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Case Report

OCHRONOSIS OF THE KNEE TREATED WITH TOTAL KNEE ARTHROPLASTY: A CASE REPORT

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ABSTRACT
Ochronosis is clinical manifestation of alkaptonuria characterized by brownish black pigment deposition in skin, cartilage and other soft tissues. Alkaptonuria is a rare metabolic disorder, with defect in metabolism of amino acids tyrosine and phenyl alanine. It can cause severe cartilage destruction in large joints and the vertebral column. Knee joint involvement, which occurs at relatively early ages, can be quite restrictive. Arthroplasty may be the treatment of choice in these patients because of limited mobility and diffuse involvement of the joint. We report a 52-year-old lady who had Ochronosis of the knee treated with Total Knee Arthroplsty.

Keywords: *Ochronosis, Alkaptonuria, Arthroplasty*

INTRODUCTION
Alkaptonuria affects one in 1,00,000 to 2, 50,000 live birth (Zatková *et al.,* 2000; Kumar and Rajasekaran, 2003) due to mutation of Homogentisate 1 2 (HGO) dioxygenase on Chromosome 3q. During the process of aminoacid metabolism intermediate producthomogentisate acid (HGA) generated is converted to methyl acetoacetic acid which is degraded and excreted in urine. Homogentisate 1, 2 dicoxygenate enzyme is responsible for this conversation is defective in ochronosis. This leads to accumulation of HGA in tissues. HGA polymerises into a brownish black pigment which gets deposited and in cartilage, bone, synovial tissue, tendons, heart valves, kidneys and skin.
The manifestation of ochronosis is secondary to pigment deposition and its implication. HGA is also excreted in urine and when exposed to air causes dark colored urine, which could be the only manifestation in childhood. Symptoms are seen only by 4^{th} to 5^{th} decade (Gaines, 1989). Pigment deposition occurs in articular cartilage leading to brittle cartilage eventually fragmentation and degenerative arthritis (Kumar and Rajasekaran, 2003). The intervertebral disc affection includes pigmentation, ossification and degenerative changes (Albers *et al.,* 1992). The deposition in tendon and ligaments leads to inflammation, calcification and rupture (Kumar and Rajasekaran, 2003; Mannoni *et al.,* 2004). Skin, cartilage and connective tissue deposition leads to pigmentation of ear lobes, sclera. Excess HGA in renal secretion causes kidney stone. Valvular calcification manifests as murmurs and failure. Knee is the commonest joint affected, however hip, shoulder joint and vertebra are also affected (Acar *et al.,* 2013). Pigment deposition also occurs in smaller joints however clinical manfestion of arthritis is rare. The clinical feature is similar to Rheumatoid arthritis and ankylosing spondylitis (Acar *et al.,* 2013). Ochroniticarthropathy primarily presents as degenerative arthropathy and is a slow progressive disease.
NASIDS, Physiotherapy and exercises are proven to be beneficial. Vitamin C 1 g/day is found to retard conversion of HGA to polymers. The benefit of Nitisinone, inhibitor of enzyme HGaoxygenase is doubtful. At present only symptomatic treatment of disease manfestation is the treatment of choice. Oschronoticarthropathy presents as OA, ankylosis in late stages. Arthroplasty of knee and hip in the treatment of choice to decrease pain and restore function (Kerimogulu *et al.,* 2005).

CASES
A 52 years old lady came with history of pain in both knee joins more in left since 5 years. Difficulty in climbing stairs and squatting. On Examination she was found to have arthritic changes with Crepitus and painful range of motion –> 0 – 90^0only. All conservative measures given previously had failed. On radiology found degenerative Tricompart mental changes [figure 1].

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General Physical examination did not reveal any significant changes and physically fit for surgery. She was advised TKR .Knee was approached through standard medial parapatellararthrotomy [Figure 2]. On opening the knee we found dark pigmentation of all the particular surfaces. Extensive loss of particular cartilage and all compartments was affected. The menisci were brittle with pigmentation [Figure 3-4]. She underwent standard posterior stabilized cemented total knee replacement (Johnson and Johnson – Depuysynthes) [Figures 6-7].

