

## Case Report

### Black burden or Taylor the saviour: A case report

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### Abstract

Alkaptonuria, also called Black bone disease or Black urine disease is a rare metabolic disorder due to a deficiency of the enzyme Homogentisate 1,2 dioxygenase, which helps in tyrosine metabolism. Homogentisic acid gets converted to maleylacetoacetic acid with the help of this enzyme Homogentisate 1,2 dioxygenase. Hence deficiency of the enzyme leads to the accumulation of metabolite Homogentisic acid, in the tyrosine pathway. This accumulation may cause various manifestations in different parts of the body. Mostly they present with arthropathy, and most patients present with musculoskeletal issues and land up in orthopaedic surgeries. Clinical presentation of this disease should be kept in mind because arthropathy of the spine makes difficult access to subarachnoid space, thus posing challenge to anaesthesiologist. Ochronosis is the bluish-black discoloration of certain tissues, such as the ear cartilage and the ocular tissue, seen with alkaptonuria.

**Keywords:** Alkaptonuria, Homogentisic acid, Ochronosis, Bamboo spine, Taylor's approach

### Background

Arthropathy is the most common complication of Alkaptonuria. Deposits of Homogentisic acid in connective tissue and inter-vertebral joints increase stiffness of

joined with restricted movements. Although there is no specific management for this disease, symptomatic management is followed in a wide set of patients. In these group of patients, increased stiffness and reduced extension of cervical spine also leads

to difficult in laryngoscopy and endotracheal intubation. Preoperative evaluation of such patient for regional anaesthesia, say Spinal or Epidural anaesthesia should be thoroughly done. X-ray lumbo-sacral spine, CT imaging of Lumbo-sacral spine and USG of Lumbar vertebra will give a clue in predicting difficult neuraxial anaesthesia.

### Case Presentation

A 60-year-old female, known hypertensive, diagnosed to have Osteoarthritis of Right hip was posted for Right Total Hip Replacement. Preoperative evaluation of this patient was done 3 days before surgery. On evaluation, she was on anti-hypertensive drug with good blood pressure control. Her airway assessment was almost normal, except for limited neck extension. Lumbar spines were examined, and inter-vertebral spine spaces were narrowed. All the above spine findings were attributed to degeneration changes from the Anaesthesiologist's point of view. Routine examination of her eye to evaluate anaemia showed some scattered brownish pigmentation over the sclera of both the eyes which was not considered to be a significant finding, considering her age factor. At the end of our Pre Anaesthetic examination, she was counselled for Spinal anaesthesia and consent for the same was obtained.

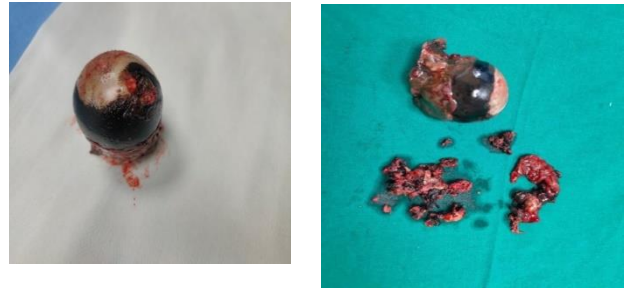
On the day of surgery, patient was shifted inside Operation theatre, appropriate monitors were connected and flow of IV fluid was ensured. She was once again explained about spinal anaesthesia briefly and procedure was started with patient in sitting position. 27 Gauge Whitacre (pencil point tip) needle was used to identify sub arachnoid space through midline inter-spinous space of L2 and L3, but we couldn't reach sub arachnoid space in spite of couple of attempts. Subsequently, inter-spinous spaces of L3-L4 and L4-L5 were also tried multiple times. Every time the needle hit the bone and we couldn't enter sub arachnoid space. Spinal needle was changed to 26 G Quincke tip needle and same spaces were tried both via midline and paramedian approach. All the approaches turned out futile, hit the bone and couldn't reach sub arachnoid space. Failure of all the above approaches, Taylors approach was performed; a variation of the



paramedian approach, described by Taylor, was carried out at the L5-S1 inter-space, the

largest inter-laminar inter-space of the vertebral column. A 26 G Quincke spinal needle was inserted in a cephalo-medial direction through a skin wheel raised 1 cm medial and 1 cm caudal to the lowermost prominence of the posterior superior iliac spine which is a useful approach in patients with ankylosing spondylosis where the spines will be fused like a bamboo. Taylor's approach of Spinal anaesthesia was performed successfully using 26G Quincke needle and 0.5% Inj. Bupivacaine Heavy 3 ml and Inj. Buprenorphine 30 mcg (as additive) was injected. After achieving adequate level

Next day in the ward when patient was questioned further regarding her past history and incidents, she revealed that she used to have dark red coloured urine during her childhood but didn't notice it further. On re-examining her eyes, we could notice symmetrical brownish pigmentation over sclera in both the eyes. X-ray Hip was taken which revealed fused spinal vertebrae resembling bamboo spine appearance with reduced joint space. All the history and clinical findings supported to diagnose Alkaptonuria in this patient. Fused spines in this condition were the main reason behind difficulty getting a spinal anaesthesia.



of block, patient was positioned, and surgery was started.

During intra operative period when joints were exposed blackish discoloration of femoral head, cartilages and joint spaces were noticed. Urine was also dark coloured. Surgery was over and patient shifted to post operative ward.



## Discussion

Alkaptonuria or Ochronosis has three main features,

(1). Darkening of the urine upon contact with air.

Homogentisic acid is oxidized to form a pigment-like polymeric material responsible for the black color of standing urine or after exposure to an alkaline agent.

(2). Ochronosis (bluish-black pigmentation of connective tissue).

Accumulation of Homogentisic acid and its oxidation products - benzoquinone acetic acid in connective tissue leads to ochronosis – brown pigmentation of the sclera.

(3). Arthritis.

It begins in the spine. Degenerative changes, mainly in intervertebral disks, may be seen throughout the vertebral column, where the lumbar spine is the most commonly affected. With the progression of the disease, it may



resemble those of ankylosing spondylitis. Patients may complain of stiffness in their lower back with no other symptoms or signs of lumbar spine disease. It may lead to disk space narrowing, widespread disk calcifications, osteophytosis with calcification of the intervertebral ligaments. Half of the individuals require at least one hip joint replacement by the age of 55 years.

Pigment deposition is also seen in heart endocardium, valves, and kidneys. Therefore, patients may have valvular disease, nephrolithiasis, and other renal complications in the advanced age.

## Conclusion

Taylor's approach could provide a reliable and less traumatic alternative to midline approach for lumbar puncture particularly in deformed spine due to scoliosis, severe arthritis, fused spines because of Ochronosis or prior spine surgery, which will pose a challenge, due to anatomical and technical difficulty, for establishing a successful subarachnoid block.

## References

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