

Taylor's Approach to Rescue: A Case of Undiagnosed Alkaptonuria Undergoing Bilateral Total Knee Replacement

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Alkaptonuria is a rare inherited autosomal recessive metabolic disorder due to the deficiency of homogentisic acid oxidase resulting in accumulation of homogentisic acid in collagen and cartilaginous tissue resulting in stiffening of joints, spine and involving other systems which pose challenges to anaesthesiologist in providing anaesthesia. Here we present a patient with undiagnosed alkaptonuria who was posted for bilateral total knee replacement. Since these patients usually have reduced intervertebral spaces, neuraxial anaesthesia was difficult with classic approach. Taylor's approach was used for neuraxial anaesthesia and, post-operative analgesia was achieved with continuous femoral nerve block with catheter placement.

Key words: alkaptonuria, Taylor's approach, total knee replacement, regional anaesthesia, femoral nerve block

Introduction

Alkaptonuria is a rare autosomal recessive inherited metabolic disorder where the enzyme homogentisic acid oxidase is deficient which is needed for tyrosine metabolism. It is characterized by the triad of homogentisic aciduria, ochronosis, and arthritis.^{1,2} Its reported incidence is about 1 in 2,50,000 to 1 in 10,00,000.³ The clinical manifestations are due to the accumulation of an ochre coloured homogentisic acid in the cartilages and connective tissues.²

We present a case of alkaptonuria posted for total knee replacement for whom classical neuraxial technique failed, so Taylor's approach was used as a rescue technique and post-operative analgesia was achieved by continuous femoral nerve block.

Case report

A 58-year-old male patient with a BMI of 21.3kg/m², and bilateral knee joint osteoarthritis came for bilateral total knee replacement. He presented with complaints of bilateral knee joint pain for the past 6 months, hindering his daily activities. He also had low back pain and joint stiffness. He had no other comorbidities and had a good effort tolerance. On examination, vitals were stable. Airway assessment revealed Mallampati class 4 and restricted neck extension with adequate mouth opening. Examination of lumbar spine showed narrowed intervertebral spaces on palpation. Blood investigations were within normal limits. Chest X-Ray, ECG and echocardiogram were unremarkable. Initially the patient was posted for left knee arthroplasty. We planned to proceed under combined spinal epidural anesthesia after obtaining informed consent. After positioning the patient seated, 18G Touhy epidural needle was attempted at the L3-L4 space under strict aseptic precautions. Multiple attempts in both midline and paramedian approaches at different levels were unsuccessful. Hence the procedure was abandoned. Then we attempted to give a subarachnoid block at L3-L4 space both paramedian and midline approaches using a 25 G Quincke needle but failed. So as a final attempt

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Taylor's approach was considered and was successful. After free flow of CSF, 2.8 ml of 0.5% heavy bupivacaine and 0.2 ml of 30 mcg clonidine were given. Block was adequate and achieved up to T6 level. Intra-operatively, the orthopedic surgeon alerted us regarding the dark pigmentation of the knee joint cartilage. Ochronosis was then implicated to be the cause for arthritis and the difficulty in providing neuraxial anaesthesia. Throughout the procedure patient was hemodynamically stable. Surgery lasted for 2 hours with a blood loss of 600 ml. Postoperatively pain was managed with a continuous femoral nerve block infusion with 5ml/hr of 0.25% levobupivacaine. Femoral catheter was left in place for 48 hours post operatively and patient was pain free with a visual analogue score of less than 3 throughout the 48 hour period. No rescue analgesic was needed, and patient was able to mobilize.

Post operatively the patient was further evaluated for suspected alkaptonuria. On systemic examination, X-ray lumbar spine showed degenerative changes with disc space narrowing and calcification indicating Baffle's sign of spine.

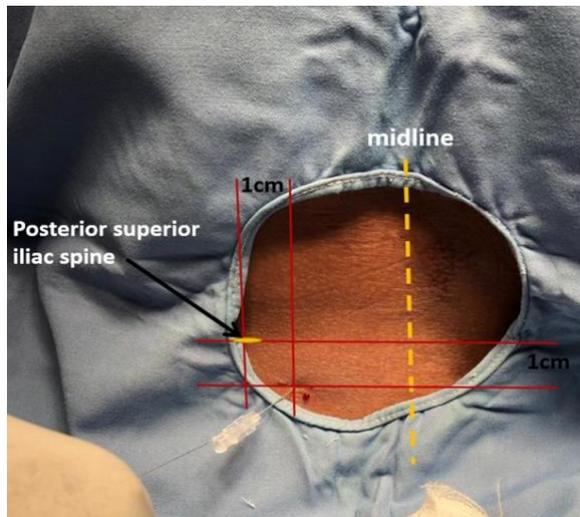


Figure 1. Taylor's approach

Urine was tested which turned black after few hours on exposure to air. Histopathological evaluation showed characteristic extensive brownish black discoloration involving the cartilage and fibro collagen confirming the

diagnosis of endogenous ochronosis (Alkaptonuria).



