McCune-Albright Syndrome with Pathologic Compression Fracture of the Vertebrae: Report of a Case with Imaging Manifestations

DA-CHUNG LIOU  CHIAO-YI LU

Department of Radiology, Military Tsoting Hospital

We describe an 18 year-old man with unusually extensive polyostotic fibrous dysplasia, which associated with scoliosis of the thoracic spine and pathologic compression fractures of T8-9, L1, and L3. Because of extensive involvement of the spine, thoracic costae, left pelvic bones and femur as well as endocrine dysfunction and skin pigmentation, he was admitted for surgery. Plain radiographs and computed tomographic scans showed expansile osteolytic lesions of the bilateral thoracic costae.

These lesions presented with intermediate signal intensity on T1-weighted magnetic resonance images, hyperintensity on T2-weighted images, and faint enhancement on gadolinium-enhanced T1-weighted images. These clinical and imaging findings may be useful in the diagnosis of McCune-Albright syndrome.

Key word: Bone, Fibrous dysplasia; McCune-Albright syndrome; Spine, fractures

Fibrous dysplasia occurs in either a monostotic form or a polyostotic form. The polyostotic form of the disease is characterized by predominantly unilateral involvement and can involve the whole body. This condition can be associated with skin pigmentation (café-au-lait spots) and endocrine abnormalities leading to accelerated maturation and sexual precocity. This form is called as McCune-Albright syndrome [1].

We present an interesting case of McCune-Albright syndrome in which an 18-year-old patient had extensive polyostotic fibrous dysplasia involving his whole body. The patient presented with scoliosis of the thoracic spine and pathologic compression fractures of T8-9, L1, and L3.

CASE REPORT

This 18-year-old man was referred to our hospital for surgery because of back pain that occurred intermittently for more than 5 years. In addition, he was ashamed of his body configuration, which was affected by severe scoliosis of the thoracic spine with pathologic compression fractures of T8-9, L1, and L3. The patient had a history of a pathologic fracture of his left femur when he was 13 years old. He underwent an operation and bone biopsy at Taipei Veterans General Hospital, where McCune-Albright syndrome was diagnosed.

On his present admission, the patient received a series of radiographic examinations, including radiography of the chest and kidneys, ureters and bladder, computed tomography (CT) of the chest with three-dimensional (3D) reconstruction of thoracic costae and magnetic resonance imaging (MRI) of the whole spines. Chest images showed scoliosis of the thoracic spine, deformity of the thoracic cage, and expansile costae on both sides (Fig. 1a). Chest CT images showed osteolytic changes in the medulla of bilateral costae, in which the soft tissues had been replaced (Fig. 1b). The 3D-reconstructed images showed multi-
locular, well defined, expansile bony lesions of the bilateral costae (Fig. 1c). Lastly, T1-weighted images of MRI of thoracic and lumbar spines showed intermediate signal intensity (Fig. 1d) and high signal intensity on T2-weighted images (Fig. 1e). Fat-saturated, gadolinium-enhanced T1-weighted images depicted heterogeneous enhancement (Fig. 1f). The images also demonstrated involvement of the cervical spines, bilateral costae, left sacrum, and iliac bones. The T8-9, L1, and L3 bodies also had pathologic com-

**Figure 1.** a. The chest film showed scoliosis of T spine (black arrowhead) and bilateral expansile costae (black arrows). b. Noncontrast CT image showed osteolytic change of medullas of bilateral costae with soft tissue replacement (black arrows). c. Chest CT with 3D-reconstruction showed multilocular, well-defined, expansile bony lesions of the bilateral costae (white arrows). Scoliosis of T spine and pathologic compression fractures of T8,9 are also noted (black arrowheads). d. MRI. Sagittal T1-weighted image (544/10, TR/TE) of the L spine without gadolinium-DTPA administration showed intermediate signal intensity on T9,12 and L1-5 bodies and pathologic compression fractures of L1,3 bodies (white arrowheads). e. MRI. Coronal T2-weighted image (4000/92, TR/TE) of the reformatted T-L spine without gadolinium-DTPA administration showed high signal intensity on T-L spines and bilateral costae (white arrows). f. MRI. Coronal T1-weighted image (615/11, TR/TE) of the T spine with gadolinium-DTPA administration and fat saturation showed marginal enhancement on the T6 body (white arrowhead) and faint enhancement on right side costae (white arrow).
pression fractures (Fig. 1d, 1e). In addition, the patient had skin pigmentation (café-au-lait spots) on his right side trunk and an deformed enlargement of his head (Fig. 1g, 1h). All of these findings revealed the characteristics of McCune-Albright syndrome in this patient.

DISCUSSION

Fibrous dysplasia is a disorder of the bony structures characterized by fibrous replacement of normal medullary bone due to dysfunctional bone-forming elements [1, 2, 3]. This disorder accounts for approximately 7% of all benign bone lesions [2]. Fibrous dysplasia commonly affects a single segment (monostotic form, 85%) and less commonly results in widespread, diffuse and generalized skeletal involvement (polyostotic form, 15%) [2, 3, 4, 5]. The polyostotic form of the condition is associated with café-au-lait spots and endocrine abnormalities leading to accelerated maturation and sexual precocity, which is known as McCune-Albright syndrome [1].

