



**Figure 1:** Vertebral column sclerosis

The patient had received general anaesthetic 4 years before for fracture of the shaft of the femur. The fracture had malunited resulting in deformity, and a redo interlocking nail femur was planned. Patient was explained the need and the procedure of fiberoptic guided awake intubation, in case of failure of regional anaesthesia. The patient was 36 kg in weight, 138 cm tall with short limbs, frontal bossing, macrocephaly with no mental retardation, receding mandible and irregular dentition. Airway assessment revealed a high arched palate, restricted neck extension and a modified Mallampati airway grade 2b. Difficult intubation was anticipated. In view of AAD, advanced airway gadgets to prevent neck movements during laryngoscopy and intubation were kept ready. Substantial sclerosis of vertebral bodies was noticed in spine radiographs with almost fused spine in the lumbar region, thereby increasing the technical difficulty in performing the neuraxial blockade.

The patient was pre-medicated with injection pantoprazole 40 mg and alprazolam 0.25 mg. The patient was verbally assured and could be comfortably positioned sitting for the neuraxial block. Needle through needle technique was used for CSE. Block was performed by paramedian approach in L2-L3 intervertebral space and was successful in the first attempt. Bupivacaine 0.5% heavy (10 mg) with fentanyl 12.5 µg was deposited intrathecally. The epidural catheter was fixed at 7 cm mark (space identified by loss of resistance to air at 2 cm from the skin). A sensory analgesia up to T10 dermatome was achieved. The patient was carefully positioned with adequate padding of pressure points. Tranexamic acid 10 mg/kg intravenous was administered during surgery. A fixed dose continuous epidural infusion containing

## Marble bone disease and the Anaesthesiologist

Sir,

Osteopetrosis is an uncommon genetic disease encountered by anaesthesiologists and is rarely reported.<sup>[1]</sup> The various associated anatomical and physiological alterations are challenging. We report the successful management of a 35-year-old female with the condition, scheduled for revision surgery for malunion of femur under combined spinal-epidural block (CSE). Pre-operatively, difficult airway, atlantoaxial dislocation (AAD) and significant bony sclerosis of entire vertebral column were identified [Figure 1].

bupivacaine 0.125% with fentanyl 1 µg/ml was started at 5 ml/h at the end of surgery for post-operative analgesia. Patient exhibited stable haemodynamics during surgery. The epidural catheter was removed on 3<sup>rd</sup> post-operative day. The patient did not exhibit any signs of post-operative neurological deterioration.

Osteopetrotic patients<sup>[2]</sup> vary greatly in their presentation and severity. The simplest form is osteopoikilosis, noticed as incidental finding of osteopetrosis on radiographs. Autosomal recessive variant has a neonatal onset with fractures, short stature, compressive neuropathies, hypocalcaemia, tetanic seizures, pancytopenia, mental retardation, skin and immune system involvement and renal tubular acidosis.

The onset of primarily skeletal manifestations such as fractures, osteoarthritis of hip joint and osteomyelitis of the mandible at adolescence is typical of autosomal dominant variant. Common findings are cranial nerve compressions manifesting as visual and hearing loss, nutritional anaemia, thrombocytopaenia, brittle and osteomyelitic bones (more prone to bleed), hypocalcaemic seizures with secondary hyperparathyroidism and muscular hypotonia. Judicious use of transfusions is warranted as significant immunosuppression is a feature of this disease. Nasal space is reduced by bony encroachments with disturbed facial anatomy complicating nasal intubation. Fibreoptic guided awake oral intubation is preferred if general anaesthesia is contemplated.<sup>[3]</sup>

The anaesthetic options are limited and technically difficult with both airway and spine involved in the disease process. The osteopetrotic bones are difficult to drill and surgical time can escalate. We were able to successfully place a central neuraxial block in our patient, but this may not always be the best anaesthetic. Hence, pre-operative identification of anaesthetic complexities is essential to plan safe perioperative course.

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Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/0019-5049.158779