CT and MRI of fibrous dysplasia of the spine

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Objectives: The purpose of this study was to present the CT and MRI findings of patients with fibrous dysplasia (FD) of the spine.

Methods: Among the patients with pathologically proven skeletal FD, 12 (8 males and 4 females; mean age, 43 years) who were evaluated with either spine CT or MRI were included. The number and location of the involved vertebral segments, the presence of lytic lesions, ground-glass opacity (GGO), an expansile nature, cortical disruption, a sclerotic rim, a decrease in body height and contour deformity were examined on CT scans ($n=12$), while signal intensity, enhancement patterns and the presence of a dark signal rim on the lesion were examined using MRI ($n=9$).

Results: Nine patients had polyostotic FD, including one with an isolated spinal localisation, while three had monostotic FD. An expansile nature ($n=3$) and osteolytic lesions with GGO ($n=3$) were seen. On CT images, GGO was noted in all patients. An expansile nature ($n=11$) and presence of lytic lesions ($n=11$) were noted. A decrease in body height ($n=9$) and sclerotic rim formation ($n=9$) were indicated. Contour deformities were visible in six patients. The MRI findings were non-specific. Dark signal rims were visible on MRI in seven patients.

Conclusion: Typical imaging findings of extraspinal FD were noted on spine CT scans. These characteristic CT imaging findings of spinal FD may be helpful in differentiating FD from other common spine diseases.

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Fibrous dysplasia (FD) is a developmental defect of osteoblastic differentiation and bone maturation of unknown origin [1]. FD may affect the skeleton either in isolation (monostotic FD, rate, 70–80%; and polyostotic FD, rate, 20–30%) or in variable combination with endocrine and cutaneous abnormalities (McCune–Albright syndrome) [2–5]. Although patients afflicted with FD may present at any age, they are typically young and in the first and second decades of life [6]. After puberty, dysplastic areas rarely expand [7]. In general, the progression of the skeletal lesions tends to be exhausted once patients have reached adult age [7]. The diagnosis of FD is difficult in adult patients because cases in older patients, especially those involving the spine, are rare.

FD represents approximately 7% of all benign tumour-like bone lesions [8]. However, the spine is affected in only 2.5% of cases, and FD of the spine is very rarely observed without there being disease elsewhere in the body [9]. Spinal involvement occurs mostly in the polyostotic form of FD; it is unusual for it to occur in the monostotic form [9, 10]. When present in the elderly with multiple vertebral lesions, a biopsy may be indicated

because metastatic disease or multiple myeloma may simulate a benign non-aggressive process. Therefore, active diagnosis and radiological familiarity of spinal FD are thought to be essential for preventing unnecessary examinations or procedures.

The purpose of this study was to present the CT and MRI findings of spinal FD, which is rare, and to assess the efficacy of CT in the diagnosis of FD.

Methods and materials

This study was approved by the Institutional Review Board of our institution; because it was a retrospective study, informed consent was not required. Pathology databases at our institution and at three other institutions were searched for cases of pathologically proven skeletal FD in patients examined between January 2005 and March 2010. All patients with skeletal FD and spinal involvement who underwent spine CT and/or MRI were included. A total of 12 patients (8 males and 4 females; mean age, 43 years; age range, 12–62 years) were included in the study. Six patients underwent an initial spine CT and/or MRI owing to spinal symptoms such as back pain or radiculopathy. The other six patients underwent spine CT and/or MRI after the incidental detection of spinal abnormalities on images obtained

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owing to extraspinal symptoms. In addition, 6 of the 12 patients were diagnosed with FD by biopsy or excision of the vertebrae or sacrum. The remaining six patients underwent a biopsy or an operation on sites other than the spine. The total extent of skeletal involvement in patients with polyostotic FD was determined by all imaging modalities listed on picture archiving and communication system.

