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Research Paper

Total Knee Arthroplasty in an Ochranotic Knee a Case Report

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I. INTRODUCTION

Ochranosis is a rare autosomal recessive metabolic disorder caused by deficiency of Homogentisic acid oxidase enzyme that leads to early degeneration of cartilages and connective tissues in patients of Alkaptonuria.[1] The enzyme is produced predominantly by kidney and liver, and responsible for conversion of Homogentisic acid(HGA) into Maleylactoacetic acid, and its deficiency is characterised by excretion of Homogentisic acid in urine, deposition of oxidised Homogentisic acid pigment in articular cartilages, connective tissue, apocrine glands and dermis. This causes dark discoloration of skin, sclera, ear cartilage, joint cartilage and urine. The diagnosis is often delayed because of its low prevalence and mild early symptoms. In suspicious case, adding Lime(Sodium hydroxide-NaOH) to the urine to alkalinize it will produce a dark brown discolouration and confirmation of diagnosis can be done with measurement of HGA levels in urine sample.[2 3]

Ochranosis is caused due to the interference of Homogentisic acid deposits which polymerises and discolours and weakens the connective tissue leading to brittle tissue which eventually gets chronically inflamed, degenerated and leads to osteoarthritis. The patients are usually asymptomatic and arthropathy appears after the fourth decade.[4] The knee is the most common site of orthopaedic abnormality, other sites involved being the hips, shoulder, sacroiliac joint and pubic symphysis.[5] They can also cause lumbar intervertebral disc calcification and disc space narrowing.

There is no definitive cure for ochranosis currently. The usefulness of phenylalanine and tyrosine restricted diet in adults are yet to be demonstrated. Symptomatic treatment with adequate analgesia and physiotherapy is recommended. However, in ochranotic osteoarthritis, total joint replacement reports good results similar to osteoarthritic patients without ochranosis.[6]

II. CASE REPORT

A 52 year old male, presented with 4 year history of pain and stiffness of the right knee. On physical examination, the right knee presented with swelling, joint line tenderness and restricted range of motion with crepitus. There was a fixed flexion deformity of 5 degrees a Genu varus deformity of 10 degrees.

Radiographically, the right knee showed degenerative changes and osteophytes in tibiofemoral and patellofemoral compartments of knee and mild-moderate narrowing of joint spaces, subchondral sclerosis, irregularities of joint surface.

The physical and radiographic findings were consistent with the diagnosis of end stage tri compartmental osteoarthritis with genu varum and fixed flexion deformity. The patient was listed for cemented total knee arthroplasty. During the surgery, significant blackish discolouration of the cartilages and surrounding soft tissues were incidentally discovered. The surgery was uneventful and no increased bleeding was noted. The patient was treated with analgesics and antibiotics and the drain was removed after 24 hours.

Post operatively, physical examination revealed dark pigmentation of sclera . His urine also turned dark on addition of NaOH and his urine HGA level was measured to confirm the diagnosis of Alkaptonuria. After surgery, radiography revealed satisfactory alignment and implant placement. The patient was mobilised after 24 hours. He underwent post operative physiotherapy as per institute protocols . The Fixed flexion and genu varum deformity was corrected and range of motion in right knee was satisfactory and comparable to normal knee.

III. DISCUSSION

Ochranosis is a musculoskeletal manifestation seen in patients with alkaptonuria. Alkaptonuria is a rare autosomal recessive metabolic disorder with prevalence of 1:19000 to 1:1,000,000.[7] Patients with alkaptonuria have mutation in chromosome 3 between 3q21 and 3q23, which encodes for the HGD gene[8] causing deficiency of Homogentisic acid oxidase, which is predominantly active in kidney liver and bowel. This enzyme is responsible for the conversion of Homogentisic acid to Maley-lacetoacetic acid in the Phenylalanine and Tyrosine metabolic pathway. This leads to the accumulation of homogentisic acid in all connective tissues and consequently elevated levels of homogentisic acid are excreted by the kidneys in urine. When this homogentisic acid in the urine is exposed to air, it gets oxidised and this produce the characteristic dark colour of the urine in ochranosis. Diagnosis can be confirmed by estimating the amount of HGA in urine using gas liquid chromatography, thin layer chromatography or enzymatic spectrometry.[9]

Ochranosis arthropathy is caused by the increased accumulation of pigments in the cartilage leading to decreased cross-linkage of collagen, which impairs the strength of the articular cartilage[1] [10] This causes fragmentation and formation of loose bodies. The cartilages fragments may adhere to the synovial membrane and cause fibrosis. Osteophytes and subchondral cyst may cause clinical signs including pain and limited range of motion.[11]

Patients with alkaptonuria are usually asymptomatic and symptomatic ochranosis appears after the fourth decade, clinically resembling osteoarthritis.[4] A plausible explanation for that is the renal tubular excretion of HGA is very effective in the early years but becomes less so with age.[2] In most cases pre operative diagnosis is usually difficult due to low prevalence and high suspicion is required .The previously published literature, often diagnosed the patient intra operatively and was not suspected until exploration of joint found dark discolouration.[4] [12] Similarly, the diagnosis was made intra-operatively in our case, and other physical findings were noted retrospectively in the post operative period. In our case, ochranosis of the joints, visual confirmation of the blackening of meniscus in right knee was incidentally identified during TKR.

