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Hypertrophic Pulmonary Osteoarthropathy without clubbing of the digits detected by Tc-99m MDP bone scintigraphy

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ABSTRACT

Isolated hypertrophic pulmonary osteoarthropathy (HPOA) is rarely reported. HPOA typically occurs concurrently with clubbing, periostitis and polyarthritis. We reported a case of pulmonary malignancy in whom HPOA demonstrated in bone scintigraphy.

Keywords: hypertrophic pulmonary osteoarthropathy (HPOA), clubbing, bone scintigraphy

Introduction

Hypertrophic pulmonary osteoarthropathy (HPOA) is a syndrome characterized by coexistence of digital clubbing, periostitis of the tubular bones and polyarthritis. We report a patient with lung malignancy with HPOA demonstrated on bone scintigraphy but without presence of clubbing. There are 2 types of HPOA, primary which is familial form of HPOA and secondary form of HPOA which associated with a wide variety of medical conditions. The periosteal new bone formation of periostitis is clinically manifested as painful limbs. Periostitis can be diagnosed by imaging studies and bone scintigraphy with Technetium 99m Methylene Diphosphonate (MDP) is a sensitive investigation in the detection of HPOA.

Case report

A 56 year-old gentleman, active smoker, presented with a 2 months history of right scapular pain. He experienced mild-to-moderate persistent right scapular pain and later associated with non-productive cough and right upper chest pain. A chest X-ray film showed right upper zone mass. Initial bronchoscopy showed partial left vocal cord palsy and no mass intrabronchially. Endobronchial ultrasound (EBUS) performed and tissue sample came back as adenocarcinoma of right lung. CECT thorax reported as right apical lung mass.

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Radiological investigations led to the diagnosis of right apical lung carcinoma, confirmed to be non-small cell cancer on biopsy. His ECOG/PS is 0 and the main complaint is persistent mild-to-moderate right scapular pain associated with pain over right upper limb and unilateral joints, which he prefers to, immobilize his right upper limb. He received concurrent chemoradiotherapy (daily radiotherapy concurrent with weekly chemotherapy) which completed in 5 weeks' time. CT scan post treatment showed features of treatment responsive disease. He was referred for bone scan to rule out bone metastasis. On examination, he is on right arm sling and obvious muscle dystrophy can be observed. There is no evidence of digital clubbing. Diffuse peri-articular tenderness was present around the right shoulder without clinical synovitis.Bone scintigraphy was undertaken for suspected metastatic disease. This revealed a pattern of uptake characteristic of HPOA. Bone scan demonstrated increased peripheral periosteal and cortical uptake seen in the right humerus, radius and ulna, and bilateral tibia. X-ray of right upper limb showed thinning of cortex of humerus, radius and ulna.

Discussion

Hypertrophic pulmonary osteoarthropathy (HPOA) is a syndrome characterized by coexistence of digital clubbing, periostitis of the tubular bones and polyarthritis [1,2]. The triad has been associated with numerous diseases which categorized into 2 types; primary and secondary. The term pachydermoperiostitis first used by Touraine and co-workers in 1935 [1] to describe the primary form of

HPOA which is a rare familial condition, about 3-5% of patients with HPOA have primary HPOA [4] and more common in males [1, 3]. Secondary HPOA is more common, about 95-97% of cases [4] and associated with malignant and non-malignant conditions. These conditions include primary and secondary lung carcinoma, pulmonary fibrosis and mesothelioma, carcinoma of liver and gut, inflammatory bowel disease, congenital cyanotic heart disease and many other rarer conditions. Prevalence is higher in non-small cell lung carcinoma (NSCLC) compared to small cell lung carcinoma (SCLC) [5].Regardless of the aetiology, clubbing is the commonest manifestation of the syndrome and periostitis is the hallmark of HPOA. The presence of periostitis and arthritis without clubbing can make the diagnosis of an incomplete form of HPOA [6-7]. Lately, an increasing number of cases of HPOA without clubbing have been reported in the medical literature [6-7]. Our case met at least 2 diagnostic criteria. Initially, he had painful right shoulder joint and subsequently painful right upper limb joints. It has been reported that periostitis or HPOA can precede underlying disease by several months [1]. HPOA usually occurs bilaterally regardless of the side of the tumour[3]. Bone scintigraphy is very sensitive in detecting HPOA. The scintigraphic finding characteristics of HPOA are bilateral symmetrical linear uptake of the radiopharmaceutical along the cortical margins of the long bones which is also known as “tram line” or “double stripe” sign. The commonest sites were tibia and fibula, and the second commonest sites were the femur and the radius and ulna. The absence of medullary cavity involvement of the long bones and absence of axial skeletal involvement distinguish HPOA from metastatic involvement of the bones[4].The underlying pathogenesis of HPOA remains unclear and many theories has been proposed. In the megakaryocyte or platelet clump hypothesis, it is postulated that there are increased platelet derived growth factors PDGF, vascular endothelial growth factor (VEGF) and growth hormone[8]. Others are humoral theories include a possible role for steroids, cytokines, growth factors and autonomic dysfunction [7].

Conclusion

Our case demonstrates the unusual finding of HPOA in the absence of digital clubbing in patient with non-small cell lung carcinoma. He is diagnosed with HPOA after findings on bone scintigraphy and X-ray in association with his main complaint of right upper limb diffuse pain and right shoulder joint pain.

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