

Detection of Unknown sites of multiple enchondroma (Ollier's Disease) mimicking like metastasis using bone scintigraphy

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ABSTRACT

Ollier's disease characterized by multiple skeletal enchondroma is a rare noninherited disease of unknown etiology. Majority of the skeletal enchondroma are present in the metaphyses and diaphysis of tubular limb bones. Ollier's disease has a predilection for unilateral distribution. Malignant changes in Ollier's disease may occur in adult patients. Radionuclide bone scanning is one method used to assess lesions depicted on radiographs or magnetic resonance images that are presumed to be enchondromas. Furthermore, a bone scan may give a clue to the multifocality of the disease. We report a case of right first phalangeal enchondroma in a 23-year-old male, who underwent bone scintigraphy detected multifocal asymmetric right side involvement of radius, humerus, femur, and tibia which confirm a diagnosis of Ollier's disease.

Key words: Bone scintigraphy, enchondroma, metastasis, Ollier's disease

INTRODUCTION

Enchondroma is a common benign tumor originating in the medullary cavity of bone and is composed chiefly of hyaline cartilage. It is believed that enchondroma arises from misplaced cartilage rests from the growth plate and usually begins its development in the metaphyseal region. It may be metabolically active and continue to grow.^[1] There is asymmetric involvement of the limbs, with one side being exclusively or predominantly involved.^[2] The cartilage tumors in enchondromatosis are asymptomatic and are detected as incidental findings. Technetium-99m methylene diphosphonate (MDP) whole body bone scan is a sensitive investigation to ascertain the complete extent of skeletal involvement particularly the asymptomatic sites.^[3]

CASE REPORT

A 23-year-old man, presented with a history of right-hand enchondroma underwent surgery before 7 years. He developed right forearm pain and X-ray showed lytic area without cortical thinning in radius [Figure 1]. He was referred for bone scintigraphy which showed intense tracer uptake in the right radius, right humerus, right femur and right tibia [Figure 2]. Static images of hand shows intense uptake in the right 1st and 2nd metacarpal and 2nd phalanx [Figure 3]. The lesions predominantly involved the tubular limb bones sparing the axial skeleton. Bone scan revealed extra sites of involvement at right humerus, right femoral, and tibial shafts. Bone scan appearance of asymmetrical unilateral involvement was reported to be consistent with multiple enchondroma seen in Ollier's disease.

DISCUSSION

Ollier's disease, a rare nonhereditary disorder characterized by multiple enchondromas with a predilection for unilateral distribution, was initially described by Ollier in 1899.^[3] The characteristic features of the disease are created by

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Figure 1: X-ray radiograph of the forearm showing lytic lesion in radius without cortical thinning

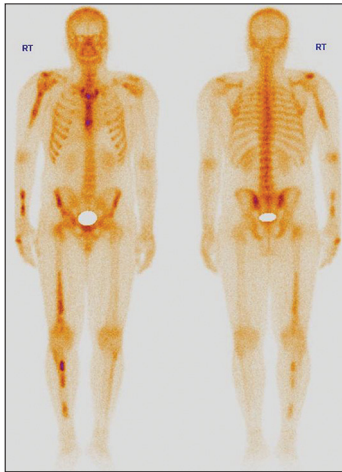


Figure 2: Technetium-99m methylene diphosphonate whole body bone scan showing intense uptake in the right radius, right humerus, right femur, and right tibia, all located asymmetrically in the right side of the body



Figure 3: Static images of hand showing intense uptake in right 1st and 2nd metacarpal and 2nd phalanx

persisting cartilage masses in the metaphyses and diaphysis, which are formed by subperiosteal deposition of cartilage. In fact, enchondromas tend to occupy the diaphyseal region in the short tubular bones and the metaphyseal region in the long bones.^[4]

The pattern of limb involvement is usually asymmetrical, with one side being exclusively or predominantly involved. The disease is usually detected during early childhood.^[5] Notable clinical problems are progressive shortening of the involved extremity, angular deformity, pathological fractures, and malignant transformation in 20–50% of cases.^[6]

The cartilage tumors in enchondromatosis are asymptomatic and are detected as incidental findings. However, the cartilage tumors in enchondromatosis may be numerous and large, producing severe deformities. The radiography features are characteristic, as the unmineralized nodules of the cartilage produce well-circumscribed oval lucencies that are surrounded by a thin rim of radiodense bone (O-ring sign). If the matrix calcifies, it is detected as irregular opacities.^[7]

Radionuclide bone scanning is one method used to assess lesions depicted on radiographs or magnetic resonance images that are presumed to be enchondromas.^[8] Enchondroma can have high fluorodeoxyglucose and MDP uptake on scintigraphic imaging and occasionally may mimic metastatic lesions in patients being screened for metastatic disease.^[9] Furthermore, a bone scan may give a clue to the multifocal nature of the disease. Unilateral increased uptake along with limb length discrepancy, which are distinct features of Ollier disease.^[9,10] It has been emphasized that Ollier's disease usually stops spontaneously with skeletal maturity; therefore, any lesion showing activity or increased uptake after termination of the growth period requires a thorough examination.^[11]

CONCLUSION

Ollier's disease, a rare nonhereditary disorder characterized by multiple enchondromas with a predilection for unilateral distribution. Radionuclide bone scanning is one method used to assess lesions depicted on radiographs or magnetic resonance images that are presumed to be enchondromas. Furthermore, a bone scan may give a clue to the multifocal nature of the disease.

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