

Clinical and Surgical Insights on Bilateral Total Knee Arthroplasty in Ochronotic Arthropathy: A Case-based Review

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Learning Point of the Article:

Simultaneous bilateral total knee arthroplasty is safe and effective treatment for end-stage ochronotic arthropathy of knee; however, a thorough history and clinical examination with a very high index of clinical suspicion are needed for pre-operative diagnosis and anticipation of potential complications during the surgery and early rehabilitation.

Abstract

Introduction: Ochronotic arthropathy (Oca) is a consequence of alkaptonuria, a rare systemic-inherited metabolic disorder leading to accumulation of homogentisic acid in articular cartilage and subsequent early degeneration of the joints. Only few cases of Oca managed with bilateral total knee arthroplasty (TKA) has been described in the literature so far. We aim to discuss surgical pearls, pitfalls, and clinical outcome of Oca of knees managed with simultaneous bilateral TKA.

Case Presentation: The patient was a 52-year-old female presented to outpatient facility with severe bilateral knee pain and difficult ambulation. After clinical and radiological diagnosis of osteoarthritis, she was posted for bilateral TKA. During arthrotomy, blackening of articular cartilage, quadriceps and patellar tendon, and synovium was noted. Subchondral bone was free of pigmentation though, seemed osteopenic while taking bone cuts. Right knee was implanted with cruciate retaining components with ultracongruent insert; while on the left side, posterior stabilized components were used. Diagnosis of ochronosis was made retrospectively with characteristic subtle pigmentation of nails, palms, pinnae, and urine homogentisic levels. Two years follow-up of the patient remained satisfactory without any major complications.

Conclusion: Oca may present with intra-operative surprise to the surgeon if not anticipated preoperatively. Simultaneous bilateral TKA is safe and effective treatment for end-stage arthritis. However, it is difficult to diagnose it preoperatively; a high clinical suspicion leads to meticulous assessment and counseling regarding potential anesthetic concerns, choice of implant, and possible intra-operative and post-operative complications reported sporadically though.

Keywords: Alkaptonuria, blackjoints, ochronosis, ochronotic arthropathy, total knee arthroplasty.

Introduction

Alkaptonuria (OMIM 203500) is one of the autosomal recessive metabolic disorders with global incidence reported to occur as 1:100,000–1:250,000 live births [1]. It was described by Archibald Garrod in 1902 as the first disorder in human known to follow the principles of autosomal recessive Mendelian inheritance [1]. Ochronosis is a Greek word which literally means yellowish discoloration (ocher-yellow), given after

microscopic appearance of yellowish discoloration of tissues and first described by Virchow in 1866. Alkaptonuria occurs due to deficiency of enzyme homogentisic acid (HA) 1,2-dioxygenase in tyrosine metabolism characterized by accumulation of HA as high as 100 folds. This leads to oxidative conversion of HA to melanin like polymer, also known as ochronotic pigment (blackish-brown) which has higher affinity for collagen rich connective tissues such as subcutaneous tissue, cartilage, tendon,

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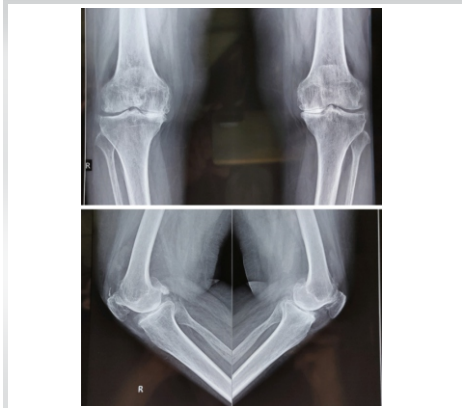


Figure 1: Pre-operative radiograph of both knee joints showing loss of medial joint space, osteophytes, subchondral sclerosis, varus deformity, and patellofemoral osteophytes suggestive of end-stage arthritis.

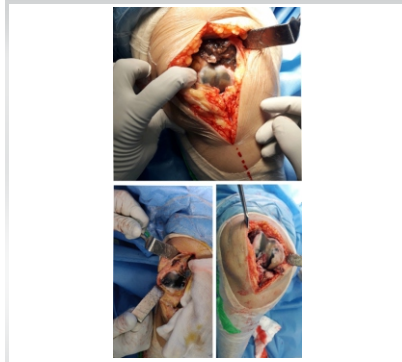


Figure 2: Intra-operative photographs showing synovial pigmentation and hypertrophy (top), discoloration and degeneration of patellar cartilage, patellar tendon and quadriceps tendon pigmentation (bottom left) and characteristic pigmentation of tibiofemoral cartilage, menisci, and cruciate ligaments.



Figure 3: Immediate post-operative radiograph depicting well cemented, mechanically aligned cruciate retaining prosthesis for the right knee and posterior stabilized for the left knee.

ligament, sclera, intervertebral disc (IVD), heart valves, and intima of blood vessels [2]. Excessive HA is excreted in urine so, when urine is exposed to air, the color turns dark because of oxidation of metabolic by-products to the polymer.

