INTRODUCTION

Buschke-Ollendorff syndrome is a rare hereditary disease characterized by small nodular opacities osteopoikilosis in bones and collagenoma and/or elastoma in the skin. Roberts et al. have identified Buschke-Ollendorff syndrome, indicating that osteopoikilosis may be associated with dermatofibrotic changes [2].

CASE REPORT

A 22-year-old male patient with pain and tenderness on his wrist was taken for radiography. On radiographs, uniform, multiple opacities with a millimeter diameter consistent with osteopoikilosis were noted around in the wrists, fingers and metacarps and evaluated as compatible with osteopoikilosis (Figure 1).

Considering that osteopoikilosis can be seen in various bones, PA lung radiography, pelvic radiograph, knee and ankle radiographs were obtained by including shoulder joints. In these radiographs, osteopoikilosis-compatible appearance was also observed in the vicinity of various joints such as the shoulders (Figure 2) and ankles.

During the examination of painful swellings in different parts of the body skin lesions of the right scapular region, which can be considered as connective tissue nevus in the diameters between 2-5 mm, were detected and the collagenoma was interpreted in favor (Figure 3).

In addition, 2-5 cm swellings were detected in the soft tissues of the right gluteal region and left cruris in the palpation of painful areas. These nodular swellings were also evaluated by painful nodular, soft tissue ultrasonography (Figure 4).

Bilateral thoracolumbar vertebrae radiographs taken due to the present back pain were normal and there were no appearances in favor of osteopoikilosis. Thoracic magnetic resonance imaging (MRI) was performed due to the continuation of back pain and MRI showed mild hypointense on T1-weighted MR imaging (Figure 5), and hyper intense signal on T2-weighted axial (Figure 5) imaging, and showed hyper intense signal on fat suppressed T2-weighted sagital image (Figure 5) in 11th thoracic vertebra corpus. Routine blood tests and abdominal ultrasonography of the case were within normal limits.

Key Words: Buschke-Ollendorff syndrome; Radiography; Hereditary disease
Histopathologically, multiple compact bone islets are seen in osteopoikilosis. Therefore the cases are usually clinically asymptomatic [5]. They are detected incidentally [4,6]. However, the similarity of radiological appearance of such lesions should be differentiated from sclerotic-osteoblastic metastases, bone mastocytosis and tuberous sclerosis [4]. In our case, the lesions were detected incidentally with hand-wrist radiographs radiograms were taken due to trauma.

Osteopoikilosis can be detected alone, but may be associated with some clinical conditions [1]. Aortic coarctation, urogenital defects and/or variations, puberty precox, some organ anomalies; endocrine disorders such as diabetes mellitus, spinal canal stenosis or dental and facial anomalies are reported as congenital accompanist anomalies with osteopoikilosis [7]. In one case, osteopoikilosis and osteosarcoma were reported to be associated with active osteogenesis [8]. In Gürul-Sebe-Baftaran syndrome, the association of osteopoikilosis with dacrocyctitis has been demonstrated [9].

In our case, there was no problem of endocrine disorders and no organ pathologies as abdominal or thoracic region. However, there was a vertebral hemangioma in corpus of 11th dorsal vertebra, which was not previously described in the literature. Approximately 25% of the patients with osteopoikilosis have yellow-white, papular skin lesions localized on the lower back, buttocks, thighs and arms, which can show symmetry [1,2,10]. The most common of these lesions are the nevus in the connective tissue. The most common form of the nevi are elastomes, which are composed of a kind of flexible connective tissue. In individuals affected by this syndrome, there may be less frequently called nevus called collagenoma which develops from collagen connective tissue. Discoid lupus erythematosus and keloid may be associated lesions of this syndrome [4,11].

For these reasons, patients with osteopoikilosis should be informed about their current conditions and if possible, their radiographs and reports should be given to them. Otherwise, they may be subjected to unnecessary examination and exposure to ionizing radiation for differential diagnosis and accompanying pathologies once they apply to other centers [12].

CONCLUSION

We suggest that multiple small compact bone islets (osteopoikilosis), which we frequently encounter in daily practice, can be reported in the reports based on incidental detection of bone and skin lesions.

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