

It is important for pediatricians to consider molecular testing of *UBR1* gene not only for the confirmation of diagnosis in the affected child but also for confirming carrier status in both parents and to offer appropriate counseling to the family.

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Primary Vertebral Lymphoma Presenting with Fracture

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We report a 15-year-old girl admitted with back pain and multifocal osteolytic lesions without systemic symptoms at T7, L5, and S1 spinal vertebrae. The child was diagnosed as having primary multifocal osseous lymphoma, in which multiple bones are involved in the absence of lymph node or visceral disease for at least 6 months following initial presentation.

Key words: Bone, Lymphoma, Vertebra.

Primarily lymphoma of bone occurs rarely in children and accounts nearly 2.8 to 5.9 percent of Non-Hodgkin lymphomas [1,2]. The incidence of a single vertebral lesion is reported to be 1.7% of all primary lymphoma of bones [3]. Most of the involved bones are long bones of the extremity, like femur [1]. The disease may resemble fracture, trauma and mimic inflammatory, neuropathic, and infectious conditions with these symptoms [4,5].

CASE REPORT

A 15-year-old girl was admitted with back pain. On physical examination, there was tenderness on thoraco-

lumbar vertebrae. There was no history of trauma. Lymphadenopathy, mass and organomegaly were not detected. Laboratory data were as follows: Hb: 12g/dL, WBC: 6500/mm³, Platelet: 300000/mm³, sedimentation: 14 mm/h, LDH: 146 U/L, renal and liver function tests were normal. Thoracic vertebra X-ray showed lytic lesions on T7 vertebrae. Thoracic computed tomography (CT) showed reduced T7 vertebral corpus height, and lytic, hypodense areas in the L5 and S1 vertebrae. 18F-Fluorodeoxyglucose positron emission tomography (18F-FDG-PET) revealed increased activity on vertebral corpus of T7, T11 and L4 vertebra and normal lungs. Bone marrow aspiration and biopsy were normal. Pathologic



FIG. 1. T1 weighted sagittal image of the thoracic spine shows wedge shaped compression fracture of the thoracic 7 vertebra body. Spinal canal calibration is normal.

examination of the bone biopsy from T7 vertebra revealed high-grade B-cell lymphoma. The patient was diagnosed as primary bone lymphoma and LMB-89 chemotherapy treatment was started. Three months later, magnetic resonance imaging (MRI) showed heterogeneous hyper intense lesions on the right side of sacrum, right iliac bone and acetabular roof and left femoral neck, which were assessed as necrotic lesions. 18-F-FDG-PET examination revealed increased FDG uptake on right sacroiliac joint and sacrum; however, left side had normal FDG uptake. Six months later, significant improvement was detected on PET. The treatment was stopped 9 months later with no active lesion on bones. The patient is now in remission for 66 months.

DISCUSSION

Primary bone lymphoma is a rare disease occurs primarily in the bone without an involvement of any other site in the body. The most involved bones are femur, tibia, mandible, mastoid, maxilla, zygomatic arch, rib, clavicle, vertebrae, scapula, ulna, talus and calcaneous [6,7].

The most common presenting complaints are pain, swelling, mass, fever, weight loss, night-pain, limp, irritability, pathologic fracture, and neurologic symptoms [7]. The initial and only symptom in our case was back pain due to vertebral fracture. Thus, in patients like our case with limited symptoms; it is difficult to make differential

diagnosis. The mean delay from the onset of symptoms until the final diagnosis was reported as 6.2 months (range, 0 to 2.5 years) [7]. The causes of delay were most often nonspecific initial presentation like nonspecific pain and/or swelling which can be attributed as musculoskeletal pain, such as muscle strain or synovitis. Difficulty in interpretation of the histological findings is a less commonly reported reason of delay [7]. Our patient was diagnosed 6 months after the pain began.

Pediatric primary bone lymphoma consists of large cell lymphoma, lymphoblastic lymphoma, small, noncleaved-cell lymphoma, and unclassified [6]. Pediatric diffuse large cell lymphoma has a favorable prognosis from others [8]. Back pain and vertebral fracture are the two complaints that can be commonly seen in children with trauma, arthritis, and infections. This can lead serious delay in diagnose. Unresolved pain and fracture despite analgesic treatment may be a good pointer to the possibility of a lymphoma.

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