Radiographs of the spine were obtained for all 12 patients and were investigated for presence or absence of lesions. After detecting lesions, we described the abnormal findings, presence of osteolytic or sclerotic lesions and compression fractures. CT examinations were performed with multidetector-row CT (4- or 16-channel) scanners in all 12 patients. The imaging parameters were as follows: 120 kVp, 230–250 mAs and slice thickness, 1.3–2.5 mm. In nine patients, MRI scans were obtained with a 1.5 T unit using a spine array coil. The pulse sequences consisted of axial and sagittal T_1 weighted [repetition time (TR) range, 500–834 ms; echo delay time (TE) range, 10–13 ms] and turbo spin-echo (TSE) T_2 weighted (TR range, 2900–4820 ms; TE range, 105–143 ms) images without fat suppression. Six patients had fat-suppressed contrast-enhanced axial (TR range, 400–815 ms; TE range, 10–14 ms) and sagittal TSE T_1 weighted (TR range, 432–650 ms; TE range, 10–13 ms) sequences. The slice thickness was 3–4 mm for sagittal images and 5–6 mm for axial images. The fields of view were 130–170 mm for axial images and 230–280 mm for sagittal images. The matrix size ranged from 256×179 to 320×352. Two radiologists independently reviewed the CT and MR images and determined the results by consensus. The number and location of the involved vertebral segments, presence or absence of lytic lesions, ground-glass opacity (GGO), expansile nature, cortical disruption, sclerotic rims, decrease in body height and cortical contour deformities were examined on CT scans. Signal intensity, enhancement patterns and the presence or absence of dark signal rims on the lesions were examined on MR images. The signal intensities of the lesions were compared with the signal intensity of the spinal cord.

A lower or iso-signal intensity compared with that of the spinal cord was defined as “low signal”, and a higher signal intensity compared with that of the spinal cord was defined as “high signal”.

Results

The imaging modalities used and the type and site of FD for all study patients are summarised in Table 1. Nine patients had the polyostotic type and three had the monostotic type. Among the nine patients with the polyostotic type, one had an isolated spinal localisation. FD affected the vertebral body and posterior element or sacral body and ala in 8 of the 12 patients. Isolated vertebral body involvement of FD was noted in a further three patients. For the remaining patient, FD was located in the sacral ala alone.

On plain radiography, no abnormal findings were detected in two patients. Eight patients had compression fractures of the vertebrae and five had peripheral sclerosis within the vertebral bodies. An expansile nature showing endosteal scalloping in the cortex of the vertebral body was noted in three patients. Osteolytic lesions with GGO were visible in three patients (Figure 1a).

The results of CT imaging are presented in Table 2. GGO was noted in all cases (100%). An expansile nature was present in 11 patients (92%). Lytic lesions were present in 11 patients (92%; Figure 1b). A decrease in body height occurred in 9 patients (75%), and sclerotic rim formation was present in 9 patients (75%; Figure 1c). Contour deformity with bone remodelling was visible in 6 patients (50%; Figure 1b). Cortical disruption was not seen in any patients.

The MRI findings of patients with spinal FD were non-specific in this study (Table 3). On T_1 weighted images, low signal intensity lesions were present in 6 patients (6/9, 67%; Figure 2a), and heterogeneous signal intensity lesions were present in 3 patients (3/9, 33%). On T_2

Table 1 Imaging modalities used and types and sites of fibrous dysplasia in all patients

Patient	Age (years)	Gender	Type	Site	Imaging modality
1	51	M	Polyostotic	Craniofacial bones, sternum, rib, spine (C1–4, T1–4, 5, 7, 11, 12, L5), iliac bones, scapula	CT
2	12	M	Polyostotic	Rib, spine (T3–all L), scapula, sternum, foot, hand, humerus	CT
3	44	F	Polyostotic	Rib, spine (C1, 2, 5, all T, L-vertebrae and sacrum), pelvic bones, skull, PNS, femur, ulna, radius, humerus, tibia, fibula	CT
4	47	M	Polyostotic	Spine (C4, 6, 7, T6–9, L2), rib, clavicle, sternum	CT, MR (CE)
5	62	M	Polyostotic	Rib, spine (C2, 4, T4–12, L5), pelvic bones, scapula, femur, humerus	CT, MR (CE)
6	30	F	Polyostotic	Spine (T8), rib	CT, MR
7	60	M	Polyostotic	Spine (L4, 5), skull, PNS	CT, MR
8	41	M	Polyostotic	Spine (L2–5), PNS, shoulder, rib	CT, MR
9	43	F	Monostotic	Sacral ala	CT, MR (CE)
10	42	F	Monostotic	Spine (T12)	CT, MR (CE)
11	41	M	Polyostotic with isolated spinal localisation	Spine (L1, 2)	CT, MR (CE)
12	38	M	Monostotic	Sacral body and ala	CT, MR (CE)

