



Anesthetic management of emergency cesarean section in a woman with lipoid proteinosis

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ABSTRACT

Lipoid proteinosis is a rare autosomal recessive disorder of generalized thickening of the skin, mucosa, and certain viscera due to deposition of numerous small clumps of proteins that give the skin a yellowish color. Lipoid deposits in upper respiratory tract, mucous membranes of the mouth and in vocal cords may interfere with airway management during general anesthesia, and hence the regional techniques are usually preferred in known cases.

Here the authors present a typical known case of lipoid proteinosis, which presented for emergency cesarean section due to abruptio placentae. She was meticulously managed with spinal anesthesia.

Key words: Lipoid proteinosis; Anesthesia, General; Anesthesia, Regional; Difficult airway; Endotracheal intubation

Citation: Nouredine K, Shibli K, EL Boursaly I, Jain SD, Lemtai Z. Anesthetic management of emergency cesarean section in a woman with lipoid proteinosis. *Anaesth Pain & Intensive Care* 2018;22(4):515-517

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Received: 4 Nov 2018

Reviewed: 5 Dec 2018

Corrected & Accepted: 31 Dec 2018

INTRODUCTION

This is a case of a full-term pregnant woman with a rare disease called lipoid proteinosis (LP), who presented for an emergency lower segment cesarean section (LSCS).

LP¹ is a rare autosomal recessive disorder of generalized thickening of the skin, mucosa, and certain viscera. The condition results from the formation of numerous small clumps of proteins that appear in the skin giving it a yellowish color, deposits in upper respiratory tract, mucous membranes of the mouth and the eyelids; but it does not impair vision.

Due to the deposits in the vocal cords, the first noticeable symptom is usually a hoarse voice. In infants a weak cry is a sign of this disorder. The voice abnormalities persist throughout the person's life and may ultimately cause difficulty in speaking or eventually complete loss of speech.² The pharyngeal and laryngeal involvement of throat, tonsils, and lips

can result in breathing problems and may predispose to upper respiratory tract infections.

The tongue may have a smooth appearance but it is thick and shortened with thickened frenulum to make the extension of the tongue difficult. Hence, all the features of difficult airway are present in LP patients.

Sufferers with this condition have hair loss affecting their scalp (alopecia) eye lashes and eyebrows.^{3,4} The skin and mucous membranes are often fragile in children with LP, leading to bleeding and scabbing following minor trauma. These problems often first appear in infancy in the mouth and on the face and limbs.

Neurologic features are also common in people with LP.^{4,5} Affected individuals may have recurrent seizures (epilepsy) or behavioral and neurological problems, which can include headaches, aggressive behaviors, paranoia, hallucinations, short-term memory loss, and absence of fear.^{4,5} Deposits can be

found in brain and some internal organs, including stomach, duodenum and the colon. The deposits in these tissues often do not cause any symptoms and may disappear over time.

In our settings, at the time of patient presentation and upon immediate literature search, no such case could be found to verify the most suitable anesthesia technique for an emergency LSCS in LP patients. The caution maintained is for structural and physical changes in the body tissue composition especially affecting the upper airway adversely, from an early stage of life.

CASE REPORT

Our patient was a 26 years old gravida 1, para 0, 32 weeks gestation presented with a complaint of abdominal and back pain and feeling of fainting in the street. On examination by the obstetricians, she had an irritable uterus but resting tone was present inbetween the contractions. CTG in progress was normal but suggested uterine contractions by clinical examination and pain suggestive of possible abruptio placentae. On the basis of obstetrician's examination, a decision was made to deliver the baby by emergency LSCS. Additionally, the baby's presentation was breech. The obstetric anesthesia team was called in to the labour room for emergency LSCS. The patient was a known case of LP since childhood. She had no specific history of surgical procedure in the past. However, at that point the upper and lower airway concerns were raised.

On clinical examination she had hoarse voice which she claimed to have since childhood. However, there was no weakness of the voice ever since. The other features included occasional dry mouth, impaired tongue mobility and thickness of the oral mucosa. ENT consultation was requested but could not be done due to the time constraint and emergency situation.

The patient had a history of difficulty in breathing and swallowing; however, on auscultation, her chest was clear with bilateral equal air entry and chest movement. There was no wheezing although the patient had a history of recurrent chest infections.

Cardiovascular and abdominal examinations were normal. In musculoskeletal system, there were obvious skin changes of LP disease e.g. a waxy yellow dermal infiltration with recurrent blistering seen on the face and extremities. Skin thickening was generalized, most obvious on the face and axilla, warty papules and plaques at site of frictions, elbow and extensor area of arms. Mild alopecia was present.

On neurological examination – patient was alert, oriented, and gave a history of an attack of generalized involuntary abnormal body movement following

distress (severe generalized dystonia) with loss of consciousness in the past. There was no history of trauma at any occasion. No tongue bite or loss of sphincter control was reported and the seizure continued for 15 min with no post ictal confusion or focal neurologic deficit. CT scan and EEG requested but could not be done.

