International Journal of Research in Orthopaedics
Shah V et al. Int J Res Orthop. 2023 Mar;9(2):462-466
http://www.ijoro.org

DOI: https://dx.doi.org/10.18203/issn.2455-4510.IntJResOrthop20230484

Case Report

Total hip replacement in a black hip: a case report and review of literature

Vikram Shah[1], Javahir Pachore[2], Jayesh Patil[1], Pranay Gujjar[1], Amit Kumar Sinha[1]*

[1]Shalby Hospital, Ahmedabad, Gujarat, India
[2]Department of Hip Replacement Surgery, Shalby hospital, Ahmedabad, Gujarat, India

Received: 15 December 2022
Accepted: 08 January 2023

***Correspondence:**
Dr. Amit Kumar Sinha,
E-mail: amitsinhamgims@gmail.com

ABSTRACT

Alkaptonuria is a rare metabolic disease leading to the accumulation of a blue-black pigment namely homogentisic acid in the cartilaginous tissue and body fluids giving them a black color. It is an autosomal recessive disease due to the deficiency of the hepatic enzyme oxidase which results in the accumulation of homogentisic acid in the skin, cartilage, and collagenous tissue giving them a black color. Herein we report a case of 65 years old gentleman who presented to our emergency department post domestic fall on his left hip, after which pain and swelling developed around the left hip and he was unable to bear weight with restricted mobility around the same.

Keywords: Alkaptonuria, Total hip replacement, Black hip

INTRODUCTION

Alkaptonuria is a rare metabolic disease leading to the accumulation of a blue-black pigment namely homogentisic acid in the cartilaginous tissue and body fluids giving them a black color. It has a prevalence in the range of 1 in 5 million to 1 in 20 million people.[1]

Though rare in the Indian scenario, the disease is reported in a high frequency in Slovakia and the Dominican Republic.[2,3]

It is an autosomal recessive disease due to the deficiency of the hepatic enzyme oxidase which results in the accumulation of homogentisic acid in the skin, cartilage, and collagenous tissue giving them a black color (Figure 1).[4]

This accumulation results in the discoloration of body fluids namely sweat and urine to black color and degeneration of joints, particularly the shoulders, spine, hips, and knees.

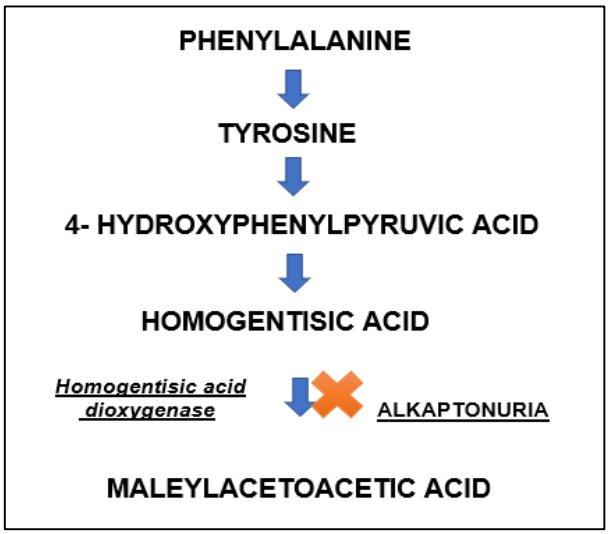

Figure 1: The pathway for the metabolism of phenylalanine. A deficiency of oxidase enzyme leads to failure in the oxidation of homogentisic acid leading to alkaptonuria.

CASE REPORT

Herein we report a case of 65 years old gentleman who presented to our emergency department post domestic fall on his left hip, after which pain and swelling developed around the left hip and he was unable to bear weight with restricted mobility around the same. The ipsilateral knee and other hips and knees were clinically normal.

On further questioning, he revealed that previously he had pain in both his hips and knees and could walk without support but with difficulty. He also gave a history of blackish discoloration of his urine and his clothes staining black with his sweat. On Examination, there was bluish-black discoloration of both his palms, and blackish discoloration of both pinna and sclera as well (Figure 2).