C, cervical spine; CE, contrast-enhanced axial and sagittal spin-echo T_1 weighted images; F, female; L, lumbar spine; M, male; PNS, paranasal sinus; T, thoracic spine.

Figure 1 A 61-year-old male with polyostotic fibrous dysplasia involving the lumbar vertebrae. (a) Plain lateral view of the lumbar spine showing decreased height of the L3, 4, and 5 vertebral bodies. An expansile osteolytic lesion with faint ground-glass opacity (GGO) is noted in the body of L4. There is a peripheral rim and diffuse sclerotic lesions within the body of L5. (b) Axial CT scan at the L4 level showing a lesion with GGO within the expansile osteolytic lesion in the vertebral body. In addition, sclerotic lesions are seen in the vertebral body and posterior element. (c) Reformatted sagittal CT scan showing the contour deformities with bone remodelling due to an expansile nature and compression fractures in the L4 and 5 vertebrae.

weighted images, all lesions showed heterogeneous signal intensities with variable signals (Figure 2b). Dark signal rims were visible in seven patients (Figure 2). Homogeneous enhancements of the lesions were seen in 3 patients (3/6, 50%). Faint enhancement and peripheral nodular enhancement were present in 2 (2/6, 33%) and 1 patient(s) (1/6, 17%), respectively.

Discussion

FD, a benign affliction of the skeleton, consists of one or more foci that are composed of cellular fibrous tissue containing irregular bone trabeculae that lead to distortion and structural weakness of the bone [7]. Involvement of the spinal column in FD is uncommon,

Table 2 Characteristic CT findings of spinal fibrous dysplasia in all patients

Patient	Locations	CT findings of the spine						
		Lytic lesions	GGO	Expansile nature	Cortical disruption	Sclerotic rim	Decrease in body height	Contour deformities
1	Most posterior elements, rarely body	+	+	+	–	–	–	–
2	Bodies and posterior elements	+	+	+	–	+	+	+
3	Bodies and posterior elements	+	+	+	–	+	+	+
4	Bodies and posterior elements, contiguous invasion of the posterior aspect of the rib	–	+	+	–	–	+	+
5	Bodies and posterior elements	+	+	+	–	+	+	+
6	Body and posterior element	+	+	+	–	+	+	+
7	Bodies and posterior elements	+	+	+	–	+	+	+
8	Bodies	+	+	+	–	+	+	–
9	Sacral ala	+	+	+	–	+	–	–
10	Body	+	+	+	–	+	+	–
11	Bodies	+	+	+	–	–	+	–
12	Sacral body and ala	+	+	–	–	+	–	–

GGO, ground-glass opacity.

Table 3 The results of MRI findings of spinal fibrous dysplasia in nine patients

Patient	Age (years)	Gender	T ₁ weighted images	T ₂ weighted images	Enhancement pattern	Dark signal rim
1	47	M	Iso to low SI	Iso to high SI	Faint peripheral	–
2	62	M	Low SI	Heterogeneous low SI with several small cysts	Faint	+
3	30	F	Low SI	Iso to low SI	N/A	+
4	60	M	Low SI	Mixed low and high SI	N/A	+
5	41	M	Low SI	Intermediate SI with several high SI portions	N/A	+
6	43	F	Mixed low and high SI	Intermediate to high SI	Peripheral nodular	+
7	42	F	Low SI	Intermediate to high SI	Homogeneous	+
8	41	M	Low SI with dark foci	Heterogeneous high SI with dark foci	Homogeneous	–
9	38	M	Low SI	Intermediate to high SI	Homogeneous	+

F, female; FD, fibrous dysplasia; M, male; N/A, not apparent; SI, signal intensity.

although axial bones are more often involved in the polyostotic form [5]. Of the cases examined in this study, nine included patients with polyostotic FD, of whom one had an isolated spinal localisation. The remaining three patients had monostotic spinal manifestations.

