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A00 Anaesthetic management of a parturient with lamellar ichthyosis undergoing a caesarean section

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Introduction

Lamellar ichthyosis is a rare autosomal recessive skin condition. It is one of many ichthyoses or skin conditions associated with hyperkeratosis. There is limited literature available regarding successful anaesthetic techniques in this patient population. We describe a case of a parturient with this rare skin condition who presented for caesarean section.

Case report

A 36 year old primigravid female, known Lamellar ichthyosis, Gestational Diabetes Mellitus and Pregnancy Induced Hypertension, presented at 37 weeks and 5 days gestation for lower segment caesarean section (LSCS) due to failure to progress in labour. Prior to learning she was pregnant, isoretinoic acid was used to help manage her skin condition. She had an uneventful pregnancy and no previous surgeries. A generalised non-painful scaly rash was noted, with profound thickening of skin covering her palms, which was associated with reduced joint mobility, and bilateral ectropion. Her Mallampati score was IV, thyromental distance 6.5cm and temporomandibular joint mobility Class A. Her BMI was 35. An 18G peripheral intravenous access was obtained and secured with crepe bandage, as no medical tape available could adhere to her skin. Routine premedication of ranitidine 50mg and metoclopramide were administered intravenously. A spinal anaesthetic was performed under strict aseptic conditions, using 0.5% chlorhexidine in 70% isopropyl alcohol, with no untoward skin reactions. 2% lidocaine was infiltrated subcutaneously over the L4-L5 interspace. A 25G sprotte spinal needle was used to inject fentanyl 15mcg and 2.7 mls of heavy bupivacaine intrathecally. No adverse reactions were noted. There was loss of cold sensation to the dermatomal level T2. Standard monitoring as per AAGBI guidelines was employed. Anaesthesia was uneventful. A live male neonate was delivered with APGAR scores of 9 and 10. The patient was ambulant post operatively with full sensation and motor control of her lower limbs. After six months, she maintains full neurological function.



Image 1: Patient with Lamellar Ichthyosis showing generalized scaly rash

Discussion

Lamellar ichthyosis is a rare autosomal recessive skin condition that is estimated to affect 1 in 200,000-300,000 persons in the Caribbean and Latin America. It is caused by mutations in the epidermal Transglutaminase 1 gene, which is an enzyme responsible for cross-linking epidermal proteins during the formation and maturation of the stratum corneum. Those born with this condition are initially encased in a collodion membrane, which dries and peels off during the first few weeks of life, leaving non -bullous scaling skin. It is associated with tight skin which can split superficially or deep. This group poses many anaesthetic challenges, some of which include difficult airway and aspiration risk, difficult intravenous access, limited joint mobility and predisposition to hypothermia. A regional anaesthetic was chosen to minimise these risks. There are few documented cases about anesthetic methods employed in this patient population. Of the reported cases, four of these were done under general anaesthesia and in children. This is the first reported case of the successful conduct of subarachnoid anaesthesia in this group of patients, using standard cleaning solutions.

References

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