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A study on total knee replacement in alkaptonuric patient

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Abstract
Introduction: Ochronosis arthropathy (OcA) is a rare condition which may be treated with total knee arthroplasty (TKA) at the end stage. The condition is often discovered only intraoperatively and the ideal choice of TKA is unknown.
Methodology: In the present study we studied 5 patients with worsening chronic bilateral mechanical knee pain had failed conservative therapy. Posterior stabilised (PS), cemented TKA and patella resurfacing was performed. Intraoperatively, collagenous structures such as the menisci and cartilage were noted to be black. Histological examination showed deposition of large amorphous brown material suggestive of ochronosis. He recovered well and underwent TKA of the contralateral knee the following year. At 2 years post index TKA, his outcome scores improved and he was satisfied.
Discussion and Conclusion: With increasing TKA performed worldwide, a surgeon may eventually be surprised by the above findings once in their lifetime. However, as the pathogenesis of OcA appears to be inflammatory in nature, we suggest using cemented PS TKA with resurfacing of the patella.

Keywords: TKA, ochronosis, collagenous structure, cartilage

Introduction
Alkaptonuria is an autosomal recessive disorder caused by the deficiency of homogentisate 1, 2 dioxygenase activity. It was first described by Virchow in 1866. Alkaptonuria is disorder of tyrosine metabolism due to deficiency of homogentisic oxidase characterized by excretion of homogentisic acid in urine, deposition of oxidized homoitate pigments in connective tissues, the dermis, apocrine glands and articular cartilages (ochronosis). The pathogenesis of the disease is the polymerization of deposited HGA that discolors and weakens the connective tissue, ultimately resulting in brittle tissue that is easily disrupted and leads to chronic inflammation, degeneration, and eventually osteoarthritis [1]. Patients with alkaptonuria are usually asymptomatic and arthropathy appears after the fourth decade [2]. Systemically, there is thickening and blueeblack or grayeblue discoloration of ear cartilage. Other body locations include the eyelids, sclera, foreheads, cheeks, axillae, genitals, nail beds, buccal mucosa, larynx, tympanic eardrum and nasal tip. Simultaneously, this discoloration can occur in skin, tendons, ligaments, costochondral junctions, sclera, heart valves, the intima of blood vessels and cause lumbar intervertebral disc calcification and disc space narrowing [3-5]. The knee is the most common site of orthopedic abnormality. Other sites of involvement are hips, shoulders, sacroiliac joints and the pubic symphysis [6]. There is currently no definitive cure for alkaptonuric ochronosis. However, total joint replacement in published cases of ochronotic osteoarthritis report good results similar to osteoarthritic patients without ochronosis [7]. We report the 5 patients with a family history of ochronosis, who developed degenerative arthritis of the knee.

Materials and Methods
- Sample size: 5 patients (60 years/ Male)
- CHIEF COMPLAINTS

1. Bilateral Knee pain
2. Low back ache
3. Inability to walk - 1 Month

Apparently asymptomatic before 2 years. Developed pain in both knees and low back ache which was gradual in onset and progressive. Pain aggravated on walking and was relieved on taking rest & medication

The patient's father who died at the age of eighty, had in later life developed a marked stoop and had severe arthritis of many joints.

General Examination

- Patient is conscious, coherent, cooperative moderately built and nourished.
- Brownish pigmentation of sclera of both eye's, the cartilage of external ears.
- The pigment deposits were not tender.
- Loss of lumbar lordosis
- No pallor/ icterus/ cyanosis/ clubbing/ lymphadenopathy/ pedal edema
- The patient had stable vitals

Local Examination

Inspection

Inverted dumbell shaped swelling seen in the suprapatellar region bilaterally (Left > Right).

Palpation

- Tenderness is present.
- No local rise of temperature.
- Swelling appreciated in the suprapatellar fossa.

Movements

- Fixed flexion deformity: 20° Left knee
  20° Right knee
- Range of motion
- 20°-90°(active) up to 100° passive

Systematic Examination

- CVS: S1, S2 heard. No audible murmurs.
- CNS: No abnormality detected
- Respiratory: Normal Vesicular Breath Sounds
- P/A: Soft, Non tender, No organomegaly.