Figure 2. Intra-operative, blackish blue discoloration of knee joint



Figure 3. Baffle's spine

After 4 days, patient was again posted for right knee total replacement which was successfully managed with planned Taylor's approach and continuous femoral nerve block. Patient was discharged after 10 days without any complications.

Discussion

Ochronosis refers to systemic deposition of ochre coloured pigment in cartilages and collagenous tissues. There are two different types of

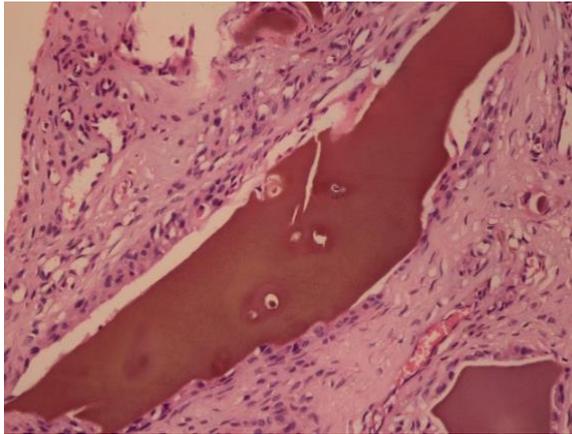


Figure 4. HPE showing brownish black discoloration



Figure 5. Sonographic image showing femoral nerve anatomy

Table 1: Showing systemic manifestations of alkaptonuria and anaesthetic implications		
Systems	Manifestation	Anaesthetic implication
Skeletal system	Pain, cartilage destruction in shoulder, hip and knee joint results in joint space loss. Mild osteophytes and tendon abnormalities	Careful positioning
Spine	Degenerative changes in lumbar and cervical spine, disc space narrowing and calcification, vertebral osteoporosis and osteophytosis, Baffle's spine. Restricted range of motion of cervical spine, neurological symptoms	Preoperative neurological examination Difficult regional blocks Difficult laryngoscopy and intubation
Airway	Pigment deposition in the larynx, trachea and narrowing of glottic opening	Small endotracheal tube Difficult airway cart should be readily available
Respiratory system	Restrictive pulmonary disease	Preoperative pulmonary function testing
Cardiovascular system	Valvular heart disease, accelerated atherosclerosis, coronary artery disease, conduction abnormalities	Preoperative 12 lead ECG, Echocardiography, Intensive intraoperative monitoring

Ochronosis; exogenous and endogenous. Ochronosis affects the skeletal, respiratory, cardiovascular, genitourinary, cutaneous and ocular systems.⁴ These clinical manifestations become evident only around fourth decade of life due to the pigment deposition within the affected tissues.⁵

Because of spine involvement both general anesthesia and central neuraxial anesthesia

possess challenges to the anaesthetist. Hence a variation of the paramedian approach, the lumbosacral approach described by Taylor can be used as a rescue anaesthetic technique. This technique is carried out at L5-S1 interlaminar space 1cm medial and 1 cm caudal to the lower most prominence of the posterior inferior iliac spine in a cephalo-medial direction. This is an alternative technique when standard approach at

L3-4 and L4-5 spaces fails. This is a very useful method in case of spinal fusion, arthritic joints, spinal deformity, skin infection in the lumbar region or in other conditions in which the usual approach is difficult or impossible.^{6,8}

It is the lowest and widest lumbar space with least chances of trauma to the spinal cord. This space is least affected by arthritic and degenerative changes and, hence Taylor's approach is a better alternative to midline approach.⁹ It is described in various textbooks as an alternative to standard midline approach, but the success depends on the acquisition of knowledge and skills to perform it. Frequent hands-on training and teaching this technique increase the safety and success rate in difficult cases. Chalopathy *et al*⁹ conducted a study to know the success rate of Taylor's approach in patients with deformed spines among residents and found 94% success rate with a 6% acceptable failure rate. There are many case reports in the literature on Taylor's approach for various spinal deformities but for alkaptonuria the literature is very little as the condition itself is quite rare.

In a study by Shanthanna *et al*¹⁰ showed superiority of femoral nerve block over epidural in patients undergoing total knee replacement. Peripheral nerve block has the advantage of sparing motor control in one leg to allow for early mobilization with crutches, avoid post-operative delirium and epidural associated systemic side effects.

Conclusion

Anaesthetists should have a good knowledge about various anaesthetic techniques and a flexible anaesthetic plan that best suits the patient to proceed when encountered with unexpected problems as in this case. Taylor's approach for spinal anaesthesia is indeed a rescue technique in cases of difficult spine and avoids the airway manipulation in patients with anticipated difficult intubation by avoiding general anaesthesia.

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