

Isolated Paget's Disease of Frontal Bone: A Case Report

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There are several reports about Paget's disease. We report a case who presented with headache and found to be have isolated Paget's disease of frontal bone with normal serum alkaline phosphatase which was not reported previously.

Key words: Paget disease, frontal bone, alkaline phosphatase.

Introduction

Paget's disease (PD) of bone is a disorder of bone remodeling, involving increased bone resorption and formation (1). It affects 3-4% of the population in northern Europe over 40 years of age (2). Diagnosis is usually made by elevated levels of serum alkaline phosphatase (ALP), urinary hydroxyproline, X-ray and scintigraphic findings (3).

PD most commonly involves the axial skeleton, but it can affect any area. The skull can be involved in 42% of patients with PD. In the majority of patients, PD affects at the least two bones (polyostotic), but in one third of patients only one bone is affected (monostotic or isolated) (4-6). We report a case who presented with headache and found to be have isolated PD of frontal bone and normal alkaline phosphatase levels which was not reported previously.

Case

66 years old, female patient admitted to Endocrinology Department because of severe headache. She had headache for 3 years. It usually starts from the mastoid region and then spreads to the whole face. She was being awakened at nights because of her pain. Her biochemical parameters in serum were normal [calcium: 9.6 mg/dl (8.6-10.2), phosphorus: 3.5 mg/dl (2.7-4.5), ALP: 131 U/L (0-270), Parathormone: 63.7 pg/ml (12-72)].

Paranasale and cranial tomography was normal. There was an osteoporotic and sclerotic area in the

frontal region in direct X-ray (Figure 1). After that finding the bone scintigraphy was taken. There was enhanced activity in the frontoparietal region in Tc-99m whole body bone scintigraphy (Figure 2,3). Although the ALP level was normal, ALP electrophoresis had done. And it was normal. (Liver 48.7%, intestinal 12%, bone origin 39.3%). Osteocalcin level was 16.16 ng/ml (normal range: 11-45). According to these findings, the patient accepted to be had isolated frontal bone PD.

Discussion

PD is a chronic, progressive bone disease. The disease may be monostotic (isolated) and polyostotic (4-6). Polyostotic involvement and activity are common at the time of diagnosis (7). Skull (25-65%), spine (30-75%), pelvis (30-75%) and proximal long bones are the most involved bones (2). The disease can be divided into three phases of increasing clinical and radiographic severity: 1) initial resorptive/osteolytic phase, 2) mid-phase, mixed osteoblastic/osteoclastic hyperplasia and 3) late sclerotic phase (4). Our patient may be accepted as mid-phase with dominantly osteoblastic hyperplasia according to bone scan.

The diagnosis of PD is based on a combination of the medical history and physical examination (particularly when increased heat or bone deformity is present), laboratory tests (measurement of serum ALP, and some instances, a urinary marker of bone resorption), and radiographic studies (bone scans and conventional radiographs) (8).

The origin of the PD is unknown, and it is frequently asymptomatic; however, the patient may present with symptoms depending on the bones

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Figure 1.



Figure 2.



Figure 3.

involved. Although neurologic symptoms are uncommon, headache, dementia, cerebellar dysfunction, cranial neuropathies, myelopathies and radiculopathies can be seen due to pagetic involvement of the skull (3,9). Our patient also admitted to our clinic because of severe headache. We detected osteoporotic and sclerotic areas in the frontal region on direct X-ray. In addition, scintigraphic activity was also enhanced in the fronto-parietal region.

There are many biochemical markers for PD. The serum ALP level is probably the most useful blood test in the diagnostic evaluation. The degree of ALP elevation of the ALP level is generally a measure of metabolic activity. For reasons not well understood, the highest levels usually are seen in patients whose involved sites include the skull. Minimally elevated or normal levels of ALP in patients with a single site of Paget's disease change may represent progressive disease at that site (isolated PD). Elevated ALP level usually provides bone turnover, about 15% patient present with normal serum ALP level and isoenzymes of ALP like our patient described above (10). The underlying mechanism of normal ALP levels is not well understood.

There are several reports about the monostotic involvement of Paget disease. Temporal bone (11), maxillofacial (12), vertebra (13), radius (14), costal region (15) hand (16) and patella involvements (17) had been reported in literature. Although skull involvement was commonly seen, there are no reports in the English literature showing the isolated involvement of frontal bone.

Finally, we report a case that had isolated PD of frontal bone and normal alkaline phosphatase levels which was not reported previously. Patients should be followed indefinitely because of the increased risk of malignant transformation in patients with longstanding PD.

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