Case Report

Advanced Clinical Anesthesiology & Pain Medicine

Anaesthetic Management of a Rare Case of Cantu Syndrome

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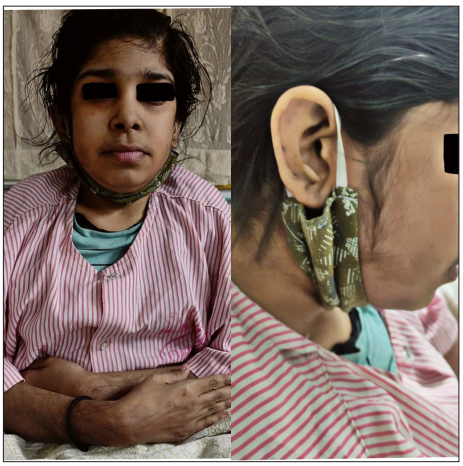
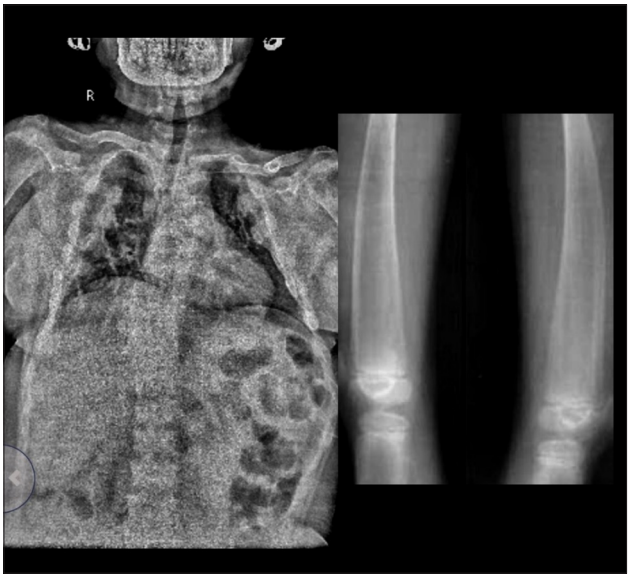
We anaesthetists encounter various cases with rare syndromes which although are posted for common surgeries, but the coexisting pathophysiological abnormalities is a concern while administering anaesthesia.

Cantu syndrome is a rare condition featuring congenital hypertrichosis, coarse facial features cardiac and skeletal abnormalities (Concolino et al., 2000; Grange et al., 2006). The patient we report here is a 26-year-old female dwarf posted for implant removal and re-fixation of left sided neck of femur fracture. History of repeated falls, similar fractures and surgery for the same was elicited.

The various co-existing pathological changes and the challenges encountered during administration of anaesthesia makes this rare case reportable. The patient was diagnosed with this syndrome at our hospital and was not aware of it.
Pre-anaesthetic assessment revealed dwarfism with height of about 127cm, grossly kyphoscoliotic dorso-lumbar spine, short neck with limited movements, limited mouth opening and macroglossia (Grange et al., 2006; Faden et al., 2009). Patient had no eye-to- eye contact with anaesthetist and had an incoherent speech.





Blood investigations revealed Hypokalemia (K-3) and raised serum Alkaline Phosphatase (Sr.ALP-775 IU/L) with rest of the values in normal range. ECG showed Left Axis Deviation, T wave inversion in leads V2-V6 and ST segment sagging in V3-V6. 2D-ECHO revealed enlarged cardiac chambers and pulmonary artery hypertension with minimal pericardial effusion (Robertson et al., 1999; Lazalde et al., 2000).

A multidisciplinary approach involving physicians, cardiologists and pulmonologists was considered immediately after the element of doubt regarding diagnosis. Case was posted a semi-emergency basis and the patient was optimised with potassium correction. Risk benefit ratio was explained to the relatives and written informed high risk consent was obtained. Regional anaesthesia was planned.

After noting baseline parameters, preloading with 300 ml of crystalloid was done and epidural space was identified with 18 G Tuohy's needle followed by low dose spinal anaesthesia with Inj.Bupivacaine (H) 0.5% 1.6 cc. Epidural test dose using

3cc of Inj. Lignocaine Adrenaline 2% was given 45 minutes after the commencement of surgery. Throughout 175 minutes of surgery, patient was haemodynamically stable. The ECG followed a similar pattern as the pre-operative picture. Total blood loss was around 700 ml which was replaced with 1 pint of whole blood. Patient also received 1 colloid and 2 crystalloids. Patient withstood the procedure well.

Postoperative analgesia was maintained with Inj.Bupivacaine 0.125% infusion for the 1st 24 hours and Inj.Fentanyl 2mcg/hr infusion in normal saline over the next 24 hours (Holley & van, 1988). Epidural catheter was removed after 48 hours.

Diagnosis of this syndrome is based on clinical features. There is only one case reported by O'Brien JJ et al, where they've summarised a 7 year old child who was operated multiple times for different surgeries, all under general anaesthesia.

Though the management of a single case doesn't define the line of treatment but we report this case with the objective that this rare case report may help clinicians in diagnosis of any such cases which otherwise may get neglected as happened in our case.

Anaesthetic challenges encountered

- Difficult airway due to macroglossia, limited neck movements with short neck.
- Compromised respiratory and cardiac functions (pulmonary function tests revealed obstructive disease pattern) with pericardial effusion and dilated heart.
- Electrolyte imbalance like hypokalemia and the related cardiac risk.
- Difficulty in administering regional anaesthesia due to kyphoscoliotic spine.
- Excessive intraoperative blood loss due to previously operated long bone fracture with implant in situ which was impacted due to fall.

This case with many anticipated anaesthesia challenges though taken care of was well managed in regional anaesthesia. Stable haemodynamic in view of cardiac anomalies is the prerequisite in these cases. Low dose regional anaesthesia was planned keeping in mind respiratory compromise and the prevalent covid 19 pandemic. In case of failed block, general anaesthesia preparation was kept ready with agents like etomidate, sevoflurane and fentanyl which are known to maintain haemodynamic stability. Such rare cases need vigilant diagnosis and multidisciplinary approach to prevent a grave outcome and good counselling should never be overlooked.

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