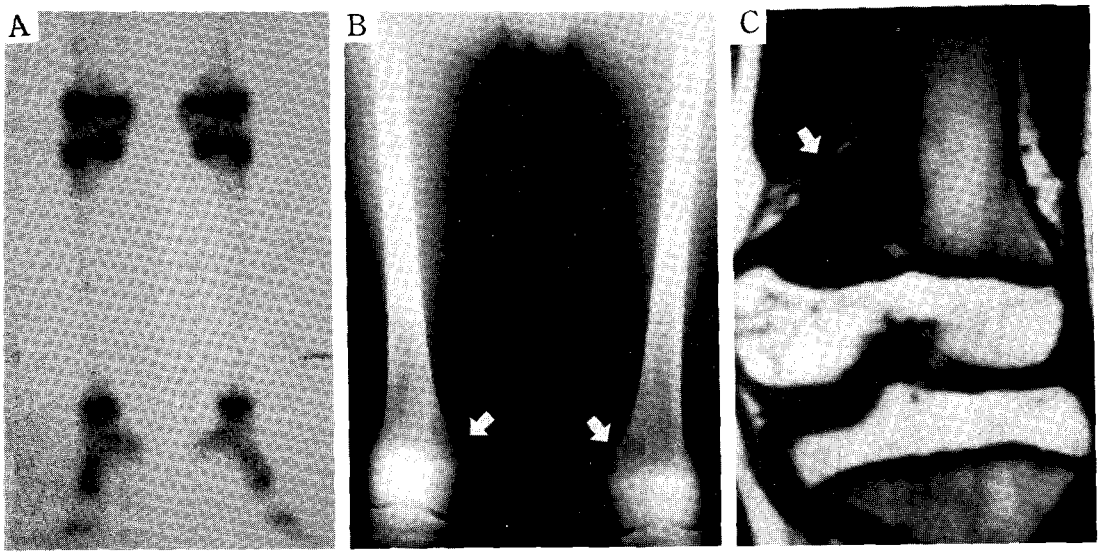

Fig. 3. Fibrous dysplasia, Medial metaphysis of both femur. 20-year-old man.
(A) The scintigram showed no increased uptake.
(B) The roentgenogram showed two radiolucent areas of ground-glass appearance with sclerotic rim.
(C) The T1 weighted MRI showed low signal lesions. (variable signal on T2 weighted MRI)

areas of ground-glass appearance with sclerotic rim. The T1 weighted MRI showed low signal lesions (variable signal on T2 weighted MRI).

Fig. 4 shows the light and polarizing microscopic findings of a 30-year-old man with monostotic fibrous dysplasia of fibula. A photomicrograph of H and E stain demonstrates a central osseous spicule surrounded by fibrous tissue, devoid of osteoblastic rimming. A photomicrograph with polarization microscopy shows a random fibrillar pattern of collagen tissue and cells in woven bone. The demonstration of woven bone is essential in establishing a fibrous dysplasia. The polarized light helps confirm the random distribution of the matrix. Lamellar bone would project in an organized pattern.

DISCUSSION

Fibrous dysplasia (FD) was first described by Albright and his associates in 1937 in conjunction with the identification of children with precocious puberty and cafe-au-lait spots, simulating neurofibromatosis. Later, Lichtenstein and then Jaffe described the bony lesions as occurring separately from the endocrine system (an endocrine/hormonal cause is suggested in the Albright's syndrome). In considering the clinical features, the cause remains unknown. A slight preponderance for females exists. The usual age of presentation is in the first two decades of life.

The two major forms of the disorder exist: monostotic and polyostotic-, the polyostotic form showing a tendency to a monomelic distribution. The Albright syndrome consists of florid skeletal involvement, precocious puberty and pigmentation of the skin, mainly on the back and predominantly unilateral in distribution. The lesions of the skin may be reminiscent of the cafe-au-lait spots of neurofibromatosis. The clinical features include skeletal deformities and occasionally pain. The skeletal

