AJBMS AFGHANISTAN
ISSN: 3005-6632

Afghanistan Journal of Basic Medical Sciences

2026 Jan 3(1): 102-106.

ajbms.knu.edu.af

Ochronotic Arthropathy Presenting as Severe Degenerative Arthritis in a Patient Undergoing Total Hip Arthroplasty: A Case Report

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ARTICLE INFO

Type: Case Report

Received: 16 April, 2025

Accepted: 27 Nov, 2025

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To cite this article:

Emami Meybodi MK, Peypar MH, Abdorrashidi H, Mohammadi MR, Shater MM, Afkhami. Ochronotic Arthropathy Presenting as Severe Degenerative Arthritis in a Patient Undergoing Total Hip Arthroplasty: A Case Report. Afghanistan Journal of Basic Medical Sciences. 2026 Jan 3(1): 102-106.

DOI: https://doi.org/10.62134/khatamuni.85

ABSTRACT

Ochronotic arthropathy is an uncommon disorder observed in individuals with alkaptonuria, characterized by the buildup of Ochronotic pigment. This case report provides the clinical presentation, diagnosis, and successful surgical treatment of a 64-year-old Asian in (Baqiyatollah Hospital, Tehran, Iran) female patient with ochronotic arthropathy, an uncommon disease characterized by the accumulation of dark pigments in the tissues of the joints. The patient initially exhibited progressive pain and limited mobility in the left hip, ultimately diagnosed as severe degenerative arthritis. Despite conservative management, such as analgesic medication and physical therapy, the symptoms continued to linger. The surgical procedure involved uncemented total hip arthroplasty (THA) using the anterolateral approach. During the 6-month check-up, the patient showed a satisfactory range of motion and no report of pain or discomfort. Having a thorough knowledge of ochronosis and its various forms is crucial for promptly diagnosing and efficiently managing the condition. Our presented case demonstrated the effectiveness of total hip arthroplasty (THA) in treating ochronotic arthropathy.

Keywords: Arthroplasty; Alkaptonuria; Arthropathy; Ochronosis; Arthritis

Introduction

Ochronosis is a condition where the connective tissue shows signs of alkaptonuria, caused by an inactive gene located on chromosome 3q21–23 (1). This gene is responsible for producing the enzyme of homogentisate 1,2 dioxygenase, also known as HGD (2). The outcome is the systematic accumulation of homogentisic acid (HGA), a colorless phenol, which can irreversi-

bly attach to collagen upon its conversion to benzoquinone (3). An additional consequence is that the polymerization of this molecule results in an iridescent dark hue, which imparts a tint to the damaged tissue such as all varieties of collagen (4). The incidence of Alkaptonuria (AKU) in the majority of ethnic groups is below 1 in 100,000, with a global prevalence of one in 100,000 to 250,000 individuals. The condition is uncommon, although in certain groups, its frequency might be significantly higher (5). People with ochronosis frequently experience ocular signs. An example of such is the characteristic limbal "oil-drop" lesions, which might serve as a valuable diagnostic clue (6).

The orthopedic symptoms of this condition consist of thickening and ripping of tendons, muscle damage occurring even with minor injuries, and the development of ochronotic arthropathy in the knee, hip, and shoulder joints (7). The surgical treatment options for degeneration vary according on its degree and may involve arthroscopic debridement, synovectomy, or arthroplasty. While ochronotic arthropathy is a rare and delayed consequence of AKU, it can progress quickly and aggressively. Oftentimes, the illness remains undiagnosed until the moment of arthroplasty (8, 9).

This report describes a case of ochronotic arthropathy in a patient who underwent an uncemented total hip arthroplasty (THA).

Case report

A 64-year-old Asian woman, who previously had gastritis and urolithiasis four years ago, was referred to the Department of Orthopedic Surgery at Baqiyatallah Hospital, Tehran, Iran in 2025, with progressive pain and movement restriction in the left hip in the last year. There was no record of any physical injury or signs of inflammatory arthritis. Additionally, there was no history of metabolic problems in the family. The clinical examination revealed a reduced range of motion (5) in the hip joint, along with discomfort when moving, particularly during internal rotation. No notable findings were observed in the other exams. The radiographic examination of the hip indicated the presence of degenerative arthritis, characterized by a reduction in joint space, confirming the diagnosis of left hip osteoarthritis (Fig. 1).