A B

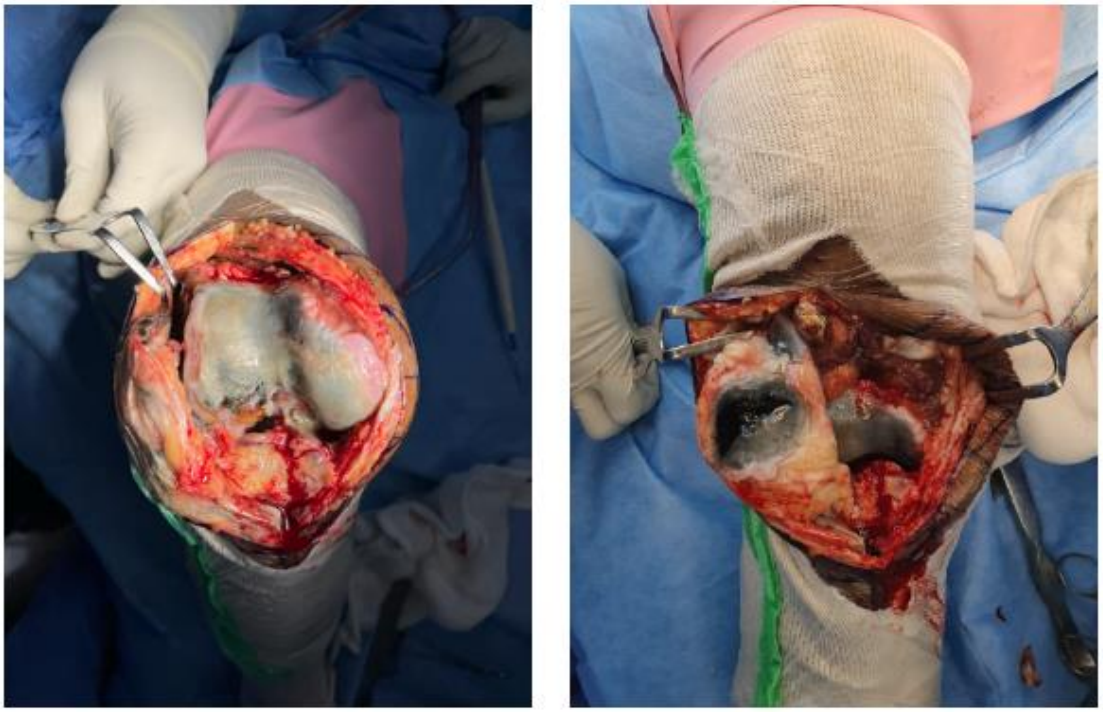

Fig A & B: Perioperative dark deposition of pigment in the knee cartilage

No definitive treatment of alkaptonuria is present currently. Symptomatic treatment of the complications of alkaptonuria is the only option. Diets low in phenylalanine and tyrosine may decrease the toxic by product HGA. Vitamin C may prevent oxidation and free radical formation. But the role of these diets in prevention of joint degeneration is not proved. Nitisinone , inhibitor of 4-hydroxyphenyl pyruvate dioxegenase, causes direct pharmacological reduction of HGA by inhibiting the tyrosine degradation pathway. Degenerative joint disease is first treated with analgesics and physiotherapy, but in case of end stage arthropathy, joint replacement is the only option.

TKA should be considered for end stage ochranotic arthropathy. Furthermore, cemented TKA offers advantages such as easier technique ensuring greater primary stability and delivery of local antibiotics when indicated.[13] A systematic review of previous publications, suggested that no particular implant design proved to be superior.[12] hence, we have performed a Cemented Cruciate sacrificing TKA.

Higher blood loss was reported in arthroplasty surgeries of patients attributed to the thickened synovium and hence the extensive synovectomy.[14] But in our case, tranexamic acid was administered pre operatively and tourniquet was used to prevent bleeding.

No specific postoperative rehabilitation guidelines are stated in literature, hence we used protocol similar to that used for standard total knee replacement in our institute. Total knee replacement has a good outcome in patients not responding to conservative management and in our case, post operative follow ups showed pain free joints and satisfactory range of motion.

IV. CONCLUSION

Ochranosis is a rare metabolic disorder affecting the cartilages characterised by dark pigment deposition. It is usually asymptomatic and diagnosis is delayed due to mild symptoms. Ochronosis arthropathy should be considered as a diagnosis when dark discoloration of connective tissue is seen intra-operatively. It may be treated satisfactorily with TKA and provides good outcome in terms of pain reduction and function for end stage arthropathy with prognosis similar to those without ochranosis.

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