The deposition of ochronotic pigment leads to attenuation in the strength of connective tissue due to alteration in mechanical properties. This leads to synovial inflammation, early degeneration of cartilage, ligaments, and tendinous structures around joints. The patients usually remain asymptomatic until 4th–5th decade when they present with multiple joints pain or chronic back pain secondary to arthritis changes in the peripheral or discovertebral joints [3]. The patients may have blackish or gray-bluish discoloration of pinnae, palms, eyelids, sclera, forehead, cheeks, axillae, nail beds, nasal tip, and soles. Simultaneous involvement of intima of blood vessels and heart walls is not infrequent. Deposition in IVDs leads to degenerative disc disease with calcification of the same. Among peripheral joints, knee is the most commonly affected joint followed by hip, shoulder, and sacroiliac joint [3].

Case Presentation

A 52-year-old female patient presented to our outpatient facility

with complaints of bilateral knee pain and swelling for the past 4–5 years. The pain was insidious in onset and disabling enough to interfere in her activities of daily living, not associated with any constitutional symptom. She had a history of lower back surgery 5 years ago for lumbar disc prolapse and had mild lower back pain on and off post-surgery. The right knee was more symptomatic than the left one. On examination, medial and lateral joint line tenderness along with patella-femoral tenderness was noted on both knees. There was significant synovial thickening along with moderate bilateral knee joint effusion which was confirmed with patellar tap. The range of motion of the right knee was 0–80° on the right and 0–90° on the left side with significant pain on 20–30° of terminal flexion. Both knee joints had about 10° of varus deformity which was partially correctable and the patient walked with antalgic gait. The anteroposterior and lateral radiographs of knee joints depicted gross narrowing of medial tibiofemoral joint spaces with subchondral sclerosis, patellofemoral and posterior recess osteophytes and overall varus alignment of both knee joints (Fig. 1).

A diagnosis of bilateral knee osteoarthritis (OA) was made and the patient was posted for bilateral total knee arthroplasty

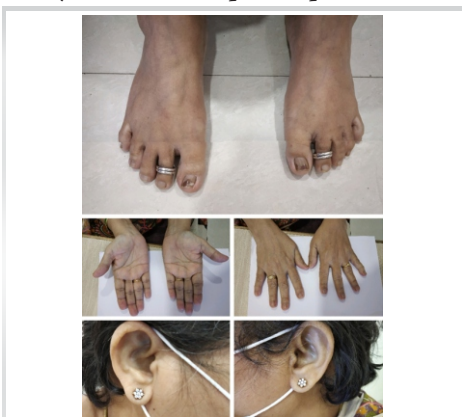


Figure 4: Clinical images showing characteristic discoloration of toe nails and dorsum of feet (top), palms and finger nails (middle), and ear pinnae (bottom).



Figure 5: Roentgenogram of lumbosacral spine and pelvis depicting diffuse osteopenia of vertebral bodies with endplate sclerosis, narrowing of disc spaces, and calcification of intervertebral discs and sclerotic-degenerative changes at symphysis pubis.



Figure 6: Follow-up radiograph (at 24 months post-operative) of both knee joints showing well aligned components without obvious radiolucent lines, osteolysis, wear, or loosening around the components.



TKA) after prior surgical clearance from an internist, cardiologist, and anesthesiologist as per protocol. During the first (right) knee surgery, there was difficulty in spinal anesthesia due to stiff spine as reported by the anesthetist. Under spinal anesthesia, after medial parapatellar arthrotomy, diffuse black discoloration of the tibiofemoral articular cartilage, menisci, joint capsule, synovium, and quadriceps tendon was noted with degenerative changes in the cartilage (Fig. 2). The synovium was hypertrophic and of dark brownish-black color. Subtotal synovectomy was done and bone cuts were completed following standard measured resection technique and trial components were inserted.

The subchondral bone was normal in appearance; however it seemed to be osteopenic than those of routine cases. The strength of ligaments and tendons seemed to be normal. During components trial, we noted positive lift-off sign (booking) of tibial tray at 70–90° of flexion which was resolved with partial release of posterior cruciate ligament (PCL) at femoral attachment. The cruciate retaining (CR) femoral and tibial components were cemented in routine manner. We used ultracongruent polyethylene liner (Curved Plus, PFC sigma, and Depuy-Synthes Warsaw Indiana) due to its proven efficiency in clinical studies [4, 5]. No excessive bleeding was noted after synovectomy and hemostasis was achieved following tourniquet deflation before closure. Drains were not used. The left knee arthroplasty was also done in similar manner except superficial medial collateral ligament (MCL) release and using posterior stabilized (PS) components. Patellofemoral OA was managed with removal of osteophytes and circumferential denervation with electrocautery on both sides. Post-operative radiographs were satisfactory in regard to component placement and alignment (Fig. 3).

