Case Report

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Paget'S Disease of Bone; A Case Report

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ABSTRACT

BACKGROUND AND OBJECTIVE: Paget's disease is a progressive skeletal disorder, which involves the gradual destruction of the bones. Regarding the low prevalence of this disease in Iran, this study aimed to present a case of Paget's disease in an asymptomatic patient.

CASE REPORT: A 44-year-old male patient with elevated alkaline phosphatase (ALP) level in yearly examinations was referred to an internal medicine clinic. Considering the normal results of other examinations, high ALP level was found to be of an osseous origin. However, no bone deformity was detected in the patient. Bone scan and simple radiography were indicative of Paget's disease considering the high osteoblastic activity in the right hip and right sides of T_1 and L_3 segments. Diagnosis of Paget's disease was confirmed by bone biopsy. The patient was initiated on oral alendronate (35 mg daily) for six months. Following this period, ALP level reduced, and the treatment is still in progress.

CONCLUSION: According to the results of this study, although Paget's disease is a skeletal disorder, it could remain asymptomatic and occur without any deformities.

KEY WORDS: *Paget's disease, Deformity, Osteoblastic*.

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Introduction

Paget's disease is a bone disorder, which affects extensive skeletal regions and is highly prevalent in European countries (1,2). In Paget's disease, osteoclasts cause bone resorption increasing the formation of thick, fragile bones, which are highly susceptible to deformity and fractures (1,3).

Through the course of Paget's disease, various symptoms emerge from the primary, secondary and following compression of the nerve tissues (1,2,4). Multiple bone involvement leads to frequent fractures, increased head circumference, blindness and deafness, foot clubbing, kyphosis, and heart failure (5). Paget's disease may inflict one single bone (25% of the cases) or multiple bones (75% of the cases) (6). Hips, spine, femur and tibia, and lumbosacral region are most commonly affected by this disorder (7). In one study, Karabas et al. described a 57-year-old female patient with maxillary swelling, and X-ray radiography confirmed Paget's disease(8). In another report, Garg et al. presented a 59-year-old male patient with mandibular osteomyelitis, who was diagnosed with Paget's disease after radiographic examination (9). Male-to-female ratio in Paget's disease is 2:1, and 90-95% of the patients are aged over 40 years (5, 10). According to previous studies, the mean age of patients with Paget's disease at diagnosis is 68.7 years (11). Considering the low prevalence of Paget's disease in Iran, we described a 44-year-old male patient diagnosed with Paget's disease due to elevated level of alkaline phosphatase (ALP) in yearly examinations. This study aimed to emphasize the fact that different diseases may manifest through various unspecific forms and symptoms. Therefore, we attempted to investigate all the aspects and symptoms of Paget's disease.

Case Report

A 44-year-old male patient was referred to an internal medicine clinic for routine check-up with high ALP level as detected by yearly examinations (954, normal maximum: 280). After obtaining informed consent, the patient was enrolled in this experiment. The patient had no medical history of abdominal and bone pain, jaundice, itching, cardiac disease and medication use. In addition, the systemic examination was normal, and no bone deformity was observed. Also, blood cell count, liver enzymes, thyroid and kidney function, and blood glucose tests were normal. To determine the cause of high ALP level, the gamma-glutamyl transferase test was performed, which yielded normal results. Liver and biliary tract ultrasound tests were normal as well. Since liver disease and bile duct are not associated with Paget's disease, and other tests were normal in our patient, elevated ALP level was considered to be of an osseous origin. According to other examinations, concentrations of calcium, phosphorus, parathyroid hormone (PTH) and vitamin D were normal. Consequently, possibilities of hyperparathyroidism and osteomalacia were ruled out. In the next stage, bone scan was performed on the patient, and due to increased osteoblastic activity in the right hip and right sides of T_1 and L_3 segments, the possibility of Paget's disease was speculated (fig 1). As requested by the radiologist, simple radiographs were obtained from these regions, which were also indicative of Paget's disease (fig 2).

