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Bone and Parathyroid Scintigraphy Findings in Sagliker Syndrome

Sağlıker Sendromunda Kemik ve Paratiroid Sintigrafisi Bulguları

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Abstract

Sagliker syndrome (SS) is a rare, exaggerated form of chronic kidney disease (CKD)-mineral and bone disorder resulting from untreated secondary hyperparathyroidism due to CKD. Herein, we describe a 34-year-old male patient whose Tc-99m-methylene diphosphonate bone scintigraphy and Tc-99m-sestamibi parathyroid scintigraphy revealed hints of SS and exhibited its defining characteristics.

Keywords: Sagliker syndrome, renal osteodistrophy, secondary hyperparathyroidism

Öz

Sağlıker sendromu (SS), kronik böbrek hastalığına (KBH) bağlı tedavi edilmemiş sekonder hiperparatiroidizm sonucu ortaya çıkan, KBH-mineral ve kemik bozukluğunun nadir görülen, abartılı bir şeklidir. Burada Tc-99m-metilen difosfonat kemik sintigrafisi ve Tc-99m-sestamibi paratiroid sintigrafisinde SS belirtileri gösteren ve SS'yi tanımlayıcı özellikler sergileyen 34 yaşında bir erkek hastayı sunuyoruz.

Anahtar kelimeler: Sağlıker sendromu, renal osteodistrofi, sekonder hiperparatiroidizm

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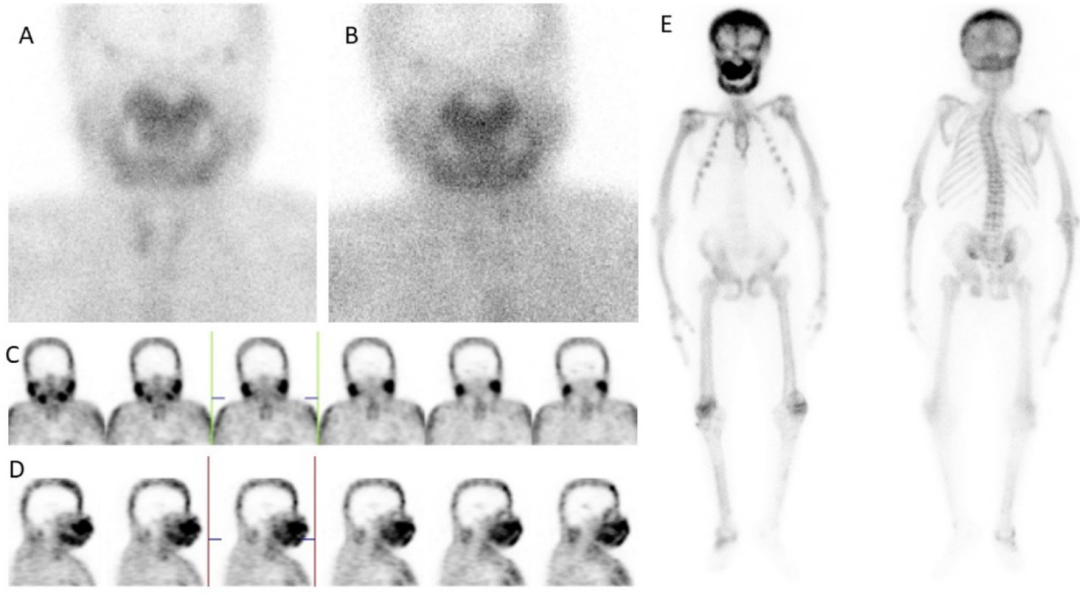

Figure 1. A 34-year-old male patient with secondary hyperparathyroidism was admitted to our department for parathyroid scintigraphy. The patient was in the hemodialysis program for 10 years because of chronic renal failure of unknown cause. Serum intact parathyroid hormone level was 3,521 pg/mL, total calcium level was 9.23 mg/dL, phosphate level was 5.4 mg/dL, 25-OH vitamin D3 level was 15.63 ng/mL, and alkaline phosphatase level was 1,375 U/L. Tc-99m-MIBI biphasic parathyroid scintigraphy (A, B) showed hyperplasia. However, on single-photon emission computerized tomography images (C, D), irregularly increased radioactivity uptake was observed in the maxilla, mandible, and calvarium, with the maxilla showing the greatest intensity. The facial deformity was noticeable, especially in the sagittal section (D). We performed whole-body bone scintigraphy (E) on a separate day to evaluate the head and other skeletal structures. Increased radioactivity uptake was observed in the axial and appendicular bones, most intensely in the maxilla and whole-body bone imaging with Tc-99m-MDP. No radioactivity uptake by the kidneys and bladder was observed. The distance between the anterior ends of the bilateral ribs increased remarkably toward the basal end in the anterior projection. The thorax had morphed into a bell-shaped mouth. There was scoliosis in the vertebral column and bending-bowing deformity in the upper extremities.