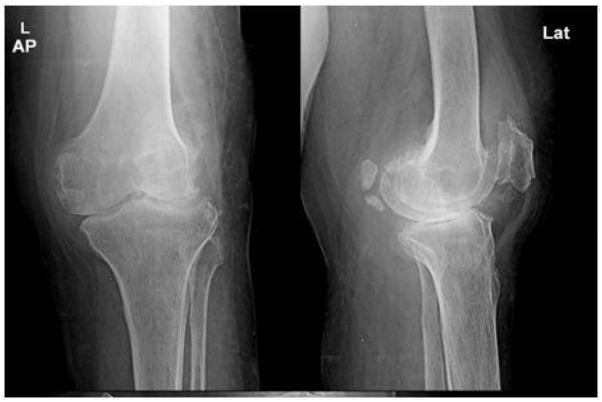


Figure 1: X- ray of knee joint showing Tricompartmental changes

Figure 2: Showing medial parapatellar arthrotomy approach

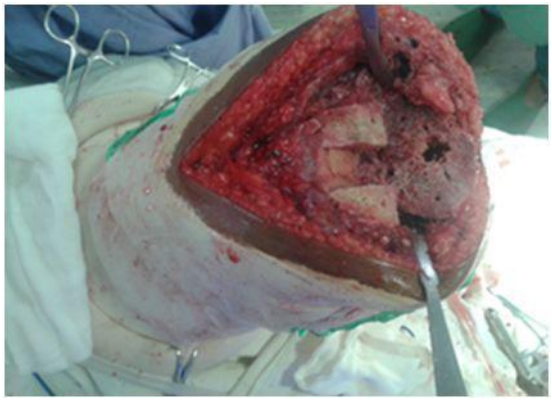
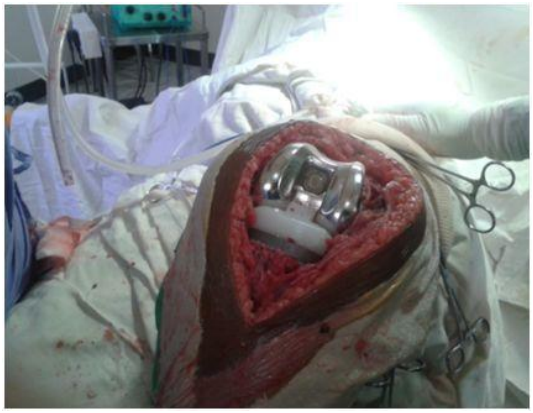
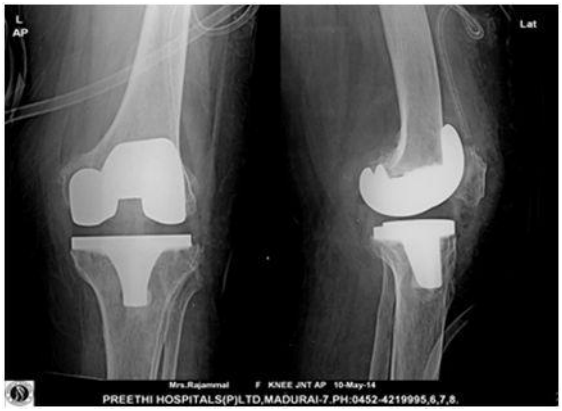


Figure 3-4: Showing dark pigmentation of all the articular surfaces and extensive loss of articular cartilage in all compartments





Figures 6-7: Standard posterior stabilized cemented total knee replacement

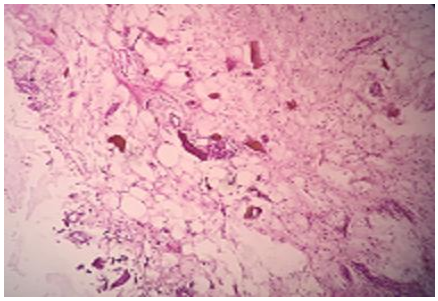


Figure 8: Histopathology of synovium showing bony trabecular hematopoietic tissue exhibiting yellow brown chronotic pigment deposits surrounded by plenty of inflammatory cells consistent with ochronosis

The cut surface and synovium was sent for histopathology Read as - Bony trabecular hematopoietic tissue exhibiting yellow brown chronotic pigment deposits surrounded by plenty of inflammatory cells consistent with ochronosis.
The post operative period was uneventful. Patient regained full range of motion from full extension to 115^0 of flexion by 6 weeks. At 1 ½ year follow-up patient did not have any complaints of knee pain with oxford score of 40 / 48 (Pre op 19/48).

DISCUSSION

Ochonoticarthropathy is clinical manifestation of Alkaptonuria. Alkaptonuria is asymptomatic and manifests in 4^{th} 5^{th} decade. Most of the cases are diagnosed intra operatively. First sign of alkaptonuria is discolouration of urine which was not seen in our patients. Discoloration of sclera and earlobe was also not seen. Being illiterate she cannot recollect any family member affected with this disorder.
Weight bearing joints like knee and hip are commonly affected on ochronioticarthropathy. Small joints may also have pigment deposition. On radiology osteophytic changes and all compartment affection may be the only feature like in our case.
At present there is not definitive treatment for ochronosis. Only symptomatic treatment of affected joints. In the review of literature most of the cases had bilateral TKR with satisfactory results. Our patient pain and oxford score improved much better compared preoperatively. We feel Arthroplasty is the treatment of choice and most of them are diagnosed intra operatively. As the cases are rare, pooled collective data of all the cases in journals are necessary to seek clear knowledge and outcome.

Conclusion

Ochronosis is characterized by severe changes and a rapid progression. It may have devastating end results, as in described our case. Since there has been no successful causal treatment to date, finally, arthroplasty may provide a solution, as of our cases have demonstrated.

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