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Eosinophilic Granuloma in a Patient with Marked Improvements of Social Activity as well as Clinical Manifestations

Takashi Isobe¹, Osamu Kataoka², Masatoshi Sumi² and Masanori Tsukuda²

A twenty-year-old male started to have low-back pain. On X-ray survey, unusual shadows of the right humerus and sacrum were found. An open biopsy confirmed a histological diagnosis of eosinophilic granuloma in the 5th lumbar spine and sacrum, followed by the second biopsy of the same diagnosis on the right humerus. A local radiation therapy was performed with 2 Gy for 3 times on the right humerus, along with low-dose continuous cyclophosphamide administration for about 17 months. As to pain and performance status (PS) of the patient, there are complete disappearance of pain and a marked improvement of PS, producing a great success in the therapy and an excellent social life i.e. a good quality of life (QOL) in the present patient.

Key Words
Eosinophilic granuloma, Local radiation therapy, Cyclophosphamide administration, Quality of life (QOL).

INTRODUCTION

Eosinophilic granulomatosis has been known to have the most common presentation of a single osteolytic lesions in a long or flat bone (1). In addition to pain and swelling over the affected area as common symptoms, there could be pathological fracture during the clinical course, producing a situation of disturbed daily life of patients. The present paper describes a case report of 20 year old male with a successful recovery from fractured

CASE PRESENTATION

Case H.F., 20-year-old male, started to have low-back pain, which was accentuated by walking in association of vague dysesthesia in the bilateral lower extremities on April 1990. He occasionally had lower abdominal pain and mictions pain suggesting of acute cystitis. On admission at the Department of Orthopedic Surgery at Kobe National Hospital on January 1992, he was found to be moderately-nourished and had a slight difficulty of walking. Physical examination revealed no eruptions on the skin, no lymphadenopathy, and no abnormality on the heart and lung. There was no hepato-splenomegaly or no abdominal mass. There was a
Figure 1. Osteolytic lesions in the upper part of the right humerus on X ray films of March 16, 1992 (left), July 14, 1992 (middle) and August 4, 1992 (right).

Sign of sciatic pain bilaterally with positive straight leg raising (SLR) test. Other neurological examination including deep tendon reflexes, pathologic reflexes, and sensory tests showed no abnormality.

Laboratory examinations on admission showed his blood type A, a serum total protein of 6.4 g/dl, albumin 3.5 g/dl, A/G ratio of 1.2, serum IgG 1580 mg/dl, IgA 335 mg/dl, IgM 200 mg/dl, and no M-protein in the serum or urine was confirmed by immunoelectrophoresis. There were plasma fibrinogen 343 mg/dl, serum total bilirubin 0.4 mg/dl, GOT 9 IU/l, GPT 38 IU/l, LDH123 IU/l, alkaline phosphatase 98 IU/l, $\gamma$-GTP 40 IU/l, CPK 113 IU/l, uric acid 3.4 mg/dl, glucose 80 mg/dl, total cholesterol 128mg/dl, $\beta$-lipoprotein 381 mg/dl, HBs-antigen negative, HBs-antibody negative, serologic tests for syphilis negative, CEA 1.4 ng/ml,
Eosinophilic granuloma

Figure 2a. X-ray films of lateral view of the lumbar spines and the sacrum, with abnormal lesions with arrows indicated of January 6, 1992 (left) and December 7, 1992 (right).

Figure 2b. CT scannings of the lumbar spines and the involved sacrum on January 24, 1992.

alpha-feto protein 1 mg/dl, sodium 138 mEq/l, potassium 3.7 mEq/l, chloride 107 mEq/l, calcium 8.5 mg/dl, and phosphorus 3.2 mg/dl. A complete blood counts included a white blood cell 10,300/µl with segmented 73%, lymphocyte 23%, monocyte 3% and basophil 1%, a red blood cell 484 X 10^4/µl, hemoglobin 14.2 mg/dl, hematocrit 40.8% and a platelet count 26.7 X 10^4/µl. In short summary, this patient did not have any abnormal levels of laboratory tests.