The decision for emergency cesarean delivery was taken by two obstetricians and the patient transferred to operating room where 16G cannula was inserted with a three-way stopcock and warm ringers lactate solution started as coloadng dose (10 to 15 ml/kg). Monitoring included blood pressure, pulse, ECG, SpO₂ and CO₂ non invasively.

Under full aseptic conditions, patient was seated and skin infiltrated with inj lignocaine 1% followed by spinal anesthesia with a 26 G pencil point needle at the level L 3/4 in the first attempt. After obtaining free flow of clear CSF, 2 ml of 0.5% bupivacaine heavy 10 mg mixed with 0.3 ml (15 µg) of fentanyl injected intrathecally.

She was positioned supine with right sided wedge under the hip and the phenylephrine infusion started at 5 µg/kg/h to maintain SBP within 20% of the baseline. The NIBP was measured at 1 min intervals during and after the delivery of the baby. Upon delivery of a healthy baby girl, 5 units of oxytocin stat was given followed by infusion of 30 units infusion via pump at a rate of 125 ml/h. Apgar scores at 1 and 5 min were 9 and 10. Arterial and venous blood samples from a doubleclamped segment of umbilical cord were sent for measurement of cord blood gases. The course of LSCS under spinal anesthesia was uneventful and patient had no incidence of nausea, vomiting, discomfort or drop in blood pressure, pulse or saturation. Patient received oxygen by hudson mask throughout surgery. Upon completion of surgery, she was transferred to the recovery room where she stayed for 2 h for monitoring and Aldrete scoring; was transferred back to the ward with aldrete scores of 10. Three days post cesarean section, she was discharged home after having received uneventful and well monitored care by the obstetric and anesthetic teams.

DISCUSSION

LP is a rare autosomal recessive disorder characterized by intercellular deposition of an amorphous hyaline material. Etiology and pathogenesis are unknown. Infantile hoarseness is a common presenting feature of the disease due to infiltration of larynx. In two-thirds of the cases, voice changes are present at birth or in early infancy as the first manifestation. Several reports of LP patients have appeared mainly in the dermatology literature, emphasizing the typical dermatologic features.

emergency cesarean section and lipid proteinosis

Urbach and Wiethe described LP in 1929. The importance of LP should not be underestimated by the anesthetist and otolaryngologists and it should be included in differential diagnosis for the management of difficult airway by the anesthetist.

Recently Haider et al⁶ reported a case of emergency cesarean section in which patient did not provide history of her disease (LP) and faced difficult intubation and could only be intubated with a small ETT size 5.5 when actually she was estimated for size 7 tube. Haider et al⁶ followed that case upto the next elective surgical operation of open cholecystectomy and administered successful and uneventful regional anesthesia for the whole surgical procedure.

In our case the patient's history was known and early concern was raised about the difficulty of securing airway in case of failed spinal or urgent general anesthesia administration if required. Due to time constraints, nothing could be done to further evaluate by ENT surgeons by microlaryngoscopy or other examinations. We relied on plan A of successful spinal anesthesia and in case of emergency control all the preparation of GA with videolaryngoscopes and electively selected small size ETTs (5 to 6.5 mm ID) were readied.

The literature suggests laryngeal instrumentation and tracheal intubation should be as gentle as possible as it may cause bleeding.⁵ Difficulties with direct laryngoscopy are expected and the use of a flexible fiberoptic laryngoscope to visualize the larynx and secure intubation is advised in known elective cases. A reduced gag reflex may occur with this condition.⁷ and thus care should be taken to avoid aspiration in the postoperative phase as well. LP patients may develop epileptic seizures,^{3,8} and this may be related to the intracranial calcification well described in

UWD.^{5,8} Therefore, avoid epileptogenic anesthetic agents to induce or sedate such patients.

In conclusion, lipid proteinosis is a rare disease; a history of progressive hoarseness with age and other historical details may alert the anesthetist. Anesthetists not familiar with this disease may mistake the laryngeal findings for more common acute upper airway conditions like recurrent laryngitis or bronchitis, especially if other markers of skin lesions are absent. Skin manifestations are usually not present at birth. These may appear at any age or not at all. Due to the upper airway narrowing, authors suggest to keep surgical airway options ready i.e. tracheostomy.

Acne-like lesions are usually present on the forehead and elbows and axilla. In our case and Haider et al⁶ case, facial acne like features were present but alone could not have alerted us in the absence of a history of LP.

We recommend that a thorough history of patients with LP lookalike skin features, abnormally dry and grossly thick tongue, unexplained hoarseness and typical acne like facial lesions must be probed with further enquiry. Antisialagogue premedication and epileptogenic drugs should be avoided. Difficult intubation should be anticipated particularly when there is gross tongue involvement, which is a late manifestation of this disease.

Conflict of interest: None

Authors' contribution:

NK: Literature search, Bibliography, Patient management

KS: Manuscript writing, Literature search

IB: Manuscript review, Admin support

SDJ & ZL: Patient management

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