

Ochronotic black meniscus during knee arthroscopy

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Abstract

Ochronotic arthropathy is a rare condition found in patients with alkaptonuria, which is a hereditary metabolic disease associated with deposition of homogentisic acid derivatives in various connective tissues of the body. We present the case of a 30-year-old woman in whom arthroscopic examination of the left knee prior to meniscectomy for bucket handle tear of medial meniscus revealed brown-black discoloration of the articular cartilage and menisci leading to the diagnosis of alkaptonuria by further laboratory evaluation. After medical and surgical treatment, patient's complaints were alleviated and no further complaints were registered, during the next follow-up.

Key words: *Alkaptonuria, arthroscopy, knee joint, ochronosis*

INTRODUCTION

Alkaptonuria is a rare autosomal recessive disease, resulting from deficiency of the enzyme homogentisic acid oxidase, which is involved in the metabolism of phenylalanine and tyrosine aminoacids. Due to partial or total deficiency of homogentisic acid oxidase in the liver and kidneys, homogentisic acid accumulates in blood, part being excreted in the urine.^[1] On exposure to the atmosphere, urine containing homogentisic acid gradually darkens to a blueish black color. Homogentisic acid and its oxidation products accumulate especially in the cartilage and tendons. It is also reported to be deposited in the skin, tympanic membrane, heart valves, sclera, cartilages of nose and ears, kidneys, pancreas, and large vessels.^[2] Due to accumulation of homogentisic acid, cartilages get a dark discoloration and become brittle and more vulnerable to mechanical stress. The diminished quality of the cartilage causes arthropathy of large joints and spine. The development of degenerative arthritis with accompanying brownish black pigmentation of the tissues and dark discoloration of the urine is called "ochronosis."

The clinical symptoms of ochronotic arthropathy generally begin after the fourth decade of life. Clinical diagnosis is made on the basis of skeletal manifestations including lower back pain, stiffness, and degenerative arthropathy of the large joints, ochronotic pigmentation of the tissues and blueish black discoloration of urine. The definitive diagnosis of the alkaptonuria may be confirmed with several biochemical tests for the quantitative presence of homogentisic acid in urine. Since there is no effective medical treatment for alkaptonuria, treatment at present

is symptomatic by local heat, physiotherapy, analgesics, external support and with surgical treatment in the form of synovectomy, arthroscopic debridement, fusion, or arthroplasty as indicated regarding the affected joint.^[3,4]

We present a patient who was diagnosed to have alkaptonuria during further laboratory tests after observing ochronotic brownish black pigmentation of the menisci and femoral cartilage during arthroscopy of the knee.

CASE REPORT

A 30-year-old woman presented with complaints of pain in the left knee along with occasional episodes of locking for 1 year. Her physical examination revealed tenderness on the medial joint line and positive McMurray's test. Ordinary x-ray images of the knees were found to be normal. Magnetic resonance imaging (MRI) of the left knee revealed bucket handle tear of medial meniscus along with partial tear of anterior cruciate ligament. She was planned for arthroscopic partial meniscectomy in view of her complaints and MRI findings. During the arthroscopic examination of her left knee joint there was mild synovial hypertrophy and a bucket handle tear was found in the medial meniscus that appeared to be brownish black in color [Figures 1, 2]. Anterior cruciate ligament was also found to be partially torn. Lateral meniscus and articular cartilage overlying medial femoral condyle also showed brownish black discoloration. Meniscectomy for bucket handle tear of medial meniscus was done and specimen was sent for histopathological examination.

Further investigations were planned for the diagnosis of ochronosis considering discoloration of the joint cartilage and menisci. The patient's freshly passed urine turned dark brown after addition of sodium hydroxide to the sample to facilitate oxidation. Ferric chloride test was also found to be positive. Histopathological examination of the excised tissue showed pigmentation and degenerative changes [Figure 3]. No pathological findings were detected with the detailed examination of the ear, sclera, skin, and cardiovascular system. Further treatment was done in the form of physiotherapy and by administering vitamin C and analgesics. After the medical and surgical treatment, the patient's complaints were alleviated and no complaints were registered, during the next follow-up.

DISCUSSION

Few reports of arthroscopic diagnosis and treatment of ochronosis have been found in the literature.^[4-8] Our patient is the fifth such case who was diagnosed arthroscopically.^[4,5,7,9] Chen *et al.*^[5] reported the arthroscopic findings of a 50-year-old male patient who was complaining of pain in both knees. Raaijmakers *et al.*^[4] reported a 49-year-old patient's knee arthroscopy findings. Thacker *et al.*^[7] reported the arthroscopic findings of a 40-year-old man who had arthroscopy of the knee and shoulder and Kural *et al.*^[9] presented a 50-year-old woman in whom arthroscopic examination of the right knee revealed brownish black discoloration of the articular cartilage and menisci leading to the diagnosis of Alkaptonuria by further laboratory examinations. Our arthroscopic findings were consistent with those of other cases in the literature. Generalized synovial hypertrophy accompanying brownish black discoloration of articular cartilage and menisci were our common arthroscopic findings with other reported cases in the literature. Generalized cartilage degeneration was not present in our case probably due to comparatively younger age of our patient. In our case there was an additional meniscus tear unlike in that of Thacker's case^[7] and similar with Chen's,^[5] Kural's^[9] and Raaijmaaker's case.^[4] Partial meniscectomy was performed for the torn medial meniscus. In our case, similar to Raaijmaaker's case^[4] arthroscopy was a diagnostic and treatment tool to remind us of ochronosis and direct us for further research. During arthroscopy, brownish black discoloration of the articular cartilage and menisci warned us to have further research for ochronosis. Macroscopic examination of the urine following alkalization and positive ferric chloride test further established our diagnosis. But we were not able to perform the laboratory tests that determine the level of homogentisic acid in urine and blood due to higher cost involved in these tests.

