

Difficult epidural in a patient with undiagnosed alkaptonuria

Sir,

Endogenous ochronosis or alkaptonuria is an extremely rare autosomal recessive inborn error of amino acid metabolism which was described by

Garrod in 1901.^[1] It occurs due to the absence of the enzyme homogentisate 1,2-dioxygenase that converts homogentisic acid to maleylacetoacetic acid in the process of tyrosine metabolism.^[2] We wish to highlight difficulties faced in securing neuraxial anaesthesia in an undiagnosed case of alkaptonuria and the significance of identifying mucocutaneous markers of ochronosis which can be missed in dark-skinned Asian patients.

A 69-year-old male, who presented with pain in both the knee and hip joints and no significant comorbidity, was posted for total replacement of the right hip joint, with a provisional diagnosis of osteoarthritis. General physical examination showed bilateral pigmented keratinised lesions on the palmar aspects and outer edges of his fingers [Figure 1a]. His vital parameters were stable and systemic examination was unremarkable. Airway examination revealed a mild restriction of neck extension and Malampatti class 3. The lumbar interspinous spaces were not distinctly felt. The electrocardiogram showed an incomplete right bundle branch block and the echocardiogram showed a degenerated sclerotic aortic valve, trivial regurgitations in the mitral and tricuspid valves and a normal left ventricular ejection fraction. Combined spinal-epidural anaesthesia was planned.

Lumbar epidural needle insertion was attempted long the midline but failed as bone was encountered with repeated attempts. The patient was then positioned on the right lateral side with the affected joint dependent and subarachnoid block through a paramedian approach was successful. The epidural space was reached after multiple attempts by the paramedian approach near L2-3 and an 18 gauge epidural catheter was successfully threaded into space. Intra-operatively, the surgeon alerted us regarding the dark colour of the cartilage around the hip joint. Ochronosis was then implicated to be the cause of arthritis and the difficulty in securing neuraxial anaesthesia. Close evaluation of the eyes revealed pigmented lesions bilaterally [Figure 1b] and the ears were found to be hard and



Figure 1: (a) Greyish-blue, pitted, scaly and non-itchy lesions on the palms. (b) Greyish scleral pigmentation

thickened. The lumbar spine radiograph obtained subsequently showed calcification of the intervertebral discs [Figure 2a]. The urine was tested which turned dark after a few hours on exposure to air and further on alkalinisation [Figure 2b]. Histopathological evaluation of the femoral head confirmed the diagnosis [Figure 2c]. The patient later recollected that his unwashed underwear showed ash-coloured stains.

Since the disease often presents in the later decades of life with symptoms closely resembling other arthropathies, the diagnosis is often missed. Anaesthesiologists encounter these patients when they are posted for replacements of the hip or knee joints and for spine surgeries. They can present with ochronotic spondyloarthropathy with decreased range of movements of the spine and hips, facet joint degeneration, spinal canal stenosis and pseudo ankylosis of the cervical joints, reduced intervertebral disc spaces and dense disc calcification that can result in difficult neuraxial anaesthesia as well as intubation.^[3,4]

Bilaterally symmetrical scleral pigmented lesions, as seen in our patient, should rouse suspicion of an underlying systemic cause.^[5] Greyish-blue, pitted, non-itchy, scaly lesions on the edges and palmar aspects of the hands, greyish discolouration of



Figure 2: (a) Intervertebral disc calcification (yellow arrows). (b) Change in the urine colour on alkalinisation and exposure to air. (c) Microscopic pigment deposition in the cartilage and in the synovium

the ear pinna and stiff and hard helical cartilage were noted in our patient can easily be missed.^[6] Calcifications in the coronaries, cardiac valves and aortic root, urinary pigment calculi and nephrotoxicity may be significant enough to alter anaesthetic management.^[2]

Although surgical implications are minimal, surgeons should involve the anaesthesiologist, physician and cardiologist early in the pre-operative period to establish a well-planned management approach.

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Conflicts of interest

There are no conflicts of interest.

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