

Diagnosing an ochronotic arthropathy during knee arthroscopy: Management of a Rare Case

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Abstract—A 60-year-old male was admitted to the outpatient clinic with a complaint of pain in the left knee. During the arthroscopy, blackening was observed in the synovium, meniscal tissue and bone surfaces. Because an improvement was not observed after arthroscopy and physical therapy, total knee replacement was applied. After two years of follow-up, patient had full range of motion with no findings of loosening. Ochronosis is one of the rare causes of knee arthritis and considering that the long-term results are satisfactory, the application of total knee replacement should not be avoided in cases where physical therapy is inadequate.

Keywords—Ochronotic arthropathy; blackening; alkaptonuria; loose body; arthroscopy.

I. INTRODUCTION

Alkaptonuria is an autosomal recessively inherited metabolic disease that presents clinical symptoms due to homogentisic acid oxidase deficiency and homogentisic acid (HGA) accumulation in the body [1,2]. HGA accumulates in various connective tissues, such as cartilages, nose, ears and sclera, thus causing a dark coloring/blackening of the tissue, also known as ochronosis [1-4]. By causing the tissues to become fragile and vulnerable to mechanical stress thus resulting in articular cartilage degeneration, ochronosis is one of the rare causes of arthritis, which usually becomes symptomatic in patients aged 40 to 50 [2,4].

The definitive diagnosis of alkaptonuria/ochronosis is made by biochemical tests that quantitatively show homogentisic acid in the urine [2,5]. Because there is no effective treatment for the ochronosis, only management of the disease is treating the related conditions, such as arthritis [2,5,6].

We are presenting a case in which we diagnose an ochronotic arthropathy during knee arthroscopy and the subsequent treatment process.

II. CASE REPORT

A 60-year-old male was admitted to the orthopedics and traumatology outpatient clinic with a complaint of pain in the left knee and a history of locked knee. In his story, he stated that he has knee pain for quite some time but got worse lately and was preventing him from doing his daily work. He has also stated that his knee got locked two times in the last three months and got loosen by the manipulative maneuvers. Physical



Fig. 1: Pre-operative anteroposterior and lateral radiography images of the patient

examination revealed tenderness in the left knee with a positive McMurray test. Flexion range of motion was 125 degrees whereas the extension range of motion was full. All distal pulses were palpable and there were no neurological symptoms. Radiographic examination revealed degenerative changes and narrowing of the joint space on both knees (Fig. 1). Magnetic resonance imaging revealed a rupture in the lateral meniscus posterior horn, with diffuse subchondral lesions in the femur and tibia, increased fluid in the suprapatellar bursa and patellar chondromalacia.

Considering the patient's story and findings, diagnostic arthroscopy was planned. During the arthroscopy, blackening was observed in the synovium, meniscal tissue and bone surfaces. Furthermore, several loose bodies in brown-black color were seen. Loose bodies were removed as much as possible and the pathological samples were taken from various tissues for examination. Widespread degenerative changes were observed in the posterior horn of the lateral meniscus and grade four cartilage damage was observed on the lateral and medial condyles and patellar surfaces. The operation was concluded after partial meniscectomy with debridement.

The patient was diagnosed as ochronotic arthropathy after histopathological examination of arthroscopic biopsy specimens. He had no complaints regarding alkaptonuria/ochronosis and no other blackening was observed anywhere in his body.

Medical treatment was initiated after surgery and patient was referred to a physical therapy and rehabilitation center at the third week follow-up, with a diagnosis of ochronotic arthropathy-related persistent knee pain after arthroscopy.

