Right Hip Pain in a 65-Year-Old Woman

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Clinical Case

We present the case of a 65-year-old woman with a personal history of cataracts, hypertension, depression, and bilateral knee osteoarthritis of 4 years. She came to our clinic due to mechanical lower back pain that had its onset 5 years prior and had no irradiation, and intense right mechanical inguinal pain for 3 months. The patient also referred progressive hearing loss and a dark coloration of urine during the past few years.

Upon examination we found a blue coloration of both ears (Figure 1). Locomotor examination showed pain upon palpation of dorsolumbar spinous processes as well as upon lumbar spine movement. The right hip had complete limitation for rotation and a flexor deformity at −30º and the right knee presented pain and crepitus upon palpation of the patella, without any signs of arthritis. Laboratory testing showed normal hemogram, immunoglobulin concentration, calcium, phosphorus, 25-OH vitamin D, parathyroid hormone, thyrotropin (TSH), antinuclear antibodies, HLA B27, and urine tests. She had an alkaline phosphatase of 141 U/L (35-104 U/L) and C-reactive protein of 9.3 mg/L (0-5 mg/L). On the pelvic x-rays (Figure 2) there was evident osteonecrosis of the right femoral head with deformity and collapse. The lumbar spine x-rays (Figure 3) showed calcification of all of the intervertebral disks and a vacuum effect and subchondral sclerosis of L5-S1. A magnetic resonance of both hips showed degenerative changes and osteonecrosis that had progressed, in the right coxofemoral joint, edema of the accompanying bone marrow, and joint synovitis. The left coxofemoral joint presented the same alterations but in an earlier stage.

Diagnosis and Progression

In order to reach the definite diagnosis, homogentisic acid was determined in a 24-hour sample of the patients' urine through photometry, showing a value of 1175 mg/L (0-0 mg/L) and confirming the diagnosis of ochronosis. It was noticed that urine turned dark upon exposure to sunlight (Figure 4). The study was completed with an echocardiogram in which an aortic valve sclerosis was seen, a densitometry with a lumbar spine T *score* of −3.24 standard deviations (SD) and of −1.51 SD on the femoral neck, bilateral hypoacusia was also evidence upon hearing examination. A total hip arthroplasty was performed on the right hip and a left hip prosthesis was placed after 8 months due to rapidly progressive joint destruction.

Discussion

Alkaptonuria is a rare autosomic recessive disease of thyrosine catabolism, characterized by homogentisic acid in urine (alkaptonuria) and in diverse, collagen-rich tissues (ochronosis). Its prevalence is approximately 1/1 000 000 inhabitants.[1] It is more frequent and severe in males, with a peak between ages 30-40. Up to 16 types of mutations

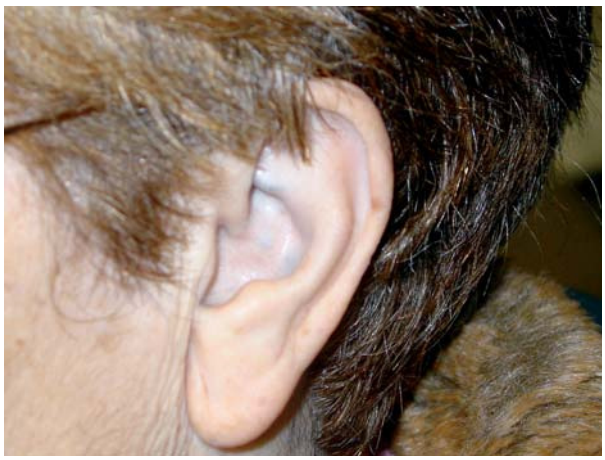

Figure 1. *Photo showing a blue coloration of the ear cartilage.*

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Figure 2. *Simple anteroposterior x-ray of the pelvis showing osteonecrosis of the right femoral head with deformity and collapse.*

of the *HGO* human gene[2] located on chromosome 3 have been identified. It has been related to HLA-B27 positive persons. Among the clinical manifestations of ochronosis, the dark blue pigmentation of the ears, malar region, and sclerae is characteristic. The outer ear wax is oftentimes black and occasionally leads to hearing loss. Urine acquires a dark brown coloration upon contact with air due to oxidation of the homogentisic acid in it. Cardiac affection can be seen, with calcification of the aortic valve, systolic heart murmurs, involvement of the supraaortic branches, and pericardium.[3] Some cases of affected cellular immune responses and secondary recurrent bacterial infections have been reported.[4] In a great deal of males, black prostatic stones can be found.[5]

With time, severe spondyloarthrosis develops characterized by progressive rigidity of the lumbar spine and loss of the lordotic curve, manifested as mechanical back pain. X-rays show characteristic calcifications and ossification of the intervertebral disks in the absence of osteophites. Occasionally, destructive arthropathies can occur, especially on the shoulders, hips and knees.[6] In our case, a rapidly progressive arthropathy of the hips with association of bilateral avascular bone necrosis, something that has not been published and may play a pathogenic role. Spontaneous rupture of tendons has been reported as an initial manifestation.[7]

The differential diagnosis with ankylosing spondylitis and osteoarthritis must be done. Current treatment is symptomatic because there is no effective therapy. Phenylalanine-deficient or vitamin C rich diets have been employed without any success. A review of cases with lumbar spine affection due to disc hernia and cord compression mentions that surgery could be useful in selected cases.[8] Another study has shown the lack of efficacy of treatment with alendronate.[9]

The interest of this case resides in the inclusion of ochronosis in the differential diagnosis of a middle-aged patient with rapid progressive destruction of the hips and chronic mechanical lower back pain.

Figure 3. *X-ray of the lumbar spine in a lateral projection: calcification all of the intervertebral discs can be seen and a vacuum effect with subchondral sclerosis of L5-S1 is evident.*

Figure 4. *Dark-colored urine sample after exposure to sunlight.*

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