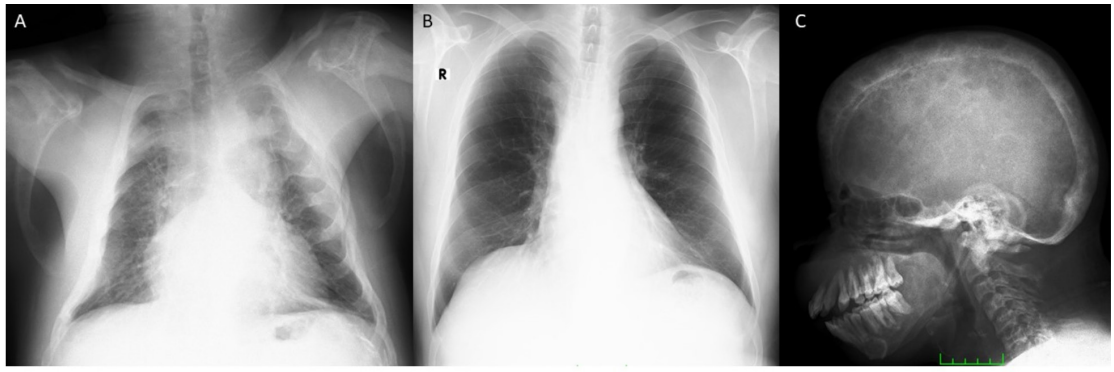

Figure 2. A posteroanterior chest radiograph (A) showed cardiomegaly, decreased lung volumes, rib fractures, and bilateral collapse of the lateral chest walls. The deformity seen in the current chest X-ray was not seen 6 years ago (B). The lateral head X-ray (C) revealed calvarial thickening and multiple lytic irregularly circumscribed lesions. In addition, there was a frontal forward malformation of the maxilla, and the lower and upper incisors were malposition. The patient was determined to have Sagliker syndrome (SS). Recently described SS is an aggravated form of renal osteodystrophy, which is an abnormal bone histology in chronic kidney disease (CKD) (1). In 1953, Cohen and Diamond (2) initially defined the signs of secondary hyperparathyroidism related to prolonged renal failure as uremic leontiasis ossea, which is marked by disfiguring craniomaxillofacial bone overgrowth (3,4). In 2004, Sagliker et al. (1) described this phenomenon in detail and named it SS. The symptoms of SS include a deformed facial appearance, maxillary and mandibular bone overgrowth, nasal bone and cartilage destruction, irregular teeth shape and localization, grade 2 maxillary malocclusion (anterior forward malformation of the upper jaws), short stature, soft and benign tissue deposits, upward curved fingertip changes, and knee and scapula deformities (1,5). In addition, SS affects the immune, neuropsychiatric, and cardiovascular systems as well as the musculoskeletal system (6). The etiology of SS remains unknown. SS is especially seen in developing countries. It is thought that financial deficiency, sociocultural deficiencies, and unwanted iatrogenic late, incomplete, or incorrect treatment methods play an essential role in the development of SS (7). After performing gene mutation studies, Demirhan et al. (8) postulated that SS may be caused by a combination of CKD, hereditary osteodystrophies, and bone dysplasias. Many patients do not notice the onset of SS because the symptoms develop slowly. Early medical and surgical treatment can prevent irreversible musculoskeletal deformities and cardiovascular disorders.

Ethics

Informed Consent: Patient consent was obtained.

Authorship Contributions

Concept: Ç.E., Ö.Ş., Design: Ç.E., Ö.Ş., Data Collection or Processing: Ç.E., Ö.Ş., Analysis or Interpretation: Ç.E., Ö.Ş., Z.A., Literature Search: Ç.E., Ö.Ş., A.E.Ş., Writing: Ç.E.

Conflict of Interest: No conflicts of interest were declared by the authors.

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