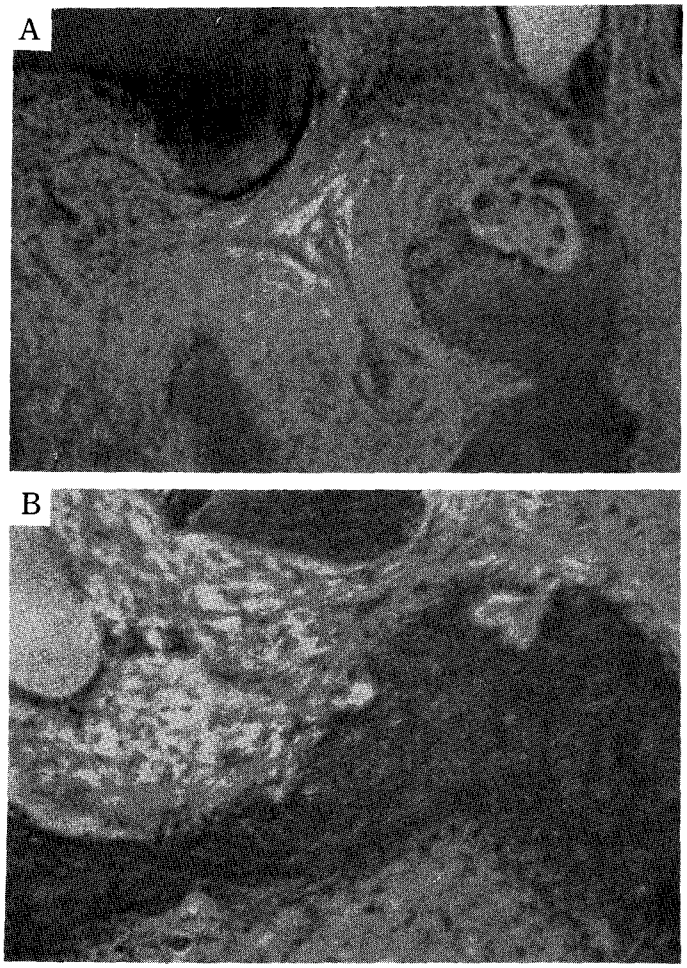

Fig. 4. Fibrous dysplasia, Fibula. 30-year-old man.
(A) A photomicrograph with H and E stain demonstrates a central osseous spicule surrounded by fibrous tissue, devoid of osteoblastic rimming.
(B) A photomicrograph with polarization microscopy shows a random fibrillar pattern of collagen tissue and cells in woven bone. The demonstration of woven bone is essential in establishing a fibrous dysplasia. Polarized light helps confirm the random distribution of the matrix. Lamellar bone would project in an organized pattern.

deformities occur in the presence of widespread involvement of the major long bones. Not infrequently, pathological fractures associated with thinning of the cortical surfaces and insufficiency fractures due to overt trauma, particularly of the major long bones, are observed.

The bones affected by the disorder, principally include the femora, innominate bones, ribs, skull and facial bones. Fordham reported[14] that in a limited series of cases with fibrous dysplasia, scintigraphic imaging had been useful in demonstrating disease where none was suspected, or in demonstrating polyostotic involvement where only monostotic disease was clinically suspected. The lesions of fibrous dysplasia tend to demonstrate striking uptake, which may represent a function of a rich vascular supply to the involved bone as well as some degree of bone remodelling. Due to the increased vascularity

of fibrous dysplastic lesions, increased concentration of bone seeking radionuclides is seen both in early perfusion and delayed static bone image.

Plain film radiography is often sufficient for diagnosis of fibrous dysplasia while bone imaging using ^{99m}Tc-MDP is more sensitive in detecting earlier lesions and polyostotic involvement[15]. Characteristic radiographic findings include expansile bubble-like lesions and ground-glass texture. The lesions may appear as radiolucent or sclerotic, depending on the degree of ossification[16~18].

In the bone-replacing phase of FD, the lesions are radiolucent, with the appearance of cyst-like areas surrounded by a thick sclerotic border. The cortex is thin and the medullary cavity widened in such instances, resulting in pathological fractures on occasion. When the lytic lesions are eccentric, the cortex may be thickened on one side. The "smoky" appearance of medullary lesions is probably due to faintly mineralized osteoid. Small islands of calcification may be present.

In the bone-forming phase, the new bone is confined mainly to the medullary cavity, in contrast to the thickening of the cortex that occurs in Paget's disease. Mainly, sclerotic and lytic lesions may be interspersed. Of interest, in lesions in the spine, the almost invariable thinning of the disk cartilage above or below the involved vertebral body, is encountered. The cause for this is unknown. Radionuclide scans often, but by no means invariably, are positive in the areas of skeletal involvement.

We demonstrated that bone imaging revealed a high percentage (93.3%, 28/30) of areas of increased uptake of ^{99m}Tc-MDP in lesions of fibrous dysplasia where roentgenogram showed expected changes. In a small number (6.7%, 2/30) of lesions, however, the increased uptake of radioisotope did not occur, although the X-rays were diagnostic. The grade of radioisotope uptake probably depends on the vascular supply and the degree of remodelling of the lesions. The findings on roentgenograms demonstrate only the distribution of bone calcium, and not the pathophysiological activity of diseased bone. Thus, bone imaging is useful to evaluate the activity or prognosis of fibrous dysplasia. When a diagnosis of fibrous dysplasia is made by bone scintigram alone, false-negative can result, since scintigraphically silent areas may exist in a small number of lesions (6.7%, 2/30) with cystic changes on the roentgenograms. Roentgenograms fail to demonstrate abnormality in only one lesion (3.3%, 1/30) of rib with increased radioisotope on the scintigrams.

CT scanning may be also useful in defining the extent of craniofacial involvement[19]. Imaging of fibrous dysplasia must differentiate between Paget's disease, osteoblastic metastasis, or fractures. It has been noted that lack of preservation of the normal outline of the bone is often seen in the expansile lesions of fibrous dysplasia. This is in contrast to Paget's disease, where the bony outline is maintained[20].

On MR, the fibrous dysplastic lesion causes an "expanded" bony contour and is characterized by decreased signal on T1-weighted image. The signal on T2-weighted MR is variable[21].

Pathologically, the medullary cavity is replaced by a material with a fibrous, gritty consistency. Microscopically, a fibrous tissue matrix predominates with occasional osseous spicules. Cartilagenous rests, as well as giant cells, may be observed sporadically. Such findings are considered characteristic[22].

In summary, bone scans are indispensable in evaluating the dynamic of bone mineral behavior and in demonstrating disease when none was suspected, or in visualizing polyostotic involvement in the cases when only monostotic disease was suspected cliniclally. Care must be taken in the diagnosis of fibrous dysplasia with bone imaging alone.

In conclusion, bone scintigram and roentgenogram are complementary procedure in the diagnosis of

fibrous dysplasia.

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