

Total Hip Arthroplasty in Ochronotic Arthropathy with quadriceps tear. A Case Report with a Review of the Literature

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Learning Point of the Article:

A comprehensive clinical assessment can help effectively plan surgery for patients with this rare condition by predicting anesthesia challenges. Total hip replacement can lead to a successful outcome in patients with ochronotic arthropathy. The use of capsulotomy can be considered for these patients. In addition, spinopelvic parameters must be taken into consideration in cases of fused spine to prevent impingement or dislocation.

Abstract

Introduction: Alkaptonuria is a rare genetic disorder characterized by the absence of the enzyme homogentisate 1,2 dioxygenase, which leads to the accumulation of homogentisic acid in connective tissues and blood, resulting in Ochronosis and osteoarthropathy.

Case Report: We present a case report of a 56-year-old male with ochronotic arthropathy and quadriceps tendon rupture who underwent a total hip replacement. The patient exhibited right hip pain, difficulty walking, and restricted movement at the hip and spine. Radiographic and magnetic resonance imaging findings confirmed significant arthritic changes in the right hip joint and a mid-substance tear in the quadriceps tendon. Intraoperatively, dark pigmentation was observed in the affected tissues, consistent with the diagnosis of Ochronosis. The literature review indicates that joint replacement surgery is the treatment of choice for ochronotic arthropathy, with patients experiencing good functional recovery. The prosthesis's long-term survival rate is comparable to primary arthroplasty, and the optimal surgical approach regarding capsule preservation remains a topic of debate.

Conclusion: Joint replacement is a feasible treatment option for patients with ochronotic arthropathy. Understanding the spine-pelvic relations and other potential complications can help surgeons plan for better recovery and functional outcomes.

Keywords: Ochronotic arthropathy, alkaptonuria, total hip replacement.

Introduction

Alkaptonuria is a rare genetic disorder with autosomal recessive inheritance which is caused by the absence of the enzyme homogentisate 1,2 dioxygenase, which is responsible for the breakdown of homogentisic acid, an intermediate in the tyrosine degradation pathway [1]. Homogentisate 1,2 dioxygenase is produced in the liver hepatocytes and kidney cells. The absence of this enzyme leads to the accumulation of homogentisic acid within the cells and blood, which then gets excreted through urine. However, due to significant accumulation and relatively

lesser excretion, the homogentisic acid accumulates in connective tissues (such as menisci, tendons, intervertebral discs, articular surfaces, sclera, and pinnae), leading to dark pigmentation, and a condition known as Ochronosis [1]. Homogentisic aciduria, Ochronosis and osteoarthropathy characterize the disorder. It presents with multisystemic involvement, with the spine, hips, and knees commonly affected in the musculoskeletal system. The patient's urine changes color on exposure to air or reducing agents [2].

The diagnosis is often suspected intraoperatively due to the

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Figure 1: The clinical image of the patient from front, side, and behind.

characteristic black pigmentation of tissues and confirmed with a histological diagnosis. We present an interesting case report of a patient with ochronotic arthropathy and quadriceps tendon rupture.

Case Report

A 56-year-old male patient came in with a chief complaint of right hip pain for the past year and difficulty walking for the last 6 months. The pain worsened with movement and improved with rest. The patient had no other significant medical history. An antalgic gait with a Trendelenburg component was observed during the clinical examination. Gross wasting of the right

gluteal and thigh region was noted, along with an extension lag over the right knee and a palpable defect over the quadriceps tendon area (Fig. 1). Movement at the hip and spine was restricted. No pigmentation or discoloration was found on the body, and the patient did not report any blackening of urine. Routine blood tests, including the rheumatological profile, came back within normal limits.

The radiography results revealed significant arthritic changes in the right hip joint, with reduced joint space and deformation of the femoral head, indicating osteonecrosis (Fig. 2). The lateral radiograph of the knee showed patella baja (Fig. 3), and the lumbar spine exhibited a calcified disk with reduced intervertebral space (Fig. 2). An magnetic resonance imaging of the right knee confirmed a mid-substance tear in the quadriceps tendon (Fig. 3).

Under spinal anesthesia, the patient underwent surgery for a total hip replacement on the right side using a modified Hardinge approach. During the procedure, we observed dark pigmentation in the hip capsule, synovium, femur head, and acetabulum articular surface (Fig. 4). Samples were taken for

histopathological examination, and we proceeded with total hip replacement involving anterior capsulotomy.

