Pregnancy in a Patient with Congenital Lamellar Icthyosis

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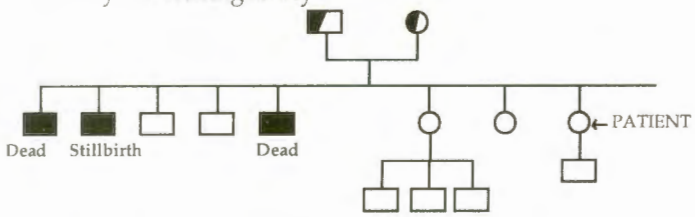
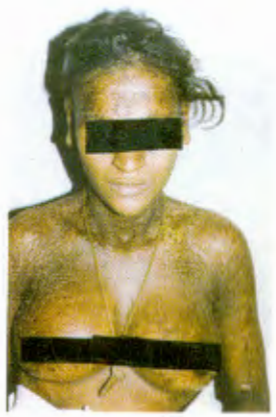
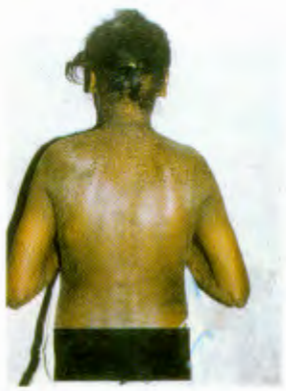
Mrs. Nagavalli, 28 year old primigravida a known case of congenital lamellar icthyosis on regular treatment presented at our antenatal out patient department with 9 months of amennorhoea and mild labour pains. She was booked and immunized in a health center. The patient was on a regular treatment with liquid paraffin and vitamin supplements for lamellar icthyosis from dermatology clinic of Govt General Hospital. The patient was the youngest of 8 children. Her parents had history of consanguinity.



On examination, patient has generalised scaling, hyperkeratosis with mild erythema. No bullae were seen. Her eyes and ears were normal. Scalp and body showed sparse hair. Patient was not anaemic and had no pedal edema. Her systemic examination was unremarkable. Per abdomen examination - uterus was 36 weeks, contracting, head unengaged; Foetal heart was good. Per vaginal examination revealed uneffaced cervix, os was closed, pelvis was average. She was admitted and baseline blood investigations were done. They were found to be within normal limits. Ultrasound showed single cephalic foetus of 36 weeks gestational age; foetal heart and movements were good. On the 5th day of admission, she complained of labour pains and was sent to labour room. She ruptured her membranes in labour ward.

In view of first degree of cephalopelvic disproportion emergency LSCS was done. Alive male baby weighing 2.8 kg was delivered. The paediatrician





found the baby healthy with no evidence of congenital icthyosis. Her postoperative period was uneventful and she was discharged on 10th day. She was advised to come for regular check up.

Congenital lamellar icthyosis is a rare autosomal recessive disorder of keratinisation. This disorder is characterized by striking degrees of hyperkeratosis but not much erythema with ectropion and ear deformities.