FD has been described in each segment of the spine, with the highest prevalence in the lumbar region [11–17]. In the patients included in this study, eight had lumbar spine involvement and seven had thoracic spine involvement. Four patients with polyostotic FD had involvement of the cervical spine. In spinal FD, the vertebral body is affected most frequently, owing to the relatively

large amount of cancellous bone [7]. Because of the close relationship to the vertebral body, involvement of the pedicles has been described in nearly all cases in which the vertebral bodies are affected [12, 15, 17–19]. In this study, lesions were located in the vertebral bodies and posterior elements or sacral bodies and alae of eight patients. However, lesions were located in the posterior elements in one of the eight patients (Patient 1 in Table 2; Figure 3). Isolated vertebral body involvement was present in three patients.

In this study, common and classic radiographic findings in extraspinal FD were not visible on plain radiographs of the spine. A decrease in vertebral body height is a common radiographic finding: when involvement of the vertebral body becomes extensive, vertebral collapse may occur, usually as a consequence of a compression fracture of one or more vertebral plates [20]. However, typical radiographic findings of FD were represented on spine CT scans, with GGO the most common CT finding. In addition, an expansile nature, lytic lesions, sclerotic rims and a decrease in body height were common CT features in our study.

Histopathologically, FD lesions are composed of fibrous tissue containing bone trabeculae, which are composed of woven bone [6, 21]. The ground-glass appearance of FD is due to the presence of many irregular spicules of bone within the fibrous stroma [17]. In our study, more than one lesion showed GGO on CT scans in all of the patients. The amount of woven bone, and the extent to which it is mineralised, will ultimately determine the radiographic density of the lesion. Endosteal scalloping of the adjacent cortex, which may be thinned, may also be noted. An expansile nature representing endosteal scalloping was seen on the CT scans of 11 patients in our study. Ultimately, the affected bone may undergo expansile remodelling secondary to the enlarging mass of fibro-osseous tissue [6]. Cellular fibrous tissue containing irregular bone trabeculae has been demonstrated as an alteration of the normal biomechanical properties of bone [7, 21]. It results in distortion and structural weakness of the bone and abnormal remodelling of the affected bone. Six patients in this study showed contour deformities of the vertebrae due to the markedly expansile nature of the

Figure 2 A 30-year-old female with polyostotic fibrous dysplasia localised in the T8 vertebra and rib. (a) Sagittal T₁ weighted MR image of the T-spine showing a well-defined lesion with low signal intensity and a thin dark signal rim in the T8 vertebral body. (b) Corresponding sagittal T₂ weighted MR image showing the lesion, which is represented with heterogeneous signal intensity and chemical shift artefact in the vertical margin.

Figure 3 A 51-year-old male with polyostotic fibrous dysplasia (FD). A homogeneous sclerotic lesion with expansile nature is represented in the right lamina of the T7 vertebra on a chest CT scan at the mediastinal level. Rib involvement of the FD is noted.

lesions. In addition, FD may be surrounded by a layer of thick, sclerotic reactive bone. This sclerotic margin can be of variable thickness and may be interrupted or incomplete. Nine patients in our study had more than one lesion with sclerotic margins and a variable pattern and thickness. In summary, on CT scans, the features of spinal FD were characterised by medullary expansion, a ground-glass matrix and a variable degree and pattern of sclerosis. Chow et al [11] suggested that, radiologically, the features of monostotic FD of the spine are similar to extraspinal lesions. We agree with previous reports that the CT features of spinal FD are similar to extraspinal lesions.