The patient recovered well with no complications. The patient was followed up at 6 weeks, 3 months, 6 months, 1 year, and 3 years. At the final follow-up, the implants are in good position (Fig. 5), and Harri's hip score improved to 88 from 37 preoperatively. The

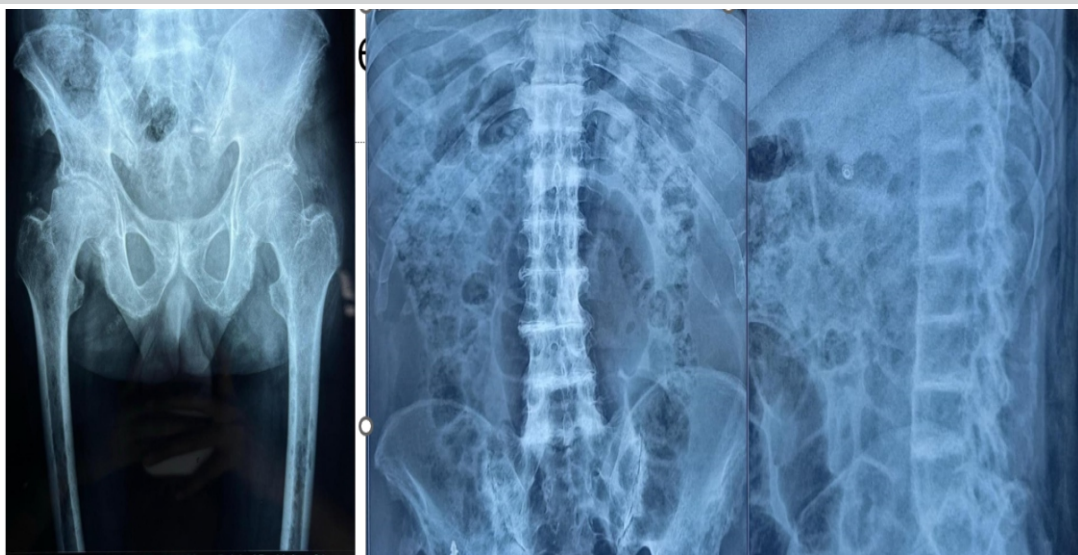


Figure 2: Pre-operative radiographs of the pelvis with both hip and lumbar spine.



Figure 3: Radiographs of right knee with thigh and magnetic resonance imaging (T2-weighted images) of right thigh with knee.

patient can extend the knee with an extensor lag of 15° and, hence, is not willing to perform quadriceps reconstruction.

Discussion

Alkaptonuria is a congenital disorder of metabolism that affects the body's connective tissue and large weight-bearing joints. It causes arthritis in the large weight-bearing joints, making movement difficult and impacting daily activities. Most patients are diagnosed incidentally with spontaneous blackish discoloration of urine, or they may develop symptoms in the 3rd–5th decade of their life. Intraoperative diagnosis becomes likely with blackish discoloration of the bursae, capsule, synovium, and cartilage. The disease process significantly

affects joints, and due to delayed presentation, joint replacement surgery becomes the treatment of choice [3].

Table (Table 1) provides a brief literature review comparing previous case reports and their key findings. The literature suggests that patients experience good functional recovery after joint replacement surgery.

The 12-year survival rate of the prosthesis is similar to that of primary arthroplasty, as reported by Spencer et al. [3]. There is no scientific evidence reported in the literature on the use of cemented versus uncemented implants in the case of ochronotic arthropathy. In their study, Cebesory et al. [4] and Fernando et al. recommended the removal of the anterior capsule due to marked thickening. Our study also found

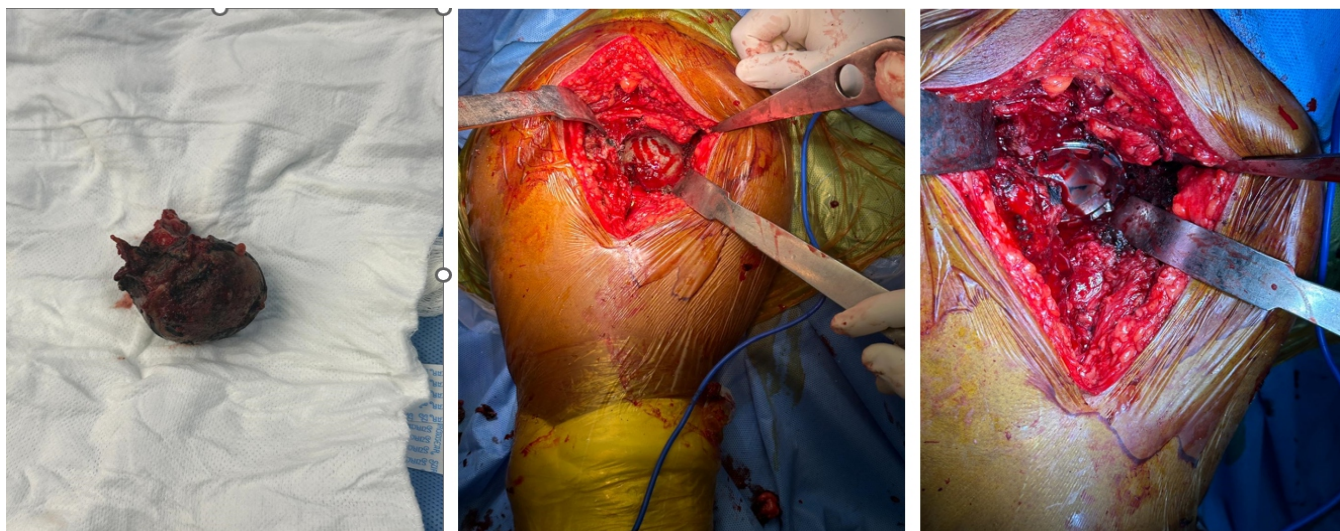


Figure 4: Intraoperative images; (a) dark-colored cut femoral head (b) black discoloration of acetabular articular surface (c) uncemented acetabular shell placement.