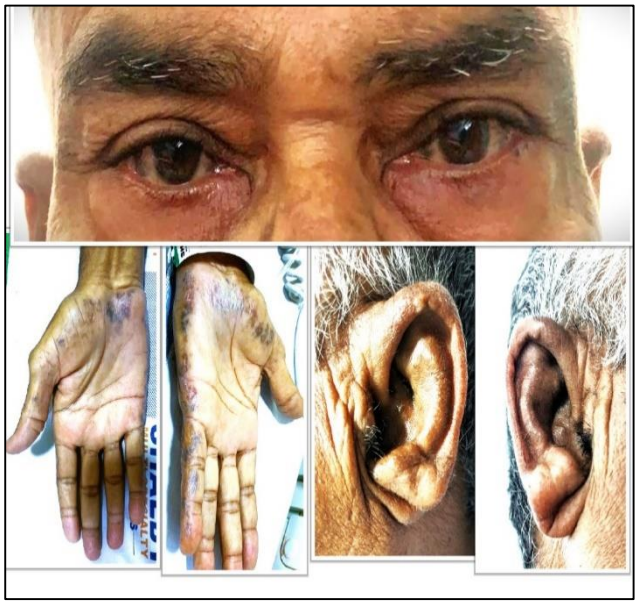

Figure 2: Bluish black discoloration of both palms, right pinna, and sclera.

He was evaluated radiologically which showed a fracture left neck of the femur, reduction of medial joint space in both of his knees, and intervertebral disc calcification with narrowing of disc spaces (Figure 3).

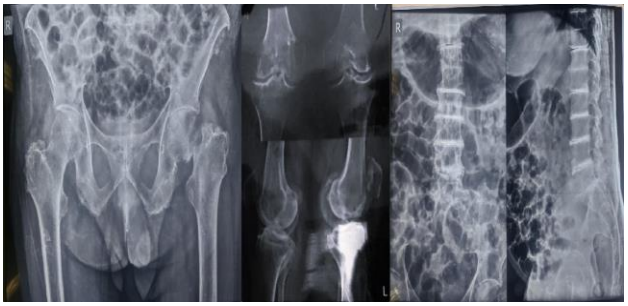

Figure 3: Radiograph showing fracture left neck of the femur, intervertebral disc calcification with reduction of lumbar lordosis and bilateral knee osteoarthritis.

The patient was planned for total hip replacement using the Direct Lateral approach (Uncemented cup and stem; metal on poly; Consensus® hip system, USA) after clearance from Anesthetist and Physician and consent from the patient side. The patient was positioned in a lateral decubitus position. After taking down the skin, superficial fascia, and tensor fascia Lata, the trochanteric bursa was resected. The fibers of the gluteus Medius were split and the anterior aspect of the Gluteus Medius, the anterior part of the Vastus lateralis, and the gluteus minimus was erased off as a sleeve or flap from the trochanteric ridge to expose the anterior capsule which had tinges of black. A neck cut was taken and the head extracted was carbon Black in color (Figure 4).

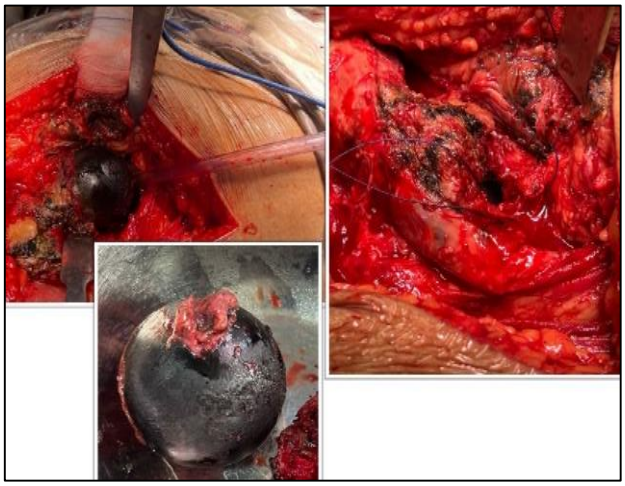

Figure 4: Blackish discoloration of capsule and head (carbon black).

Femoral part preparation with the positioning of appropriate retractors and trialing with appropriate sizes were done. The acetabulum that was exposed had hues of 'black'. The cartilage which came out after reaming was also black in color (Figure 5).

