

## Lamellar Ichthyosis in a Neonate: A Case Report

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### Summary

**Okoro BA, Okeahialam TC and Orimilikwe E. Lamellar Ichthyosis in a Neonate: A Case Report.** *Nigerian Journal of Paediatrics* 1985; 12:29. The first case of lamellar ichthyosis (collodion skin) with ectropion in a male neonate seen at the University of Nigeria Teaching Hospital, Enugu is described. It is a rare congenital skin disorder which tends to recur, although cases of complete remission have been reported. This patient has remained normal six months after discharge and he is still being followed up.

### Introduction

APART from the various forms of naevi, congenital malformations of the skin are rare among Nigerians. A case of lamellar ichthyosis in a Nigerian newborn is described in this communication.

### Case Report

A one-day male infant was admitted with a history of being born with everted, reddish, puffy eyelids and excessive peeling of the skin. The mother received antenatal care; there were no complications during pregnancy and delivery. Two other children in the family, both boys, aged 5 and 2 years, were normal at birth.

Examination showed a full-term neonate weighing 4kg with a head circumference of 31cm and normal vital signs. There was generalised

oedema with marked desquamation of the skin revealing widespread erythematous areas. On the intact parts of the body, the skin was dry and thin and appeared like a brown shining collodion membrane forming tight bands around the wrists and ankles. There was eversion and swelling of the eyelids (ectropion) (Fig 1).

Haemoglobin level was 15g/dl, while culture of swabs of skin and eyelids yielded no growth. Treatment consisted of nursing care, daily baths with hibitane in water and liberal application of vaseline (petroleum jelly) 4-hourly, to the skin. The eyes were irrigated with normal saline and chloramphenicol eye drops were instilled 4-hourly. In view of the widespread desquamation of the skin, the patient was placed on prophylactic antibiotics using *Ampiclox* syrup, 125mg 6-hourly, for 1 week. *Abidec* 0.3ml daily was also given.

By the 10th day, the desquamation of the skin was complete and a new, dry but non-scaly epidermal layer appeared (Fig 2). The infant was discharged after two weeks and at the 6th month of follow-up, the skin had remained normal.

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*Fig 1. Newborn with lamellar ichthyosis. Note glistening film over the hands and feet, which is markedly thickened over the left wrist. The collodion skin is beginning to exfoliate on the trunk.*

### Discussion

Lamellar ichthyosis is a congenital abnormality of the skin. It is inherited as an autosomal recessive trait and is present at birth. According to Brocq<sup>1</sup>, this condition was first reported by Vidal while a review of the subject was published in 1917 by Mackee and Rosen<sup>2</sup>. Since then, more cases have been reported in the literature<sup>3-5</sup>.

There is paucity of information on the occurrence of lamellar ichthyosis of the newborn in Africa as a whole. This may be due to the

rarity of the condition, or to failure in publishing cases seen. This is the first case to be reported in this hospital and it indicates that this condition though rare, occurs in our environment. Infants with lamellar ichthyosis have been called "collodion-babies" because at birth, the thickened stratum corneum forms a collodion-like envelope on the entire body. This film of stratum corneum is quite tight and tends to evert the lower eyelids causing ectropion and the lips causing eclabion<sup>6</sup>; the latter was not seen in our patient.

Ectropion involving all four eyelids is rare in lamellar ichthyosis<sup>8</sup>; in our patient, only the two upper eyelids were affected. The sequel of



*Fig 2. Newborn with lamellar ichthyosis. The collodion skin has completely exfoliated.*

ectropion varies from spontaneous resolution as in this patient to very severe complications requiring surgical correction<sup>8,9</sup>.

The outcome of the skin manifestation is variable. Reed *et al*<sup>4</sup> described 2 cases of lamellar ichthyosis with complete clearing of the skin and who remained so for life and one case that subsequently developed cataract. Wilkinson<sup>5</sup> carried out a 21-year follow-up of a patient whose skin showed transition from a "collodion membrane" appearance to a state of mild and regional ichthyosis which affected mainly the trunks and flexures. In our patient, the skin has remained normal and a long-term follow-up will be required to monitor any future changes.

#### References

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