After six weeks of physical therapy, the patient was referred to us with the recommendation of knee arthroplasty surgery, considering that he did not benefit from medical treatment and rehabilitation, as his pain was still existed. After physical examination, patient was diagnosed as ochronotic arthropathy-related gonarthrosis and total knee replacement surgery was performed with the consent of the patient. During the surgery, significant blackening was observed in patella, femoral condyles, tibia condyles and joint capsules (Fig. 2). Knee was prepared for implantation after appropriate osteotomies and extensive washing with saline solution, and components was placed. All osteotomy materials and soft tissues were sent to histopathological examination. Surgery was concluded after suturing.

Patient was discharged after mobilization, with a clean wound site and appropriate rehabilitation program. At 2-years follow-up, patient's physical examination and radiographic imaging revealed a full range of motion, with no complaints, no complications and no findings of loosening (Fig. 3).

III. DISCUSSION

Knee pain is one of the leading causes for admissions to orthopedics and traumatology outpatient clinics. Although the differential diagnosis varies according to age; sprain and strain injuries of the knee, meniscal tears and arthritis are among the most common causes of knee pain. Although arthritis can be caused due to many different reasons, osteoarthritis, in which old age plays an important role, is usually the first reason that comes to mind. Among the less common causes, alkaptonuria/ochronosis takes an important place. With an estimated incidence of 1:250.000 to 1:1.000.000, ochronosis is a relatively rare cause of arthritis [2,7]. To our knowledge, more than 200 cases of ochronotic arthropathy were reported in the literature, with less than 50 cases reported in the last five years and most of them being related to either spine or knee joint [8-15]. Thus, it is important to record such cases and this is the main strength of this case report.

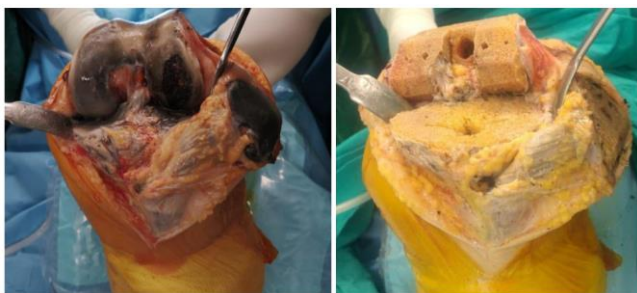


Fig. 2: Intraoperative images of the patient's knee before and after cleaning the ochronotic tissues and performing osteotomies during total knee replacement surgery



Fig. 3: After 2 years of follow-up, no signs of complication and loosening were observed in the patient's examination and radiographic imaging.

Although ochronotic arthropathy can be diagnosed during arthroscopy, by visualizing the blackening of the synovium and other connective tissues of the related joint, the definitive diagnosis is made by histopathological examination [1,6,9,16,17]. In our case, even though we have not included ochronotic arthropathy in our differential diagnoses, we have diagnosed ochronosis during the diagnostic arthroscopy and confirmed our diagnosis with histopathological examination.

The treatment of ochronotic arthropathy is similar to that of other arthritis causes; physical therapy and lifestyle recommendations playing an important role in the first step of treatment and joint replacement usually being the most effective treatment method [2,6,18,19]. In our case, after six weeks of physical therapy and medical treatment, patient still had knee pain and difficulty walking. Thus, it was inevitable to apply total knee replacement surgery to our patient.

Aydogdu et al. have reported a case of ochronotic arthropathy with a 4-year follow-up. They have reported satisfactory results with no complications after 4 years with a total knee replacement surgery [18]. Ilyas et al. have reported a 45-years-old ochronotic arthropathy with a 12 -year follow-up and stated full range of motion with no findings of loosening [20]. In our case, patient had full range of motion with no complaints and no findings of implant loosening after 2 years of follow-up. Our results are consistent with the literature.

In conclusion, ochronosis is one of the rare causes of knee arthritis and should be kept in mind in the differential diagnosis of knee pain, especially over the age of 40. The most important step in its diagnosis is arthroscopy, and the first step of the treatment is physical therapy and medical treatment. Considering that the long-term results are satisfactory, the application of total joint replacement should not be avoided in cases where physical therapy is inadequate.

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