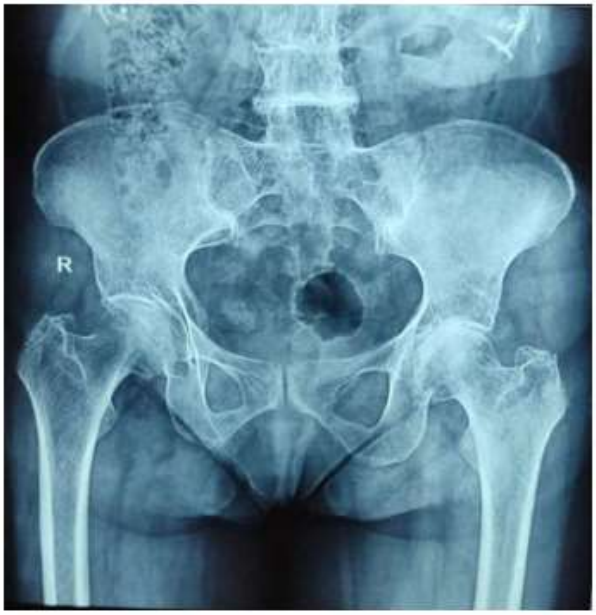

Fig. 1: Anteroposterior x-ray view of pelvis showed subchondral sclerosis with joint space reduction in the hip joint on both sides, preferably the left side (Original)

Initially, the patient underwent conservative management, which included the administration of analgesic medication and physical rehabilitation. Regrettably, these measures yielded unsatisfactory results. After a year she referred to our department. In the clinical examination of the left hip, we found limited ROM and pain, especially in internal rotation movements. The left hip showed clear radiographic signs of severe degenerative arthritis. After cardiac and anesthesia consultation and usual pre-operative tests, the patient was a candidate for uncemented THA surgery through anterolateral approach. During the procedure, both the capsule and the

surrounding tissues exhibited a dark hue resembling that of tar.

No contraindications were found during this observation, and the treatment was successfully completed without any interruptions. Further analysis revealed that the femoral head, acetabulum area, and surrounding tissues also exhibited dark pigmentation. Specimens were collected and sent for PCR culture and pathology investigation (Fig.2).

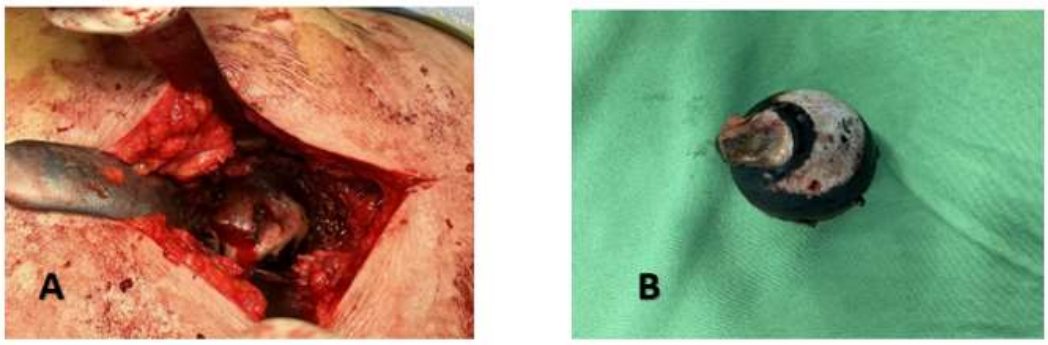

Fig. 2: Intraoperative observations. A black tissue was observed following exposure B) black capsule was observed upon exposure

In the histological study with hematoxylin and eosin (H&E), the sections showed mature bone trabecula and intervening space with a dark-brown pigment compatible with ochronosis (Fig. 3). Following the surgical treatment, the patient got a thorough checkup to identify any additional symptoms or signs of the condition. During this assessment, multiple pigments were noted in the sclera (Fig. 4). In addition, when the patient's urine was exposed to air, it turned black, and the concentration of HGA in her urine increased.