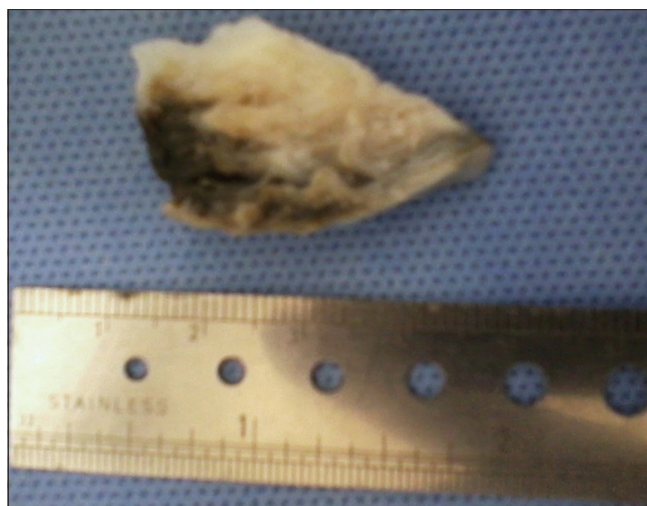


Figure 1: Excised medial meniscus showing brownish black discoloration

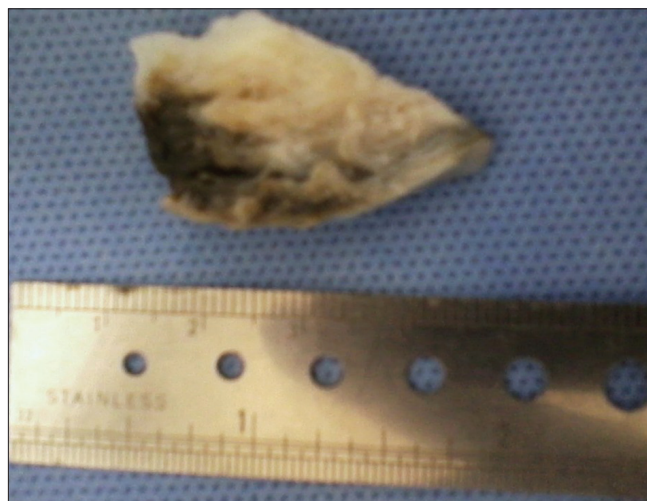


Figure 2: Excised medial meniscus (under surface)

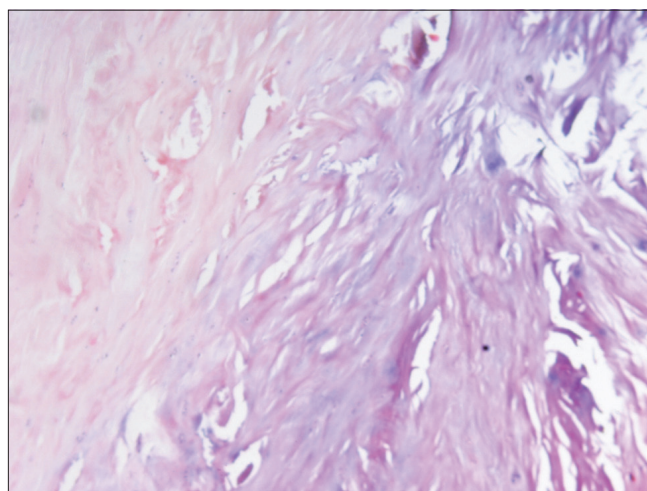


Figure 3: Histopathological section showing pigmentation and degenerative changes (x40)

Approximately 50% of the alkaptonuric patients manifest one clinical form of ochronosis after the fourth

decade. First signs are skin and soft tissue pigmentation. Diagnosis can be made during childhood by observing dark coloration of diapers or urine when left standing. Our patient's mother had noticed the dark color of her urine in early childhood. Ochronotic discoloration of the cartilage of the ears and nose may be observed at the age of 30. Other less common extra articular manifestations are ochronosis of the heart with valve stenosis and urinary tract obstruction by ochronotic calculi. No extra articular manifestations were observed in our patient. No curative treatment for this disease exists. Treatment with high dose of ascorbic acid proved to lower the levels of homogentisic acid in excreted urine,^[4] but does not prevent the occurrence of arthropathy. Nitisinone is shown to have significantly altered the excretion of homogentisic acid in urine in both human and rat studies. Nitisinone is suggested to be used for treatment of the tyrosinemia type 1 but its use in ochronotic patients has yet to be proved.^[9] Since there is no effective treatment to replace the deficient enzyme, symptomatic treatment is initiated with NSAIDs, intra-articular steroid injection and ascorbic acid. Arthroscopic debridement, synovectomy, partial, or total joint arthroplasties and fusion are the surgical treatment options.^[10] Arthroplasty seems to be the best choice for patients whose symptoms persist despite conservative treatment. Raaijmakers *et al.*^[4] reported to provide symptomatic relief with administering 400 mg/day chondroitin sulfate and 1500 mg/day glucosamine for 4 months.

Finally, ochronosis is a rare metabolic disorder affecting peripheral joints as well as spine and the soft tissues and arthroscopy has great importance in suspecting and diagnosing ochronosis.

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