CONGENITAL ICHTHYOSIS

(Harlequin Foetus—A Case Report)

by

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Introduction

The Harlequin foetus is the most severe form of congenital ichthyosis. The condition is very rare. It is because of foetal membrane which subsequently splits and desquamates. The case reported because of its rarity.

Case Report

The Patient S.R. aged 20, residence of Narsala, Pipla was admitted as a emergency case at Government Medical College Hospital on Dated 24-9-80. She had amenorrhoea of 9 months and had labour pains since early morning. She was primigravida, married 1 year back and not sure about her last menstrual period. On examination, her vital parameters were within normal limits. Per abdominal examination revealed, uterus of 34 weeks size, foetus was in left vertex anterior position, Foetal heart sounds were regular. Patient was having strong uterine pains. Per vaginal examination showed that cervix was fully effaced, fully dilated, membranes were absent, and head was at 'O station. Sutures were on L.O.A. position. Pelvis was adequate.

Pt. delivered a herlequin female foetus on 24-9-80 at 12.45 A.M. weighing 1 Kg. 750 grams. (as shown in pic) Baby expired on 3rd day. Patient was not wiling for post-mortem of the baby.

Dsicussion

In congenital Ichthyosis, the body is completely covered with thick firm abnormal skin in which marked fissuring may occur. Bilateral ectropions are frequently present in the severe cases, being due to contraction of the abnormal skin. The ears may also be completely closed.

Microscopic appearance of skin shows marked hyperkeratosis with many single nuclear horny cells present in aggregation. The sweat and sebaceous glands were decreased. Most infants with this extreme form of disease are either stillborn or die soon after birth.

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See Fig. on Art Paper VII