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Case Report

Perioperative anaesthetic management of a patient with laurence moon biedl syndrome with chronic kidney disease undergoing laparoscopic cholecystectomy: a case report

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Abstract:

Lawrence Moon Biedl syndrome is a rare genetic disorder which is autosomal recessive in nature. The rarity of this disease syndrome makes its mention scarce in medical texts. f Here, in this text, we narrate a case report of 21 year-old lady with Laurence Moon Biedl syndrome (LMBS) along with obesity, chronic kidney disease (CKD), mild left ventricular systolic dysfunction who was admitted in ICU for conservative treatment for fever, shortness of breath and bilateral pedal swelling. She was thereafter scheduled for elective laparoscopic cholecystectomy. The anaesthetic problems anticipated to be associated with this syndrome are difficult airway and perioperative hemodynamic instability due to systemic abnormalities.

Key-words: Laurence Moon Biedl Syndrome, difficult airway, CKD, anaesthetic management.

Introduction:

In 1866, Laurence and Moon, two scientists, first reported cases of retinitis pigmentosa with dwarfism, nyctalopia, mental retardation and hypogonadism.[1,2,3]. Arthur Biedl, a pathologist thereafter described an additional feature of polydactyly along with this syndrome. This disease complex was thus termed as the Laurence Moon Biedl syndrome [LMBS]. The frequency of this syndrome is 1:160000[4]. The disease spectrum also involves renal anomalies[8]. This syndrome is challenging from anaesthesiologist's standpoint due to anticipated difficult airway[8]. Presence of facial and dental abnormalities creates difficulties in the mask ventilation[9]. Bifid epiglottis or anterior laryngeal web if present lead to intubation difficulty[9] in such patients.

Case History:

A 21-year-old lady with previous diagnosis of LMBS was admitted in our ICU with complaints of fever, bilateral swelling of feet, shortness of breath, generalized weakness and decreased urine output for 7 days. Other features characteristic of LMBS such as polydactyl in both legs (figure 1), mental retardation, retinitis pigmentosa, chorioretinitis, renal and cardiac dysfunctions were also present.

On admission to ICU, her blood samples detected increased urea and creatinine (2.2 mg/dl) with low haemoglobin levels. Total leucocyte count was normal with elevated ESR, CRP and normal thyroid profile. Blood culture was positive for S.Typhi-'H'&'O'. There was raised sepsis marker [Pro-calcitonin- 2.59] indicating possibility of systemic infection. ABG analysis indicated metabolic acidosis. Urine culture was also indicative of infection.

The patient was treated conservatively under our supervision. Elevated INR was managed by 4 units of A negative FFP. Low haemoglobin was managed with 2 units of A-negative packed RBC. USG showed cholelithiasis. The patient was advised for elective cholecystectomy but was postponed due to haemodynamic instability. She was put on IV antibiotics and other supportive managements. The patient however did not require any haemodialysis. She improved gradually and was discharged after being stable. Following her up at regular intervals, we decided to go for interval cholecystectomy after 3 months.

Here, we report the anaesthetic management of a patient with LMBS scheduled for an elective cholecystectomy. On pre-anaesthetic evaluation, her parents gave history of delayed developmental milestones with mental retardation. There was also positive history of obstructive sleep apnoea. On thorough airway examination, Mallam Pati score was *III*, distances for assessment of submandibular space were all lesser than the standard for rigid laryngoscopy. Neck extension was restricted, neck flexion was adequate, Upper Lip Bite test negative. Mouth opening was 2 fingers. Limited mandibular protrusion, short fatty neck, large tongue are among the other factors contributing to anticipated difficult airway (figure 2).

On ECG there were wide QRS complex, poor R-wave progression, non-specific ST changes in multiple leads. On echocardiography, there was evidence of IHD, ejection fraction-49%, grade -*II* LV diastolic dysfunction and trivial tricuspid regurgitation. Chest skiagram revealed increased cardio-thoracic ratio.

Difficult airway cart was kept ready anticipating difficult airway. Mask ventilation of this patient was however relatively unremarkable. Since the patient was able to follow commands satisfactorily, she was prepared for awake fibreoptic intubation. Due to difficulty in identification of landmarks for airway blocks, the patient was only given intratracheal block with 2ml of 4% lignocaine and was adequately nebulised with 4% lignocaine. 10% Lignocaine spray was used for blocking glossopharyngeal nerve intra-orally. Supplemental oxygen was delivered via nasal catheter. Fibreoptic scope was introduced orally and endotracheal tube was inserted under videoscopic guidance.

