Black hip: a rare case treated by total hip replacement

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BACKGROUND AND OBJECTIVE: Ochronic arthropathy of hip (Black Hip) is a rare clinical manifestation of congenital disorder of amino acid metabolism characterized by a classic triad: (1) degenerative arthritis, (2) ochronotic pigmentation, and (3) urine that turns black on long standing or alkalinization. We report a case of ochronic arthropathy of the left hip joint that was successfully treated by total hip arthroplasty.

DESIGN AND SETTINGS: This is a case study conducted at PES Medical College, Andhra Pradesh, India.

PATIENTS AND METHODS: A 60-year-old female patient came with a history of progressive pain in her left hip joint for the last 8 months. She was diagnosed to be suffering from ochronic arthritis of left hip.

RESULTS: After tissue confirmation she was operated with total hip replacement. At the end of 2 years, the patient was symptom free without any implant loosening.

CONCLUSION: Ochronotic arthropathy is a rare metabolic disorder that can be underdiagnosed many a times. Early management is only symptomatic, and advanced cases need surgical intervention. Vitamin preparations are given because of the influence of vitamin C on tyrosine and phenylalanine metabolism. In the cases of severe degenerative arthritis of hip, total hip replacement may be considered as a surgical option.
the diagnosis of ochronosis was made.

The patient was planned for elective total hip replacement under epidural anesthesia. The hip joint was opened with Moore’s approach. During the exposure, the tissues around the joint were found to be dark in color with gross involvement of the joint capsule and the synovium (Figure 6). It was decided to abandon the procedure to have a tissue diagnosis keeping in mind the rare possibility of chronic fungal infection. The joint was debrided and the tissue was sent for histopathological examination. The histological sections of removed bone and soft tissue demonstrated the classic findings of ochronosis, including pigmented areas, reactive giant cells, and thickened synovium. Once the histopathology report confirmed the diagnosis of ochronosis, the patient was operated with reverse hybrid total hip replacement after 1 week (INDUS cemented cup and uncemented stem; Figure 7). The patient was followed up with regular intervals at 6 weeks, 3 months, 6 months, and yearly afterward. At the end of 2 years, the patient was symptom free without any loosening of the implants and was
Ochronosis or alkaptonuria is an autosomal recessive disorder due to the deficiency of homogentisic acid oxidase, which is an important enzyme in the metabolism of amino acids phenylalanine and tyrosine. Normally, this enzyme breaks down homogentisic acid to fumaric acid and acetoacetic acid. In the absence of this enzyme, homogentisic acid gets polymerized and deposited in the connective tissues. Some part of homogentisic acid gets excreted through urine and sweat. Such urine turns dark color after long standing or mixing with alkaline agent like sodium hydroxide. Confirmatory tests for diagnosis are chromatographic, enzymatic, or spectrophotometric determinations of homogentisic acid (HGA).

Patients with alkaptonuria may be asymptomatic throughout their life, but severe cases will be manifested early in the infancy like staining of the nappies on expose to air. Maximum number of cases is diagnosed in the fourth to fifth decades. These patients are presented with staining of clothes in the axillae and appearance of pigmented nodules in pinnae, sclera, or skin. Ochronosis affects the entire body and can involve cardiovascular, genitourinary, ocular, cutaneous, and musculoskeletal system.

Ochronotic arthropathy usually manifests in adult life with the progression of symptoms with age. In severe cases, arthropathy occurs much earlier. Cartilage has got special affinity for homogentisic acid pigment, and when sufficient amount is this pigment is deposited within the articular plates, it leads to the death of chondrocytes and calcification of cartilage.

Spine is the first region to get affected, with patients complaining of stiffness and low back pain followed by weight-bearing joints like hip and knee. Smaller joints do not develop arthritis, but the cartilage within these joints shows pigmentations. The deposited HGA polymer in the cartilage leads to brittleness and fragmentation, which in turn leads to degenerative arthritis. The involvement of intervertebral disks leads to pigmentation and ossification of the nucleus pulposus, leading to degenerative changes. Pigmentation may involve tendons and ligaments because of their collagen content, which leads to tendon inflammation, calcification, and rupture.

Primary treatment for ochronotic arthritis is mainly symptomatic. Physiotherapy and NSAIDS are beneficial but do not slow down the progression of the disease. Vitamin C preparations are given because of its influence on tyrosine and phenylalanine metabolism. Dietary restrictions of food containing phenylalanine and tyrosine have proved to be successful in decreasing the symptoms. In the cases of severe ochronotic arthritis, total joint replacement may be considered for symptomatic pain relief.

CONCLUSION
Ochronotic arthropathy is a rare disorder of metabolism that can be underdiagnosed many times. Sometimes it may present as a surprise to the surgeon on the operating table. Early management is only symptomatic and advanced cases need surgical intervention. As we reported, in the cases of severe degenerative arthritis of hip, total hip replacement surgery may be considered with preoperative counseling and explanation of outcomes of surgery compared to non-chronic patients.
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