Lab Investigations

- Homogentisic acid levels were 150.30 while the reference range is 1.00%

Treatment

We have performed bilateral total knee replacement.

Quadricep's snip technique was used to expose the knee joint and prevent any tendon ruptures. No intraoperative complications were encountered.

Post-operative treatment

- Patient received 5 units of packed RBC's
- Made to walk with walker support the very next day of surgery.
- Received standard physical therapy with excellent results.

Discussion

Patients presented with joint involvement have joint replacement as the final treatment option as replacement has a very good outcome as far as functional outcome is concerned [11]. Patients with alkaptonuria are usually asymptomatic and arthropathy appears after the fourth decade [2]. Ochronotic arthropathy is often diagnosed at surgery and may not be suspected until a blackened joint is found at surgery [8, 9]. Early diagnosis of ochronosis is valuable to orthopedic surgeons to avoid tendon ruptures and preoperative cardiac clearance is also important because of the risk of valve calcification [11]. In the present study the diagnosis of ochronosis was not made until exploration of the joint and confirmed after taking complete family history retrospectively after surgery. So high suspicion is required for preoperative diagnosis. Ochronosis mainly affects the cartilage and does not affect the subchondral bone. In this case, the patellar tendon had mild black discoloration and while retracting the patella a popping sound was noted which can be a sign of tearing or detachment; fortunately I did not find such. Tendon can be friable and stiff due to ochronotic involvement and one should be careful while retracting patella. We need more such case presentations to prove real incidence of such involvement. Subchondral bone was normal in appearance and free of pigments but while taking cuts it was observed that it was of soft consistency (extra soft as compared to softness of osteoporotic bone as per tactile sensations while taking cuts). The reason behind altered consistency could not be found and it is not mentioned in prior case reports. As there is also lack of data suggesting choice between cemented or non-cemented prosthesis, we chose to drill holes in cut surfaces of the femur and tibia before cementing as a part or our routine procedure. Cement was used because of soft bone. Increased intraoperative and postoperative blood loss was found in earlier case reports. This may be due to synovectomy of the hypertrophied synovium which was more than what we usually observe in other common causes of arthritis. Our protocol is to remove drain on second postoperative day. But due to increased intra and early postoperative blood loss, drain was kept up to forth postoperative day. One could consider tranexamic acid to decrease blood loss.

Fig 1: X-ray of normal man

Fig 2: X-ray of osteoarthritic Knee

Fig 3: Intraoperative procedure

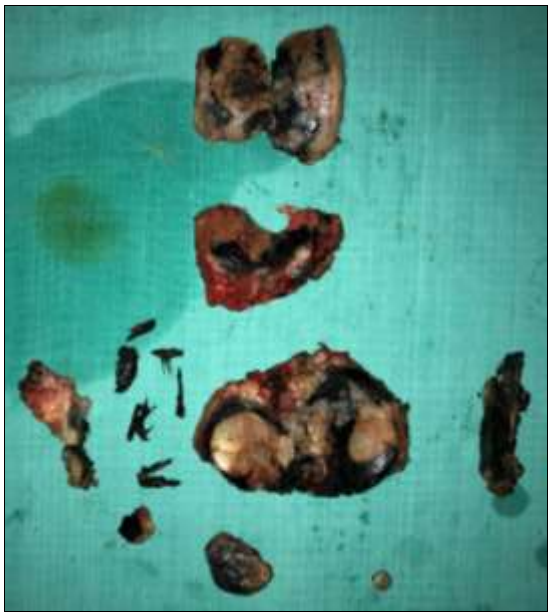

Fig 4: Black stiff menisci, articular cartilage and cut surfaces of the tibia and femur

Fig 5: Intraoperative osteoarthritis knee

Conclusion

- Alkaptonuric patients may be treated supportively. nitisinone is the drug of choice
- Total knee replacement has many complications such as deep vein thrombosis, disseminated intravascular coagulation.
- Joint replacement has excellent outcomes in a patient with significant degenerative arthropathy.

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Conflict of Interest

None

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