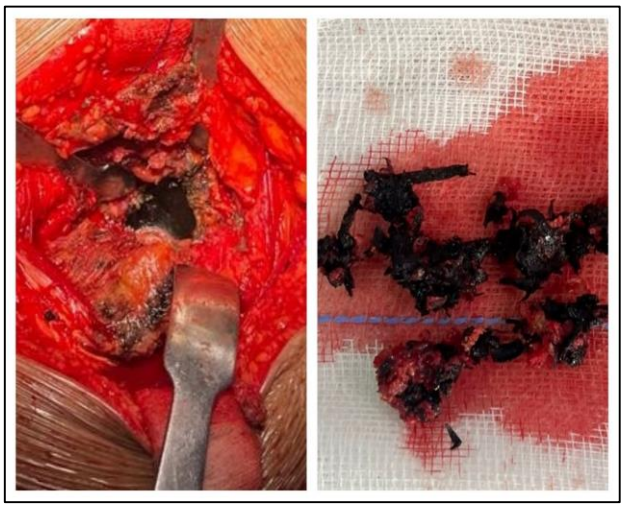

Figure 5: Acetabulum along with the cartilage obtained from reaming which was black in color.

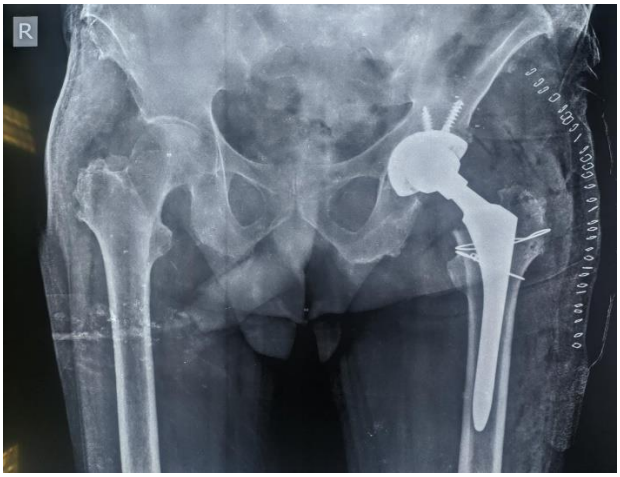

Figure 6: Post-op radiograph showing cerclage wiring and good positioning of implants.

After an appropriate reaming & trial of the cup, the original cup and liner were inserted. The original stem was positioned (during which anterior cortex was partially split vertically at the time of final stem insertion for which cerclage with two wires was done), trialing with the head was done followed by implantation of metal head and reduction of hip. After the closure of the hip capsule osteotendinous approximation of the gluteus Medius and minimus to the trochanteric ridge via two drill holes made in the trochanteric area and with the use of an 'OS' needle (Orthopedic surgeon needle) was achieved. Rest, closure was done in layers, and the patient was advised non-weight bearing (toe touch allowed) for 6 weeks. An immediate post-operative radiograph was taken which showed good positioning of implants (Consensus®, USA) (Figure 6). The diagnosis of the condition was also confirmed via histopathological examination of the tissues which showed multiple pigmented areas with reactive giant cells which are characteristic of the disease.

DISCUSSION

Alkaptonuria is a rare metabolic disease due to deficiency of the enzyme alkapton oxidase leading to accumulation of the pigment homogentisic acid which then accumulates in the cartilages of the joints leading to degenerative changes and what is referred to as "ochronotic arthropathy".[5] Patients with the disease usually lead an uneventful childhood though hyperpigmentation of urine may be seen in childhood as well.[6] Ochronosis can also be induced via exogenous factors like phenol, benzene, and hydroquinone; however, the difference is that classical arthropathy is absent in exogenous alkaptonuria.

Ochronotic arthropathy classically manifests during the third to fourth decade probably hypothesized to decreased renal clearance of the pigment with advancing age.[7] The most common areas of affection are the vertebral column manifesting as calcification of the intervertebral discs, narrowing of the disc spaces, and arthropathy of the peripheral joints particularly the hips and the knees limiting mobility and causing restriction of the activities of daily living.

Table 1: Literature review.

Articles	Year	Age/ gender (Years)	Joint	Comments	Follow up (Years)	Results
Efram et al[8]	2022	50/M	B/L knees	---	6 m	Excellent
G S et al[9]	2021	57/M	B/L knees	Extensor tendon risk of rupture; Difficult spinal anaesthesia	18 m	Excellent
Paaskesen et al[10]	2020	72/M	U/L hip + U/L Knee	---	---	Good
Zimmerly et al[11]	2019	66/M	U/L hip +U/L knee	Surgical, anesthesiological, and postoperative management require more vigilance	---	Excellent
Roca B et al.12	2016	78/M	----	Osteoarticular changes and aortic stenosis		Good
Gowda N et al[13]	2013	60/F	U/L Hip	Influence of vitamin C; Joint replacement in severe arthritis	2	Excellent
Pandey et al[14]	2011	56/M	Revision hip	Thorough preoperative evaluation in patients of alkaptonuria	--	Good
Zacharia et al[15]	2009	65/F	Excision arthroplasty	Fracture of Neck femur with scoliosis	---	Good
Ogata et al[16]	2008	75/F	U/L hip	Dura and arachnoid membranes could be damaged and made vulnerable by HGA	---	Excellent
Shimizu et al[17]	2007	74/F	U/L hip	"Fragmentation and cleft formation" in the cartilage of the femoral head	---	Good

Continued.

