ed a local clinic and was referred to our hospital for evaluation of a pathologic fracture in C4. There was no history of trauma. The physical examination showed slight deformities in his left extremities. There were no skin lesions, such as café-au-lait patches. In spite of the pathologic fracture, there was no neck pain or range of motion limitations. The neurologic examination showed tingling in both extremities and motor weakness of the left upper extremity (grade IV). Spurling and Hoffman’s signs were negative bilaterally.

The white blood cell count, electrolytes, liver and thyroid function tests, and serum concentrations of parathyroid hormone, 25-hydroxyvitamin D, serum calcium, and phosphate were within normal limits.

Radiology

Cervical spine radiographs showed that the C4 vertebral body was partially collapsed. Computed tomography (CT) demonstrated multiple lytic lesions involving the odontoid process of C2 and the body, left pedicle, and posterior elements of C4. Combined anterior and posterior decompression and reconstruction were performed. Post-operatively, the histopathologic examination confirmed FD. On the post-operative follow-up examination, the neurologic deficits had completely resolved.

Key Words: Fibrous dysplasia · Cervical spine · Elderly.

INTRODUCTION

Fibrous dysplasia (FD), first described in 1983 by Lichtenstein[8], is an uncommon benign fibro-osseous abnormality of the bone with an unknown etiology. The disorder involves a single bone (monostotic) or multiple bones (polyostotic)[4]. Polyostotic FD may be combined with cutaneous and/or endocrine abnormalities (McCune-Albright syndrome)[1]. The most common sites of the diseases are the long bones, craniofacial bones, ribs, and pelvis, whereas vertebral involvement is rare[4]. FD involving the cervical spine has rarely been reported[2,3,5,9-14]. Moreover, cervical FD presenting as a pathologic fracture in older patients is extremely rare[12]. We report a rare case of symptomatic cervical FD associated with a pathologic fracture in a 63-year-old man.

CASE REPORT

History and examination

A 63-year-old man presented with progressive weakness of the left arm and pain in the shoulder and arm. The symptoms had worsened over a period of 6 months. One week prior to admission, the weakness in the left hand was aggravated. He visit-
normal uptake in the sternum, multiple spines, left scapula, extremities, ribs, and pelvis (Fig. 3). A bone mineral density was normal (Fig. 4). Therefore, osteoporotic compression fracture was excluded. Based on the radiologic findings, the lesions were thought to be polyostotic FD of the cervical spine causing a pathologic fracture.

**Operation**

An anterior approach to C4 was performed. The involved bone was soft and white. Corpectomy of C4 was performed and the involved bone was easily removed with curettage and rongeurs. Because the entire iliac bone was also involved with FD, a human allograft block was used for reconstruction of the corpectomy site. Additional internal fixation was performed by a plate and screws at C3-5. Then, the patient was turned to the prone position for posterior stabilization. Posterior reinforcement was performed because FD partially involved the C3 and C4 vertebral bodies and posterior elements of C4; an allograft strut was used. A left posterior foraminotomy was performed for foraminal stenosis due to the hypertrophied lateral mass. An additional lateral mass screwing and fusion with allograft were performed at C3-4-5 (Fig. 5). The histopathologic examination demonstrated fibrous tissue within medullary bone and irregularly-shaped bony trabeculae embedded in the fibrous stroma, which was consistent with FD (Fig. 6).

**Post-operative course**

There were no post-operative complications. The patient’s pain, tingling, and weakness improved rapidly. After 7 days, the patient was discharged wearing a Philadelphia collar. Eight months postoperatively, the graft fusion was confirmed without FD invasion.

**DISCUSSION**

FD is a benign intramedullary fibro-osseous lesion, represent-
Cervical Fibrous Dysplasia Presenting as a Pathologic Fracture | SH Lee, et al.

The adjacent disc height is frequently decreased. A CT scan is the best diagnostic tool to demonstrate the radiographic characteristics of FD. A CT scan confirms decreased cancellous bone and cortical thinning, ballooning, or collapse of vertebrae. MRI is a sensitive tool to demonstrate the extent of tumor involvement, extent of neural compression, and differential diagnosis. FD appears as a homogeneous hypo-intense lesion on T1-weighted imaging unless there is a pathologic fracture. On T2-weighted imaging, the lesion is heterogeneous depending on the amount of fibro-osseous tissue, cellularity, cystic alterations, hemorrhage, and cartilaginous tissue. FD is homogeneously enhanced after intravenous gadolinium. The differential diagnosis of FD includes hemangiomas, giant cell tumors, aneurysmal bone cysts, Paget disease, and osteoblastomas. In older patients, multiple myeloma and metastatic carcinoma should also be considered.