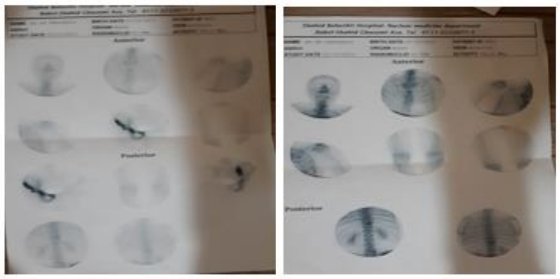


Figure 1. Increased Osteoblastic Activity in the Right Hip (slightly higher than the activity in the right sides of T_1 and L_3 segments)



Figure 2. Increased Cortical Bone Thickness and Sharp Trabecular Pattern due to Bone Resorption and Sclerosis

Moreover, complementary examination revealed a mild auditory acuity, which had been neglected by the patient. In addition, radiometric examination of the patient was indicative of sensorineural hearing loss, which was assumed to be caused by Paget's disease. To obtain a definitive diagnosis and rule out other hypotheses, pelvic bone biopsy was carried out, and

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the sample was sent for pathological examination. Paget's disease was confirmed through observing dense bone blades with mosaic patterns in the microscopic examination (fig 3). Treatment of the patient included oral alendronate administration (35 mg daily) for six months. After this period, ALP level reduced, and the treatment is still in progress.

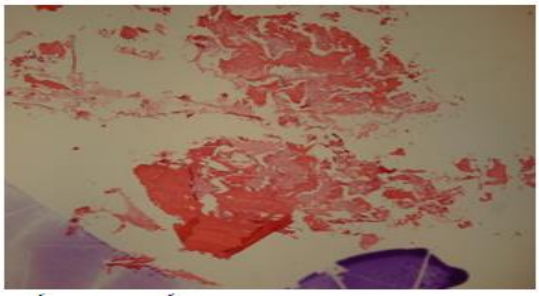


Figure 3. View of Dense Bone Blade at 40× Magnification (Haematoxylin and Eosin Stain)

Discussion

Bone deformity is the main feature of Paget's disease, which was not detected in our patient. With the exception of increased ALP level, the patient had no other symptoms of Paget's disease. Paget's disease affects diverse ethnic groups and is highly prevalent among Caucasians, while it is less prevalent among indigenous Asians (1, 3, 4). In countries with high prevalence of Paget's disease, the majority of patients are aged over 55 years (12). Histopathologically, the first stage of Paget's disease involves the lytic cycle, in which the lysis index is evident (13). Most symptoms of Paget's disease appear in the final stage, also known as the sclerotic stage, in which the sclerosing bone replaces the natural bone (14).

Consequently, patients often manifest facial deformities and increased skeletal volume, such as mandibular and maxillary hypertrophy (15). In one study, Khajavi et al. described a 17-year-old patient initially presented with progressive visual acuity in the left eye within the past two years. Visual acuity decreased in the right eye, and although the bone density had increased in that region, it was softer than expected. Bone samples were sent for pathological examination, and typical mosaic pattern of the samples confirmed the diagnosis of Paget's disease (16). In another report, Eghbali et al. described a 38-year-old patient who had referred for dental extraction since two years before. Significant facial deformity correspondent to leontiasis ossea was detected in the patient, and laboratory results were indicative of elevated ALP and PTH levels (17).In the aforementioned studies, both patients showed symptoms of Paget's disease, none of which were observed in our patient. The only symptom associated with Paget's disease in our patient was increased ALP level. According to international studies, Paget's disease more commonly affects men compared to women, and this finding has been confirmed in the Asian population (13). Furthermore, Paget's disease has the highest prevalence among elderly patients (18). According to some studies, the age at onset of Paget's disease is above 40 years, while other studies have suggested a median age of 68.7 years for this disorder (15, 19). On the other hand, few studies have reported the onset of Paget's disease at younger ages. This could be due to the fact that in some cases, Paget's disease remains asymptomatic and is identified through the sudden elevation of ALP levels (14). In conclusion, although Paget's disease is a skeletal disorder, it may remain asymptomatic and manifest without any bone deformities. Therefore, it is recommended that individuals aged over 40 years with high ALP levels be monitored for Paget's disease.

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