Fig. 3: Histological study with hematoxylin and eosin (H&E)

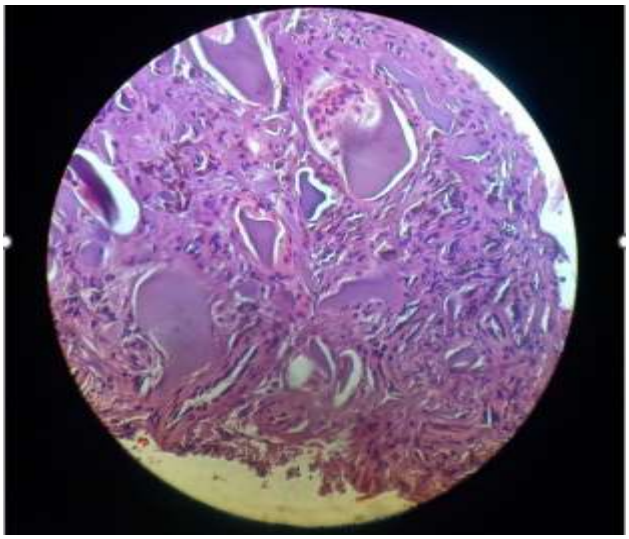

Fig. 4: Ochronotic pigmentation of the sclera

Eventually, the procedure resulted in a substantial reduction in her pain. In the latest follow-up, 6 months after the surgical procedure, the range of motion (5) in the left hip was considered satisfactory, with no reported pain or discomfort. Additionally, the radiologic studies showed no further pathological findings in femoral or acetabular components (Fig. 5).

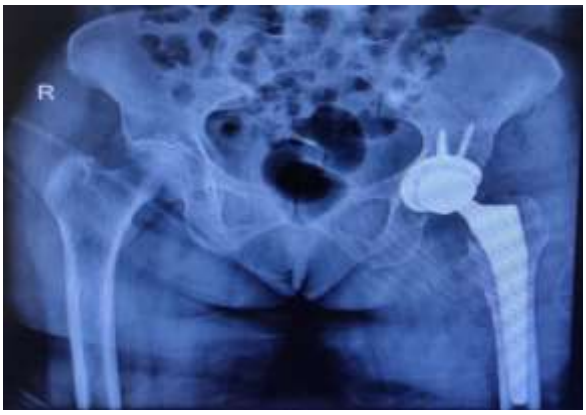

Fig. 5: Pelvic X-ray after the operation

Discussion

This case report highlights the challenges involved in diagnosing and treating ochronotic arthropathy, especially when it occurs as a late consequence of alkaptonuria. The rarity of the condition, coupled with its insidious progression, frequently results in a postponed identification, as observed in this report.

Ochronosis is a hereditary metabolic disorder caused by the lack of HGD, leading to the buildup of HGA in different organs, such as connective tissues (4). During the physical examination, our patient exhibited multiple pigmented areas in the sclera. However, Kazemi et al. also noted the presence of multiple pigmented areas in the axillary region, in addition to the sclera region (10). The favorable result of the anterolateral total hip arthroplasty (THA) indicates that surgical treatment can effectively relieve pain and enhance joint function in individuals with ochronotic arthropathy. Patients with ochronotic arthritis who had joint replacement surgery had comparable outcomes in terms of the durability and efficacy of their prosthetic joints when compared to those with osteoarthritis (11).

We found that six months after the operation, the patient's left hip had a sufficient range of motion, and there were no reports of pain or discomfort. The surgery had a beneficial effect on the patient's quality of life. Following the

surgery, PCR culture of the samples confirmed that there was no infection present.

Conclusion

However, given the potential for confusion with several infections, like mucormycosis, it is crucial to accurately diagnose this condition prior to surgery by carefully considering additional signs and symptoms of the disease, such as large joint arthralgias and pigmentation of the sclera.

Funding

No Funding

Competing interests

The authors declare no conflict of interest, financial or otherwise

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