

Inadequate Spinal Anaesthesia in a Case of Marfan Syndrome with Post Dural Puncture Headache Posted for Epidural Blood Patch

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Respected Sir,

Marfan syndrome is an autosomal dominant connective tissue disorder with a reported incidence of one in 3,000-5,000 individuals which is caused by mutation in the fibrillin-1 gene located on chromosome 15^{1,2}. It is characterized by disproportionate tall stature, aortic root aneurysms, vertebral anomalies, dural ectasia and ectopia lentis³. We present a case of failure of spinal anaesthesia, followed by post Dural Puncture Headache (PDPH) in a patient with Marfan's syndrome.

A 17 year old male patient presented to our hospital with right sided uncomplicated indirect inguinal hernia posted for elective right Lichtenstein tension free mesh repair. Pre-operative evaluation revealed no comorbidities. His height was 180 cm and weight was 52 kg. Systemic examination was unremarkable except for high arched palate. Laboratory investigations were within normal limits. Two-dimensional echocardiography showed structurally normal cardia with trivial TR (jet gradient 31 mm Hg), normal pulmonary artery pressures and ejection fraction 60%.

On the day of surgery, after obtaining informed written consent, patient was shifted to operation theatre. Spinal anaesthesia was administered after two attempts of lumbar puncture. Patient experienced mild tingling sensation of the lower limbs with inadequate level of analgesia. Hence, conventional general anaesthesia was administered to the patient. Post extubation, patient was shifted to post anaesthesia care unit.

The patient presented with PDPH after 48 hours of surgery. He was treated conservatively with adequate hydration, analgesics and caffeine. Despite two days of treatment, PDPH persisted. Hence, he was posted for placement of Epidural blood patch. Patient was reviewed. Procedural risks and benefits were explained to the patient and his attenders. Informed written consent was obtained. A 20-gauge intravenous canula was secured into right upper limb and fluids were started. After adequate hydration, patient was shifted to operation theatre. Baseline heart rate (72 bpm), blood pressure (136/80 mm Hg) and oxygen saturation (98%) were recorded. Patient was made to lie in the right lateral position.

Under sterile precautions, after local anaesthetic infiltration, 18 gauge Tuohy needle was used to identify the epidural space at L3-L4 intervertebral space (same space used in previous spinal anaesthesia) by loss of resistance technique. Simultaneously, under aseptic precautions, 20 ml of intravenous blood was withdrawn into a syringe. 16 ml of this blood was injected slowly into the epidural space with 2 ml incremental doses over a period of 5 minutes (Figure 1).

Resolution of headache and absence of appearance of any new symptoms were ensured by conversing with the patient during the procedure. The patient tolerated the procedure well. Once the procedure was performed, patient was made supine. Significant reduction in symptoms was noticed in two minutes post procedure. Immediate relief of symptoms by the blood patch is due to tamponade effect of the dural leakage while simultaneously raising

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Figure 1. Placement of epidural blood patch - Technique of epidural insertion and intravenous blood collection simultaneously.

the subarachnoid pressure^{4,5}. There was no sensory or motor neurological deficits and both limbs had a power of 5/5. Patient was comfortable and hemodynamically stable. The patient was monitored for 45 minutes in the post anaesthesia care unit and shifted to ward.

Dural ectasia is the widening of the dural sac often seen in patients with Marfan syndrome (63-92%)⁶. Associated increase in CSF volume due to dural ectasia and the erratic spread of the intrathecal local anaesthetic increases the rate of spinal anaesthesia failure. The association between connective tissue disorder and PDPH is explained from the disorganization of dural elastic fibres. In patients with

connective tissue disorder, the dura remains lax allowing persistent CSF leakage though the dural defect increasing the incidence of both, occurrence of PDPH and placement of Epidural Blood Patch for headache resolution⁷.

Our observation in this case concludes that increased incidence of inadequate spinal anaesthesia, development of PDPH and requirement of Epidural Blood Patch in patients with connective tissue disorders needs to be anticipated while administration of anaesthesia so as to manage them efficiently.

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