Intravenous cannulation was secured with 22G cannula on the left hand. Midazolam 0.04mg/kg and glycopyrrolate 4mcg/kg were given intravenously half an hour before induction. Propofol 1.5mg/kg, fentanyl 1mcg/kg and atracurium 0.25mg/kg were administered

intravenously to facilitate intubation. General anaesthesia was maintained with gaseous mixture of 1:1 O_2 and N_2O. The surgery was completed within 45 mins without any intra-operative complications. The neuromuscular blockade was adequately reversed using Train of four (TOF). The patient was extubated when she gained full consciousness and able to follow commands. During the peri-operative period the patient was given intravenous fluids judiciously while monitoring urine output. Nephrotoxic drugs were avoided during the peri-operative and postoperative course and fluid was restricted to 1ml/kg/hr.

Discussion:

This disease syndrome comprises of two disorders, Laurence-Moon and Bardet-Biedl syndrome. It is mainly diagnosed based on clinical features. Scarcity of literature on this topic makes it to be mentioned by most medical textbooks as a congenital cause of retinal disease[8]. In India, till now only about 15 cases of LMBS have been reported[5]. Most of these cases are reported from the southern part of India[6,7].

The anaesthetic problems associated with this syndrome include obesity (weight- 63kg, BMI-37.2 kg/m^2), short neck with renal abnormalities and difficult venous access [8]. There are no data in the literature to suggest preferred anaesthetic and analgesic agents [8,9]. Facial abnormalities lead to difficulties in the mask ventilation whereas abnormalities of epiglottis if present create difficulty during intubation[10]. As we were anticipating difficult airway, awake oral fibreoptic intubation was performed. Mental retardation in such cases needs special mention in pre-operative preparation. Our patient was given premeditations 30 mins prior to induction and was kept with her parents in preoperative room.

In these patients, nephrotoxic agents should be avoided[8]. Perioperative fluid consumption, urine output should be recorded and post-operative electrolytes should be measured as were done in this case. Sevoflurane was avoided because of degradation products associated renal failure. We used atracurium as muscle relaxant as its metabolism is independent of renal function. Though the patient is non-diabetic, CBG monitoring and strict glycaemic control were undertaken. During the perioperative period life there are chances of threatening arrhythmias due to altered cardiological function. Therefore, perioperative ECG monitoring should be done. However renal and cardiac abnormalities did not affect the course of anaesthesia.

We successfully delivered critical care management and general anaesthesia to this patient with such a rare condition. Although we did not encounter any complication in this case, this syndrome however carries high potential for difficult airway management and hemodynamic instability due to cardiovascular and renal systemic disorders. Patients having LMBS should therefore be prepared preoperatively with extreme caution. Anaesthesiologists should be prepared for difficulties related to airway and systemic disorders.

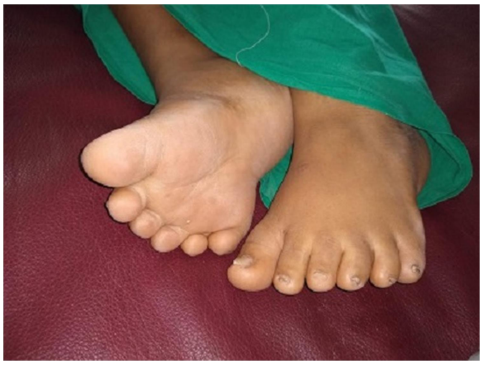

Figure 1-Polydactyl of both legs in a patient of Laurence Moon Biedl syndrome

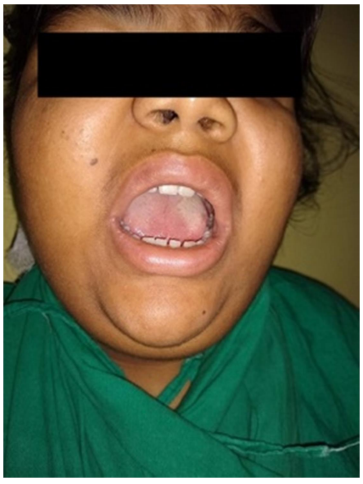

Figure 2- Patient of Laurence Moon Biedl syndrome having short, fatty neck, large tongue, Mallampati grade-*III*

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