In most cases of FD involving the non-axial skeleton, conservative management is recommended, even following pathologic fractures. The healing potential for all fracture sites has been reported to be 94%7. In vertebral FD, conservative management has also been recommended, in particular with polyostotic FD. Two cases of odontoid fractures (type II) complicating polyostotic FD, which were successfully managed with braces, have been reported11,13. However, surgical treatment is indicated for persistent pain, neurologic deficits, vertebral collapse, instability, and/or cord compression. In previous reports, different surgical methods have been performed, such as laminectomies, corpectomies, local curettage, and arthrodesis12,14. Due to the different expansion properties of FD, there is no gold standard for operative treatment. When FD is restricted to the anterior or posterior element, one approach may be sufficient. However, vertebral FD commonly involves both anterior and posterior elements, indicating approximately 2.5% of all bone neoplasms and 7% of benign neoplasms6. FD can present in a single bone (monostotic) or multiple bones (polyostotic). Monostotic FD is more frequent (70%) than polyostotic FD (27%). McCune-Albright syndrome (3%) is a type of FD in which polyostotic lesions are accompanied by café-au-lait spots, precocious sexual development, and other endocrine abnormalities. This syndrome appears to occur more frequently in females4,6.

The most common sites for FD include the ribs, femur, tibia, and humerus. Spinal involvement is unusual and occurs in 7-24% of patients with polyostotic FD, but is rare with monostotic FD4,12,13. Cervical involvement with FD is rare and approximately 30 cases of cervical FD have been reported, with 12 cases of monostotic FD13. The disease can affect every level and location of the cervical spine, and occurs more often in males than in females12. Most patients are young adults, ranging in age from 11-56 years13,14. In FD, dysplastic areas rarely expand after puberty because FD usually tends to be exhausted after cessation of bone growth12. Therefore, most pathologic fractures in FD occur in weight-bearing long bone at a young age4.

Like our case, cervical FDs presenting as a pathologic fracture are extremely rare in older patients. The possible explanation may be that unlike monostotic FD, polyostotic lesions often continue to enlarge after skeletal maturity, with an increase in pathologic fracture4.

The majority of patients with FD are asymptomatic and the diagnosis is incidental. The most common symptom of FD is localized pain4,11. In some patients, FD is first diagnosed at the time of a pathologic fracture11,13. Myelopathy or radicular sensory and/or motor loss as in our case has rarely been reported. Neurologic impairment due to cervical FD has been reported in only 6 cases8.

Radiologically, FD typically exhibits ‘ground glass’ radiolucency, reflecting random woven bone formation4. The adjacent disc height is frequently decreased. A CT scan is the best diagnostic tool to demonstrate the radiographic characteristics of FD. A CT scan confirms decreased cancellous bone and cortical thinning, ballooning, or collapse of vertebrae. MRI is a sensitive tool to demonstrate the extent of tumor involvement, extent of neural compression, and differential diagnosis. FD appears as a homogeneous hypo-intense lesion on T1-weighted imaging unless there is a pathologic fracture. On T2-weighted imaging, the lesion is heterogeneous depending on the amount of fibro-osseous tissue, cellularity, cystic alterations, hemorrhage, and cartilaginous tissue. FD is homogeneously enhanced after intravenous gadolinium. The differential diagnosis of FD includes hemangiomas, giant cell tumors, aneurysmal bone cysts, Paget disease, and osteoblastomas. In older patients, multiple myeloma and metastatic carcinoma should also be considered4,8,11.

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elements, and a combined anterior and posterior approach may be needed, as in our case. In our case, there was potential instability resulting from vertebral collapse and a pathologic fracture. Neurologic deficits also existed due to the collapsed vertebral body and the hypertrophied lateral mass on the left side. Therefore, an anterior corpectomy with anterior plating and an additional posterior laminoforaminotomy with posterior reinforcement were performed. In our case, posterior instrumentation was sufficient; however, we reasoned that anterior instrumentation was not appropriate due to partial involvement of the C3 and C4 vertebral bodies, the use of an allograft with a low fusion rate, and the posterior laminoforaminotomy. Five cases of combined anterior and posterior approaches have been reported in patients with cervical FD. In all cases, a staged combined approach was performed during a 1 week to 10 month interval. In the current case, a single-stage combined approach was performed. There were no complications associated with the surgery. In patients who are poor surgical candidates, percutaneous vertebroplasty may be used as an alternative for symptomatic FD associated with pathologic fractures.

CONCLUSION

We report a case of symptomatic cervical FD associated with a pathologic fracture in a 63-year-old man. Cervical FD presenting as a pathologic fracture is extremely rare in older patients. The current case is the first report of cervical FD presenting as a pathologic fracture in the 7th decade of life. Although most pathologic fractures in FD occur in weight-bearing long bone at a young age, it could also occur in older patients with cervical FD.

References