FD did not consistently show hypointense signal intensities on T_1 and T_2 weighted MR images, although most radiologists are of the opinion that FD is likely to show decreased signal intensity within the lesion owing to the fibrous tissue [22, 23]. Gogia et al [5] stated that the MRI characteristics of FD are variable, typically showing a signal intensity that is intermediate to low on T_1 weighted images and intermediate to high on T_2 weighted images. In our study, MRI findings of spinal FD were non-specific. MRI of the spine revealed various signal characteristics on T_1 and T_2 weighted images, with varying degrees of enhancement patterns after gadolinium administration. According to Jee et al [24], the signal intensities are variable according to the degree of bony trabeculae, cellularity, collagen fibres, cystic changes and haemorrhage within the lesions on T_2 weighted images. Also, FD may be surrounded by a layer of thick, sclerotic reactive bone, called a rind [21]. The rind is seen as a peripheral black rim on T_1 and T_2 weighted images [24]. Seven of the nine patients who had MRI scans had dark signal rims surrounding the lesions in our study.

The diagnosis of FD is difficult, especially in adult patients. Based on imaging findings and location, the differential diagnosis includes metastasis, multiple

myeloma, osteoblastoma and chronic infectious spondylitis. In cases of FD with lytic lesions, the differential diagnosis should include haemangioma, giant cell tumour and aneurysmal bone cyst; in cases of FD with blastic lesions, it should include Paget's disease and osteoblastoma [12]. Spinal FD tends to show an expansile nature, GGO and a relatively well-defined lytic lesion with a sclerotic rim or contour deformity. Osteolytic metastasis rarely presents as expansile vertebral lesions because it is invariably associated with vertebral collapse [25]. Metastasis usually shows an ill-defined edge with a wide zone of transition, which implies rapid growth. The vertebral body is more frequently involved in multiple myeloma [25]. Only rarely does multiple myeloma result in vertebral expansion, and vertebral collapse is more

Figure 4 A 47-year-old male with polyostotic fibrous dysplasia mimicking chronic infectious spondylitis. (a) Sagittal T_1 weighted MR image of the T-spine showing irregular bony fusion of T7, T8 and T9 vertebral bodies with a decrease in body height and obliteration of discs. Heterogeneous low signal intensities are noted in the spinous processes of corresponding levels. (b) Fat-suppressed contrast-enhanced sagittal T_1 weighted MR image showing the lesions, which are represented with a mild, peripheral enhancement pattern. (c) Chest CT of the mediastinum showing bony fusion of costovertebral junction with bony expansion and ground-glass opacity.

commonly seen. The rare sclerotic myeloma may also cause vertebral expansion. Osteoblastoma originates in the neural arch and often extends into the vertebral body. On CT, the lesion shows areas of mineralisation, expansile bone remodelling and sclerosis or a thin osseous shell around its margin. Mineralisation within an osteoblastoma may have the radiological appearance (rings and arcs) of a chondroid matrix. Infectious spondylitis generally shows destructive changes of two adjacent vertebral bodies and the intervening disc. Only reparative changes are seen as reactive sclerosis of the vertebral body, resulting in an ivory vertebra [25–28]. One patient in our study (Patient 4 in Table 2) had MRI findings mimicking chronic infectious spondylitis. However, in this case, bony expansions of posterior elements and posterior ribs with GGO were detected on CT scan (Figure 4).

The present study had several limitations. The first was the small number of cases examined; therefore, further studies with a larger number of cases are necessary. The second was that MRI was not performed in three patients and contrast enhancement was not performed in any patients owing to the inherent nature of the retrospective study. As a result, there were missing MRI analyses. The third limitation was that biopsies were performed in some patients in bony locations other than the spine, but the biopsy involved the spine lesions in most patients.

In conclusion, FD should be included in the differential diagnoses of patients with expansile osteolytic lesions with GGO, sclerotic margins or lesions in the vertebral bodies and posterior elements, especially when seen on spine CT scans. Moreover, these characteristic CT imaging findings of spinal FD may be helpful for diagnosing FD on spine CTs and preventing unnecessary procedures, especially in adult patients.

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