Articles	Year	Age/ gender (Years)	Joint	Comments	Follow up (Years)	Results
Kerimoglu et al[18]	2005	--	4 cases of U/L hip	Cementless total hip prosthesis	---	Good
Fisher et al[19]	2004	69/F	B/L knee + U/L hip	Severe aortic stenosis requiring valve replacement, low trauma fracture of the distal femur despite two years of alendronate therapy	5	Improvement
Moslavac et al[20]	2003	70/M	B/L knee + U/L hip	----	7	Excellent
Ayna[21]	2000	53/F	B/L hip	HLA-B27(+) woman with ochronotic arthropathy	---	Excellent
Dom et al[22]	1997	63/M	U/L hip	----	--	Good
Corrà et al[23]	1995	58/F	U/L hip	---	---	Satisfactory

Not only the musculoskeletal system, but reports are there of alkaptonuria affecting the cardiovascular system requiring urgent replacement of aortic valves and ischemic heart disease.[24]

No specific treatment has been found to date apart from focused treatment on reducing the intake of diet containing phenylalanine and tyrosine which are products from which homogentisic acid is derived and increasing the intake of ascorbic acid which has varying or no strong clinical correlation in literature. The destruction of articular cartilage in patients with alkaptonuria is extensive and appears at an average age of 55 to 60 years necessitating joint replacement which has shown good results in a review of case reports published (Table 1).

Spencer et al described 11 joint replacements in 3 patients with alkaptonuria polyarthropathy, including shoulder and elbow replacements not previously reported.[25] No prosthetic failures occurred in up to 12 years of follow-up.

Aydoğdu et al reported a 48-year-old man who had been treated with cementless total knee arthroplasty.[26] The 4-year follow-up of cementless total knee arthroplasty was satisfactory without any evidence of loosening.

Wang et al reported a 64-year male who underwent one-stage revision arthroplasty in a knee infected with *Achromobacter xylosoxidans.*[27]

Karaoğlu et al reported a 10-year follow-up in a 55-year-old man who underwent cemented knee replacement.[28]

CONCLUSION

Our literature review found no cases of revision of Joints done in patients of alkaptonuria. Since there is no treatment for this ailment at present, Total joint replacement seems to be an effective modality in improving upon the activities of daily living in these patients giving them a new quality of life with significant improvement in their activities of daily living.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Laskar FII, Sargison KD. Ochronotic arthropathy. J Bone Joint Surg. 1970;52B:653-4.
2. O'Brien WM, La Du BN, Bunim JJ. Biochemical, pathologic and clinical aspects of alcaptonuria, Ochronosis, and ochronotic arthropathy: a review of world literature (1584-1962). Am J Med. 1963;34(6):813-38.
3. Milch RA. Studies of alcaptonuria: inheritance of 47 cases in eight highly inter-related Dominican kindreds. Am J Hum Genet. 1960;12(1):76-85.
4. Virchow R. Ein Fall von allgemeiner Ochronose der Knorpel und knorpelahnlichen Theile. Virchows Arch. 1866;37:212-9.
5. O'Brien W, La Du BN, Bunim JJ: Biochemical, pathologic and clinical aspects of alcaptonuria, ochronosis, and ochronotic arthropathy. Am J Med. 1963;34:813-38.
6. Al-Mefraji SH, Al-Mefraji. Alkaptonuria in a 5-year–old boy in Iraq. East Mediterr Health J. 2008;14(3):745-6.
7. Fisher AA, Davis MW. Alkaptonuric ochronosis with aortic valve and joint replacements and femoral fracture: a case report and literature review. Clin Med Res. 2004;2(4):209-15.
8. Elafram R, Ammou AB, Romdhane MB, Sghaier M. Alkaptunuria; a case report. Int J Surg Case Rep. 2022 Nov;100:107682.
9. GS, John JT, Nair DS, Yadav S, Varghese J, T J. Ochronotic Surprise during Total Knee Replacement! A Case Report. J Orthop Case Rep. 2021;11(10):49-52.
10. Paaskesen CK, Hofbauer C. Alkaptonuria detected during knee arthroplasty treatment]. Ugeskr Laeger. 2020;182(37):V04200271.
11. Zmerly H, Moscato M, Di Gregori V. Arthroplasty in alkaptonuric ochronosis. J Popul Ther Clin Pharmacol. 2019;26(2):e20-e24.