On X-ray survey after admission, mild osteolytic lesions in the upper part of the right humerus, and unusual shadow in the sacrum were noted (Figure 1). The presence of bony destructions were confirmed by
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X-ray tomography and computed tomography (Figure 2). Thus, an open biopsy for the spine was performed around the 5th lumbar spine (L5) and sacrum (S2), with cleaning up the involved lesions by curettage. The histological diagnosis of the biopsied specimen was eosinophilic granuloma. Furthermore, on March 3rd, the second surgery was carried out for tumor resection of 5th lumbar spine and sacrum with iliac bone graft, followed by an application of body cast postoperatively. Meanwhile, the patient was found to have a high risk of bone fracture on X-ray of the right humerus, which prompted the orthopaedic surgeon to operate the lesion. On April 9th of 1992, he had the third surgery of right humerus with curettage, and the biopsied specimen identified the histological diagnosis of eosinophilic granuloma. Both biopsied materials showed similar findings of an infiltrate with foamy macrophages and admixed eosinophils, in association of lymphocytes, giant cells, proliferated osteoclast, and neutrophils, with no necrosis or no malignant cells. Cast on the operated right upper arm was applied. Postoperatively, a local radiation therapy was performed with 2 Gy for 3 times within a week, in a field of 20 X 5.5 cm with a depth of 4 cm of the right humerus by local irradiation. An additional bone lesion was found in the 5th cervical spine (C5) on X-ray (Figure 3) and CT scans, which was also confirmed to be the involvement of eosinophilic granuloma, derived from the bone, in the 5th cervical spine on an open biopsy. Histologically, this biopsied material demonstrated eosinophilic granuloma in association with plasma cells and macrophages (Figure 4). Therefore, an alkylating of cyclophosphamide was instituted orally; first with 50 mg a day with prednisolone 20 mg a day starting on July 14, 1992 for one month, then switched to a dose of cyclophosphamide 100 mg and prednisolone 10 mg for another 6 months, followed by reduced maintenance dose of cyclophosphamide 50 mg with predni-
solone 5 mg daily for subsequent 10 months until November of 1993. The institution of low-dose continuous cyclophosphamide appeared to achieve a good result to control the patient’s condition, i.e. an improvement of performance status from 4 initially to 0, with obtaining a moderate physical activity and fine social life (to go to school as a college student) after discharge from the hospital, without any cast. There was also no abnormality in laboratory data during and after combined treatments including operations three times, local irradiation 3 times and low-dose continuous administration of cyclophosphamide with or without prednisolone.

**DISCUSSION**

Eosinophilic granuloma is a localized form of distinctive histiocytosis, or Langerhans’ cell histiocytosis (1-5). It is a benign form and has a benign lesion of proliferating Langerhans’ cells. It is not severe clinically among histiocytosis syndromes and is characterized by lytic lesions of one or more bones. However, as in the present case of 20 year-old young boy, some cases develop clinically severe form or complicated situation. Because of unknown origin, the disease extended beyond its original boundaries and resulted in complete destruction of bone, or alternatively, has multifocal granulomatous lesions.
Thus, early detection of the bone lesion is certainly necessary in the diagnosis and treatment of this disease. For this purpose, X-ray morphology in a detailed study and recent advance of MR (magnetic resonance) imaging have clinically contributed. As to the X-ray morphology, there need careful observations for differentiation from Ewing's sarcoma, osteomyelitis, metastasis of carcinoma, lymphoma and multiple myeloma. In this regard, MRI study could achieve a favorable result to detect the early detection (6-8). In one study (7) of 11 patients with histologically proven eosinophilic granuloma, a marked enhancement were found on MRI by intermediate to high signal intensity on T1-weighting and high signal intensity on T2-weighting, in contrast with appearance on conventional radiography and CT. MRI was superior to other imaging methods in demonstrating bone marrow involvement and any accompanying soft tissue mass or inflammation. Another study (8) of 16 cases described that the most common MR appearance (10 cases) was a focal lesion, surrounded by an extensive, ill-defined bone marrow and soft tissue reaction with low signal intensity on T2-weighted images, considered to represent bone marrow and soft tissue edema (the flare phenomenon).

REFERENCES