Cranial and mandibular involvement is seen in most patients, though vertebral lesions are not typical [1, 2, 5]. The frequency of involvement of the cervical vertebra and the lumbar vertebra, as part of polyostotic fibrous dysplasia, has been reported to be 7% and 14%, respectively [2]. Such involvement may result in progressive debilitation and deformity. Polyostotic fibrous dysplasia usually manifests with unilateral predominance, and frequently with bony deformity. We report this case because the disease extensively involved the patient’s skull, bilateral costae, entire spine, and left hemipelvis, resulting in scoliosis and pathologic compression fractures of T8-9, L1, and L3 vertebral bodies.

Radiographic findings of fibrous dysplasia are complex and generally classified into the six types: (1) peau d’orange, (2) whorled plaquelike, (3) diffuse sclerotic, (4) cystlike with multilocular or unilocular, (5) pagetoid, and (6) chalky [6]. In these six types, the most common presentation includes multilocular, radiolucent lesions or a ground-glass appearance with bony expansion and a well-defined border. Because of its specific radiographic findings, fibrous dysplasia is frequently diagnosed on conventional radiography. CT images show expansile osteolytic lesions containing soft tissue, a well-defined cortex and faint heterogeneous enhancement after the administration of contrast material [5]. MRI findings vary, depending on the fibrous, cartilaginous and sometimes hemorrhagic components of fibrous dysplasia. T1-weighted images most often depict intermediate signal intensity; whereas T2-weighted images show low signal intensity for predominantly fibrous lesions, high signal intensity for highly cartilaginous and proteinaceous lesions and mixed signal intensity for lesions with a combination of these characteristics. The lesions show heterogeneous or peripheral rim enhancement after the administration of a gadolinium-based contrast agent [1, 2, 4].

Malignant transformation is a distinctly unusual complication and has been estimated to occur in less than 1% of all cases [3]. Malignancies occur predominantly in lesions of the femur and mandible. The most common malignant tumor is osteosarcoma, follow by
fibrosarcoma-like tumors [3]. The radiographic differential diagnosis for fibrous dysplasia includes hemangioma, Paget’s disease, aneurysmal bone cyst, neurofibromatosis, giant cell tumor and expansile metastases [2, 7]. Plain radiographs of hemangiomas show honeycomb vertebrae and a mottled pattern of increased signal intensity on T1- and T2-weighted MRI images. The incidence of these lesions is peaked between the fourth and fifth decades of life. Paget’s disease causes a bone-within-bone appearance and “ivory vertebra” on plain spinal images and hypointense area on T1- and T2-weighted MRI images. It most frequently occurs in older age (>85 years). Aneurysmal bone cysts presents with a soap-bubble pattern and lytic eccentric radiolucency on plain radiographs. These cysts progress rapidly within 6 weeks to 3 months. Neurofibromatosis showed twisted ribbon-like ribs, enlargement of the neural foramen with a dumbbell neurofibroma of the spinal nerve. Involved areas are slightly hyperintense on T1-weighted MRIs with a hyperintense periphery and hypointense cord on T2-weighted images. Giant cell tumors are solitary masses or of multiple foci in rare case. They are mostly located in the meta-epiphysis region of long bones. Lastly, metastasis was the last consideration because it has a variety of imaging presentations and because our patient did not have any primary malignant disease.

In summary, we present a case of McCune-Albright syndrome involving the skin, spines, costae, and hemipelvis with complication of scoliosis and pathologic compression fractures of T8-9, L1, and L3. We use CT with 3D-reconstruction and MRI in this case because of their high resolution, high-quality depiction of the foci and because they can be used to survey lesions in the whole body. To our knowledge, a case of McCune-Albright syndrome with this unusually severe and extensive involvement, in the absence of significant recent or remote trauma and requiring decompression and surgical fusion, has not been reported previously.

REFERENCES:

McCune-Albright症候群合併胸腰椎病理壓迫性骨折之各種影像之特性：病例報告

劉大忠 陸教義
國軍左營醫院 放射診斷科

McCune-Albright症候群是一個廣泛多發性的纖維發育異常，合併皮膚的咖啡斑和內分泌異常導致性早熟的總稱。它好發單側並且在影像學上有特殊的表現。我們的病例是一個18歲的男性，由於侵犯到頭顱、兩側肋骨、全身脊椎、左側骨盆、合併脊椎側彎和病理壓迫性骨折，加上身上的斑點和性早熟的外觀，而被證實是McCune-Albright症候群。由於是少見的先天性異常疾病，因此我們作了一系列不同的影像來評估這種疾病。

關鍵詞：纖維發育異常；McCune-Albright症候群；脊椎、骨折