

Case Report

Challenges in Anesthetic Management in a 25-Year-Old Patient with Ichthyosis

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INTRODUCTION

Ichthyosis is a group of skin disorders caused by abnormalities in skin growth and shedding, leading to excessive keratinization. There are over 50 types of ichthyoses, each with varying symptoms, but all are characterized by abnormal skin cornification.¹ The condition is rare, with less than one case per 2,000 individuals.² Ichthyosis can result in skin barrier issues, eyelid ectropion, joint mobility difficulties, and reduced sweating, posing risks for infections, sepsis, and other complications.^{1,4} Prior reports have suggested that malnutrition and vitamin deficiencies are more common in patients with ichthyosis, especially with severe forms.⁵⁻⁷ Ichthyosis can be genetic or acquired,⁸ and while there is no cure, treatments focus on managing symptoms.⁹

This report describes a unique case of an adult patient with suspected lamellar ichthyosis undergoing elective shoulder surgery. The lack of pre-operative communication about the patient's condition required problem-solving strategies that led to a delay in the surgery. The patient provided written, informed consent for the publication of this report.

CASE REPORT

A 25-year-old female underwent shoulder surgery at an outpatient surgery center. The anesthesiologist noted several physical characteristics, including thickened skin, ectropion, and an eclabium, which made preparation for surgery challenging. The team used ultrasound guidance to secure intravenous (IV) access in the antecubital fossae due to non-visible and non-palpable veins. However, the patient's skin caused the IV lines to clot, requiring a new line to be inserted in the leg at a shallow angle under ultrasound guidance. The IV was sutured into place due to the patient's skin scales preventing the use of tape or other securing methods.

During induction, the patient was given lidocaine, fentanyl, propofol, and succinylcholine. Intubation was successful with a GlideScope® on the first attempt. To secure the endotracheal tube, the team tied a gauze roll around it and circumferentially wrapped it around the patient's head. Gel lubricant was applied to the eyes and covered with gauze due to ectropion. The patient was rolled laterally during the procedure, and precautions were taken to prevent the tube from dislodging. Electrocardiogram (EKG) electrodes were secured with lateral pressure from rolled-up towels for continuous cardiac monitoring.

The patient maintained stable vitals throughout the procedure, was

extubated without difficulty, and transferred to the post-anesthesia care unit. The IV was removed post-operatively.

DISCUSSION

Lamellar ichthyosis, a non-fatal form of ichthyosis, is likely the subtype affecting the patient, given her age and facial features.¹⁰ Lamellar ichthyosis typically is identified at birth by the presentation of a collodion baby with features like ectropion and eclabium.¹¹ Despite increased risks of infection, dehydration, and malnutrition, patients with lamellar ichthyosis generally have a normal life expectancy.⁴ Alternatively, the patient could have an acquired form of ichthyosis, but without a comprehensive history, determining the exact type is challenging.¹ Regardless of the specific form, long-term and perioperative management are similar among the subtypes.^{2,12}

Ichthyoses present various anesthetic challenges. In this case, the patient's skin condition made it difficult to place an IV line without ultrasound guidance. Due to the skin's inefficacy with adhesives, the team sutured the IV in place and used alternative methods to secure the endotracheal tube and EKG electrodes.^{4,13,14} Limited mobility and difficulty with intubation due to facial involvement and reduced neck extension also are considerations.^{4,5} Lubricating gel and careful positioning have been used to address these issues in previous cases.^{4,13-15}

There are no specific contraindications to anesthesia for patients with ichthyosis,^{4,5} and both inhaled anesthetics and total intravenous anesthesia have been used.¹³⁻¹⁵ However, it is important to note that systemic retinoids, a common treatment for ichthyosis, can be harmful to the liver and bones.¹⁶ Pre-operative communication about the patient's skin condition is crucial to prepare for potential challenges. Improved communication, possibly through a perioperative surgical home model, can enhance perioperative care and patient outcomes.

REFERENCES

- Gutiérrez-Cerrajero C, Sprecher E, Paller AS, et al. Ichthyosis. *Nat Rev Dis Primers* 2023; 9(1):2. PMID: 36658199.
- Marukian NV, Choate KA. Recent advances in understanding ichthyosis pathogenesis. *F1000Res*. 2016 Jun 24;5:F1000 Faculty Rev-1497. PMID: 27408699.
- Oji V, Traupe H. Ichthyosis: Clinical manifestations and practical treatment options. *Am J Clin Dermatol* 2009; 10(6):351-364. PMID: 19824737.
- Rosenbaum T, Rosenbaum ER, Lienhart KL, Mazereeuw-Hautier J, Paller A. Anaesthesia recommendations for patients suffering from Harlequin ichthyosis. *Anesthesiologie und Intensivmedizin* 2016; 57(6):S453-S460.
- Hegde HV. Anaesthesia Recommendations for Patients Suffering from Lamellar Ichthyosis. Updated May 2014. <https://www.orphananesthesia.eu/en/rare-diseases/published-guidelines/lamellar-ichthyosis/146-lamellar-ichthyosis/file.html>. Accessed March 22, 2023.
- Tsivilika M, Kavvadas D, Karachrysa S, Sioga A, Papamitsou T. Management of harlequin ichthyosis: A brief review of the recent literature. *Children (Basel)* 2022; 9(6):893. PMID: 35740830.
- Frasconi F, Dreyfus I, Rodriguez L, et al. Prevalence and risk factors of vitamin D deficiency in inherited ichthyosis: A French prospective observational study performed in a reference center. *Orphanet J Rare Dis* 2014; 9:127. PMID: 25091406.
- Majmundar VD, Baxi K. Hereditary and Acquired Ichthyosis Vulgaris. [Updated 2023 Aug 8]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK562318/>.
- Crane JS, Paller AS. X-Linked Ichthyosis. [Updated 2024 Jan 25]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK448149/>. Accessed March 25, 2024.
- Craiglow BG. Ichthyosis in the newborn. *Semin Perinatol* 2013; 37(1):26-31. PMID: 23419760.

¹¹ Wu YC, Yao Q. Lamellar ichthyosis. *JAMA Dermatol* 2023; 159(2):210. PMID: 36576743.

¹² Paller AS, Butala S. Inherited Ichthyosis: Overview of Management. UpToDate. 2023. www.uptodate.com. Accessed March 25, 2024.

¹³ Franzen MH, LeRiger MM, Pellegrino KP, Kugler JA, Rizzo WB. Sjögren-Larsson syndrome: Anesthetic considerations and practical recommendations. *Paediatr Anaesth* 2020; 30(12):1390-1395. PMID: 33037729.

¹⁴ Rosenbaum T, Rosenbaum ER, Lienhart KL, Choate AI. Obstetric anesthesia for harlequin ichthyosis: A unique challenge. *A A Case Rep* 2015; 4(2):19-21. PMID: 25611001.

¹⁵ Smart KJ, Gruffi CA, Doherty TM. Anesthetic management of a patient with harlequin ichthyosis. *Case Rep Anesthesiol* 2021; 2021:9953320. PMID: 34350038.

¹⁶ Zaenglein AL, Leby ML, Stefanko NS, et al. Consensus recommendations for the use of retinoids in ichthyosis and other disorders of cornification in children and adolescents. *Pediatr Dermatol* 2021; 38:164-180. PMID: 33169909.

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