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NAVIGATING THE DELICATE: A CASE OF EPIDERMOLYSIS BULLOSA SIMPLEX

Paediatrics

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KEYWORDS

INTRODUCTION

Epidermolysis bullosa (EB), an inherited skin condition that is not clinically distinct at birth. There are four groups that this illness falls into. Each subtype has a highly distinct prognosis, which can range from a normal life span with only mild skin affection to severe skin affection and neonatal death. [1] All of the subtypes may manifest with severe, potentially fatal blistering and varying degrees of mucosal involvement; however, the different subtypes cannot be clinically discriminated at birth. Until a certain diagnosis and prognosis are established, managing the infant presents significant ethical challenges to both the family and medical professionals. In addressing these conundrums, the moral considerations of life and "quality of life" are challenging yet crucial. The management of the patient and the family requires these inquiries and conversations.[1]

Case Report

A term SGA male (37 weeks) baby was born to a primiparous mother via normal vaginal delivery. Baby cried at birth. While resuscitating it was noticed that baby had multiple erosions of skin over the limbs which became significant on mopping. The cutaneous examination revealed multiple erosions on both foot, right leg, right knee, right hand, right elbow, left dorsum of hand and upper lip. Anonychia was present on first three toe and first four toe nails. As per the Dermatologist advice, wet sterile gauze covering (at the site of erosion) was done along with barrier nursing. Baby was handled in a sterile environment to prevent infections. Clinically the diagnosis of Epidermolysis Bullosa Simplex was made. Further plan of skin biopsy and other investigations was discussed with the attenders. Due to financial constrains attenders took the baby to other hospital where baby succumbed to neonatal sepsis.

Conclusion

Epidermolysis Bullosa Simplex is a rare and challenging condition to treat requiring interdepartmental coordination. Special handling and treatment are required because the mother and medical personnel find it difficult when a child in pain is crying and has an unknown result. In this instance, cooperation between the dermatology department—which possesses specialised knowledge about the diagnosis and treatment of EB skin conditions—and the NICU was crucial.[3] In this instance, pain control was crucial yet challenging.[4]

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