12. Roca B, Roca M, Monferrer R. Alkaptonuria Presenting with Impressive Osteoarticular Changes and Severe Aortic Stenosis. Conn Med. 2016;80(3):139-41.
13. Gowda N, Kumar MJ, Kumar AK. Black hip: a rare case treated by total hip replacement. Ann Saudi Med. 2013;33(4):368-71.
14. Pandey R, Kumar A, Garg R, Khanna P, Darlong V. Perioperative management of patient with alkaptonuria and associated multiple comorbidities. J Anaesthesiol Clin Pharmacol. 2011;27(2):259-61.
15. Zacharia B, Chundarathil J, Ramakrishnan V, Krishnankutty RM, Veluthedath R, Puthezhath K, Varughese I. Black hip, fracture neck of femur and scoliosis: a case of ochronosis. J Inherit Metab Dis. 2009;32(1):S215-20.
16. Ogata J, Tamura K, Miyanishi K, Minami K, Haranishi Y, Tsubaki T. Anesthesia in a patient with alkaptonuric ochronosis for total hip arthroplasty]. Masui. 2008;57(4):439-42.
17. Shimizu I, Hamada T, Khalpey Z, Miyanishi K, Hara T. Ochronotic arthropathy: pathological evidence of acute destruction of the hip joint. Clin Rheumatol. 2007;26(7):1189-91.
18. Kerimoglu S, Onder C, Aynaci O, Malkoc CH. Hip arthroplasty for ochronosis. Saudi Med J. 2005;26(11):1812-4.
19. Fisher AA, Davis MW. Alkaptonuric ochronosis with aortic valve and joint replacements and femoral fracture: a case report and literature review. Clin Med Res. 2004;2(4):209-15.
20. Moslavac A, Moslavac S, Cop R. Case report of a patient with ochronosis and arthroplasty of the hip and both knees. Reumatizam. 2003;50(1):26-8.
21. Aynaci O, Onder C, Turhan AU. Bilateral hip arthroplasty for ochronotic arthropathy. Clin Rheumatol. 2000;19(2):150-2.
22. Dom K, Pittevils T. Ochronotic arthropathy: the black hip. Case report and review of the literature. Acta Orthop Belg. 1997;63(2):122-5.
23. Corrà T, Zaccala M, Galante M. Ochronotic arthropathy: rapid destructive hip osteoarthritis associated with metabolic disease. Clin Rheumatol. 1995;14(4):474-7.
24. Vavuranakis M, Triantafillidi H, Stefanadis C. Aortic stenosis and coronary artery disease caused by alkaptonuria, a rare genetic metabolic syndrome. Cardiology. 1998;90(4):302-4.
25. Spencer JM, Gibbons CL, Sharp RJ, Carr AJ, Athanasou NA. Arthroplasty for ochronotic arthritis: no failure of 11 replacements in 3 patients followed 6-12 years. Acta Orthop Scand. 2004;75(3):355-8.
26. Aydoğdu S, Cullu E, Ozsoy MH, Sur H. Cementless total knee arthroplasty in ochronotic arthropathy: a case report with a 4-year follow-up. J Arthroplasty. 2000;15(4):539-43.
27. Wang XC, Zhang XM, Cai WL, Li Z, Ma C, Liu YH et al. One-stage revision arthroplasty in a patient with ochronotic arthropathy accompanied by joint infection: A case report. World J Clin Cases. 2022;10(25):9036-43.
28. Karaoğlu S, Karaaslan F, Mermerkaya MU. The long-term result of arthroplasty in the treatment of a case of ochronotic arthropathy. Acta Orthop Traumatol Turc. 2016;50(5):584-6.

Cite this article as: Shah V, Pachore J, Patil J, Gujjar P, Sinha AK. Total hip replacement in a black hip: a case report and review of literature. Int J Res Orthop 2023;9:462-6.