Legg-Calve-Perthes disease: A must know entity for anaesthesiologists

Sir,

Legg-Calve-Perthes disease (LCPD) is osteonecrosis of the femoral head epiphysis. It is 4 times more common among boys and bilateral involvement occurs in 8–24% of cases. The reported annual incidence varies between 0.45 and 21 cases/100,000 children. The disease is usually diagnosed among children <15 years of age, with a peak onset between 5 and 8 years of age.[1] There is delayed skeletal maturation and impaired growth. In addition to congenital abnormalities, LCPD is associated with greater risk of cardiovascular diseases and diseases of the blood.[1,2] Therefore, the disease may warrant specific anaesthetic considerations during surgery.

A 120 cm tall, 40 years female having LCPD was scheduled for total hip replacement. She had impaired growth with proportionate dwarfism. She presented with pain in bilateral hips since 7 years and was diagnosed as a case of LCPD at that time by the orthopaedician on the basis of avascular necrosis of femoral heads. She was hypertensive since 4 years and was on tablet amlodipine 5 mg twice a day. She had single port-wine haemangioma of face since birth. Systemic examination and vital parameters were normal. Airway evaluation revealed limited extension of neck with Mallampati gradeIII. Blood biochemistry including platelets was within normal limits. Chest X-ray, electrocardiography and echocardiography were normal. Magnetic resonance imaging (MRI) of lumbosacral region revealed disc desiccation at L1–L2 to L5–S1 level with disc bulge at L4–L5 and L5–S1 levels compressing the anterior thecal sac and indenting the cauda equina roots. In view of spine problems, regional anaesthesia was not considered and general anaesthesia was planned. Awake fibreoptic intubation was planned, but she could not be convinced. Hence, direct laryngoscopy under general anaesthesia was planned after checking adequacy of ventilation. Induction was done with glycopyrrolate 0.2 mg, fentanyl 100 μg and propofol 100 mg administered intravenously. After checking adequacy of ventilation, succinylcholine 75 mg was administered intravenously. Direct laryngoscopy revealed Cormack-Lehane grade II and trachea was intubated successfully. Anaesthesia was maintained with sevoflurane in 67% nitrous oxide and atracurium 20 mg initially and thereafter 5 mg repeated twice. Fentanyl 50 μg was repeated intraoperatively. Surgery lasted for 2 h. At the end of surgery, neuromuscular blockade was reversed and trachea was extubated with the patient awake. Postoperatively, analgesia was provided with paracetamol infusion 100 ml at the end of surgery and repeated 8 hourly thereafter for 2 days. Postoperative course remained uneventful. X-ray pelvis after surgery revealed right femoral implant and avascular necrosis of the left femoral head [Figure 1].

Patients with LCPD present a great challenge to anaesthesiologist. These patients may have several congenital abnormalities such as congenital heart disease, Goldenhar syndrome, haemophilia, renal disease, Down syndrome, epilepsy and scoliosis.[2] Hence, these patients should be evaluated thoroughly by history, clinical examination and investigations.

These patients have a 70% higher risk of cardiovascular diseases. Of all cardiovascular diseases, hypertension and ischaemic heart disease have been more common. These patients have a 40% higher risk of diseases of blood including nutritional, haemolytic and

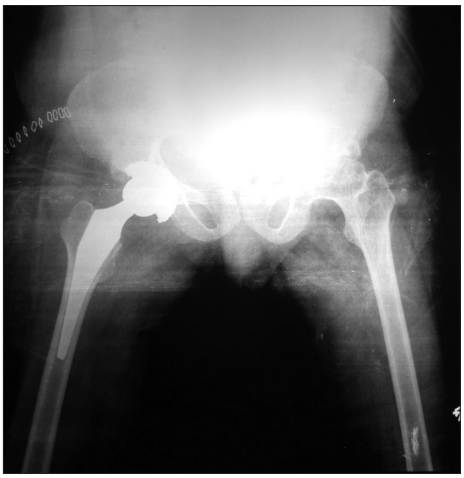

Figure 1: Right femoral implant and necrosis of the left femoral head

aplastic anaemia, purpura and other haemorrhagic conditions.[1] These patients also have increased risk of coagulation defects such as thrombophilia, activated protein C resistance, protein S deficiency and antiphospholipid antibody syndrome.[3,4] These are hypercoagulable states and patients may benefit from anticoagulant thromboprophylaxis. Our patient had no other comorbidity except hypertension that was under control with amlodipine 5 mg twice a day.

Not much literature is available regarding the choice of anaesthetic technique in patients with LCPD. In general, regional anaesthesia is chosen for total hip replacement. However, in our patient, MRI of lumbosacral region revealed disc desiccation at L1–L2 to L5–S1 level with disc bulge at L3–L4, L4–L5 and L5–S1 levels compressing the anterior thecal sac and indenting the cauda equina roots. Hence, we avoided spinal anaesthesia and chose general anaesthesia.

Spina bifida can accompany LCPD. Spinal deformities, if present, area relative contraindication to regional anaesthesia as there is difficulty in determining the cause of new neurological deficits that may appear perioperatively. The patient should be informed of the risk of neurological complications including coincidental progression of preoperative deficits. Preoperative neurological status should be documented. Furthermore, patients with preoperative neurological deficits may undergo further nerve damage more readily from needle or catheter placement, local anaesthetic systemic toxicity and vasopressor-induced neural ischaemia.[5]

There is paucity of literature regarding anaesthetic management of LCPD. However, this disease deserves special anaesthetic considerations, as in addition to avascular necrosis of femoral heads, these patients may have congenital anomalies including other skeletal abnormalities and greater risk of cardiovascular diseases and diseases of blood.

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Conflicts of interest

There are no conflicts of interest.

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