Figure 5: Post-operative radiographs after total hip replacement.

during surgery [5].

anterior capsular hypertrophy, which we subsequently removed. However, Pachore et al., in their study of 12 patients with a follow-up of 3–24 years, found good results with the preservation of the capsule

A few studies have anticipated difficulty in spinal anesthesia due to changes in the spine and have preferred general anesthesia as the safer option [6, 7]. In our case, no such difficulty was observed, probably due to the preservation of the space. In addition, spinopelvic parameters must be considered in the fused spine to prevent impingement or dislocation.

In our case, an incidental diagnosis of alkaptounuria was made intraoperatively with exposure. Following this, an uncemented total hip arthroplasty was performed. Incidental sporadic cases of alkaptounuria are found among the Indian population. Proper pre-operative assessment with appropriate surgical steps done by a competent surgeon can be sufficient for adequate management and excellent outcomes.

Conclusion

The case emphasizes the effects of alkaptounuria and ochronotic arthropathy on the musculoskeletal system, especially on the

Serial number	Author	Age/sex	Joint involved	Intervention	Salient feature
1	Cebesoy et al., 2014 [4]	46/male	Hip pain	Total hip replacement	The anterior capsule was found contracted and was removed
2	Machoochy and Razi, 2018 [8]	57/female	Hip and knee pain	Total knee and total hip replacement	Excellent outcome at 2 year
3	Gowda et al., 2013 [9]	60/female	Hip pain	Total hip replacement	Excellent outcome at 2 year
4	Rajkumar et al., 2020 [10]	27 total joint arthroplasties in 16 patients	Hip and knee pain	Total knee and total hip replacement	Improvement in Harris hip score and Knee society score at a follow-up from 12 to 132 months.
5	Siavashi et al., 2009 [11]	54/female	Hip fracture	Cemented total hip replacement	Incidental intraoperative diagnosis;
6	Fernando et al., 2018 [12]	69/female	Hip pain	Bilateral staged total hip arthroplasty	Advocated for excision of capsule, excellent follow-up after 1.5 years
7	Chu et al., 2021 [13]	48/female	Back pain		misdiagnosed as ankylosing spondylitis
8	Arasu and Prasanna, 2022 [14]	68/female	Hip pain	Total hip replacement	Excellent outcome at 1 year
		36/male	Hip pain	Bilateral staged total hip replacement.	Excellent outcome at 5 year
		56/male	Hip pain	Total hip replacement	Excellent outcome at 2 years
9	Siavashi et al., 2009 [11]	54/female	Hip fracture	Total hip replacement	
10	Al-Ajlouni et al., 2020 [15]	60/male	Hip pain	Total hip replacement	Excellent outcome at 10 years follow-up
		46/female	Hip pain	Total hip replacement	Good outcome at 3 years follow-up
		50/male	Hip pain and knee pain	Total hip replacement and total knee replacement	Good outcome at 3 years follow-up
11	Ilyas et al., 2020 [16]	45/male	Hip and knee pain	Staged total hip and knee replacement	Hip Capsular resection was done, and excellent follow-up at 12 years
12	Singh et al., 2024 [17]	43/male	Bilateral hip and knee pain	Total hip and knee replacement	Excellent follow-up at 11 years
13	Paul et al., 2024 [18]	60/female	Hip fracture	hemiarthroplasty	Thickened joint capsule present, satisfactory outcome at 13 months
14	Bhattacharjee et al., 2023 [19]	52/male	Hip pain	Bilateral total hip arthroplasty with dual mobility cup	Follow-up at 6 months
15	Current case	56/male	Hip pain with a quadriceps tear	Uncemented total hip replacement	Incidental intraoperative diagnosis; excellent outcome

Table 1: Review of existing literature with patients with ochronotic arthropathy.

hip joint. The patient's successful recovery after total hip replacement highlights the significance of joint replacement surgery as a feasible treatment for this condition. Further research may be warranted to explore the long-term outcomes and potential differences in using cemented versus uncemented implants in cases of ochronotic arthropathy.

Clinical Message

As a rare condition, a comprehensive clinical assessment can assist in effectively planning surgery for these patients by predicting anesthesia challenges, identifying any concurrent spine issues, and improving postoperative results. The use of capsulotomy can be considered in patients.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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