The diagnosis of ochronosis was confirmed retrospectively in post-operative period, while the patient revealed history of black discoloration of her urine and of family members. Careful physical examination revealed subtle blackish discoloration of her palms, helices of ear pinna, finger nails, toe nails, and skin of dorsum of the feet (Fig. 4). Her lumbosacral spine roentgenograms depicted diffuse osteopenia of vertebral bodies with endplate sclerosis, narrowing of disc spaces, and calcification of IVDs while pelvis depicted characteristic degenerative changes and sclerosis at symphysis pubis (Fig. 5). Her high urine HA levels confirmed the diagnosis.

In post-operative period, the patient had mild-to-moderate pain, which was managed with standard multimodal analgesic protocol. She was mobilized full weight bearing with walking frame on post-operative day 1. Her post-operative wound healing was uneventful and standard rehabilitation protocol was followed. Sutures were removed after 2 weeks and the patient

remained satisfied with the surgery. At the latest (24 months post-operative) follow-up, the patient was symptom free and the ranges of motion were 0–110° on the right side and 0–120° on the left side. The knee society score (KSS) was 82 and 84 on the right and left side respectively and the knee function score was recorded to be 90. The follow-up radiographs showed neutral mechanical alignment with absence of radiolucent lines, osteolysis, wear, or loosening around the components (Fig. 6).

Discussion

Ochronotic arthropathy (Oca) is a consequence of alkaptonuria, a rare systemic-inherited metabolic disorder leading to accumulation of HA in articular cartilage and subsequent early degeneration of the joints. Knee and hip joints are the most involved joints in Oca and typically the patient presents in their 5th–6th decade of life [3]. The diagnosis in our case was missed pre-operative and it came as an intra-operative surprise (due to unexpected black discoloration of the joint and synovium after the arthrotomy) which later confirmed with characteristic history, physical examination, findings of the previous discectomy surgery and spine radiographs, and urine HA levels. However, the articular cartilage remains loaded with the high levels of HA, the subchondral bone remains spared of any discoloration. Similarly, the patellar tendon, MCL, PCL, and joint capsule showed pigmentation, yet the integrity and strength were intact.

In a similar case report by Patel VG, osteopenic nature of subchondral bone and fragile tendency of patellar tendon have been described during TKA for Oca of knee. The author also reported increased blood loss (around 1150 ml total) than usual due to extensive synovectomy which caused delayed drain removal not necessitating blood transfusion [6]. Spontaneous rupture of quadriceps tendon has also been reported in few other reports [7, 8, 9]. In recently published similar report by Al Dosari et al., the authors did not report any increase in blood loss during the surgery [10]. The author also emphasized that since the integrity of ligaments was intact, the patient was implanted with the CR TKA prosthesis. Good functional outcome was reported by the author in the case.

While our knowledge of surgical treatment of chronic arthritis has been limited to few case reports only [11, 12, 13] with relatively short follow-up, Rajkumar et al. recently published the largest case series involving 27 joint arthroplasties in 16 patients with a mean follow-up of 39 months [14]. The author emphasized about the finding of multiple joint involvement with varying severity of arthritis in all patients with eight patients having two or more joints replaced. Furthermore, the author also concluded that the strength of cancellous bone was not compromised in Oca cases and use of uncemented

components in hip arthroplasty cases does not lead to early failure due to loosening. However, in the mentioned case series, one patient had prosthetic joint infection 2 months after index surgery necessitating debridement first and then explantation with two stage revision later; while another patient had aseptic loosening of cemented acetabular component after 10 years post-operative necessitating revision of the component. Lee et al., in their recently published case report with systemic review, concluded that no prosthetic design confers superiority over other in TKA for OcA [15]. However, the authors suggested using PS knee prosthesis because of suspected compromised state of ligaments.

Conclusion

OcA may present with intra-operative surprise to the surgeon if not anticipated preoperatively, particularly in cases with subtle clinical features. Anesthesia considerations include calcified cardiac valves, difficulties in spinal anesthesia due to calcified

disc spaces, and difficult fiberoptic intubation. Total joint arthroplasty is considered best choice of the treatment for end-stage arthritis, while in initial stages of knee arthritis intra-articular steroid injection, synovectomy and arthroscopic debridement have been tried with variable and inconsistent success.

The role of medical management with ascorbic acid and HA lowering agents such as nitrofurantoin remains controversial and does not show any differences in clinical outcomes.

Clinical Message

Simultaneous bilateral TKA is safe and effective treatment for end-stage arthritis. A very high index of clinical suspicion must be there to diagnose it preoperatively and scrupulous assessment and counseling of the patient is a must regarding potential complications though, reported sporadically in few cases till date.

Declaration of patient consent : The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for patient images and other clinical information to be reported in the journal. The patient's parents understand that his 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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