LAMELLAR ICHTHYOSIS OF THE NEWBORN

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Summary

A case of Lamellar ichthyosis of the newborn presenting with characteristic Clinical features is reported. Literature on the entity is reviewed.

Lamellar ichthyosis of the newborn is a rare form of congenital dermatoses which is characterised by a parchment like membrane completely enveloping the infant at birth. It is responsible for the so called collodion or Harlequin foetus. Immediately after birth, small fissures appear on the body surface and desquamation of the membrane begins with large keratinous lamellae being cast off. The word ichthyosis is derived from ichthys, Greek for fish and lamellar is diminutive of lamina, Latin for a thin plate, Thus in cases of Lamellar Ichthyosis large flat scales cover most of the body.

Review of Literature

The condition was first described by Seeligmann in 1841[1] under the title of "Epidermal Desquamation of the Newborn". The descriptive term of 'Lamellar Ichthyosis of the Newborn' was coined by Grass and Torek in 1895[9]. In the same year Ballantyne[3] described in detail a case of mild type of foetal ichthyosis and reviewed 33 cases which were reported until 1895 under many different names. Shelmire in 1955[4] collected another 15 cases from the world literature. Bloom and Godfried[5] reviewed 19 cases reported between 1955 and 1962 and added one of their own. Lenz and Altmen[6] reported 4 cases in 1968. The total number of reported cases of this entity were estimated at 103 in 1968[6].

Case Report

A 34 years old female was referred for an X-ray of the abdomen for suspected twin pregnancy. She was 4th gravida ; previous 3 children, all males, were normal. There was no history of consanguinity between the parents. Posteroanterior view of the abdomen showed a single foetus presenting by breach. No congenital anomaly was detected on the X-ray.

A female infant weighing 6 lbs. was delivered, and started breathing spontaneously. It presented a most grotesque appearance. The face showed severe ectropion of upper eyelids which only partially covered the eyeballs and eclabium of the lips with an open immobile mouth. The nose was effaced making the nasal orifices very prominent. The ears were flat and malformed and appeared as if glued to the scalp. Another striking feature was the leathery hard collodion like membrane which covered the whole body. This was apparently responsible for flexion of the extremities and eclabium of the labia majora. The horny sheet of skin had developed numerous wide deep fissures with exfoliation of large and

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small scales exposing patches of normal soft and reddish brown skin. The whole skin surface had a variegated appearance. The scalp was sparsely covered with hair but nails were normal in appearance.

The baby made groaning noises and expired within a few hours. (Fig. 1 Page No. 50).

No abnormality of the heart, lungs or other viscera was found on general examination. The skeletal survey of the newborn failed to reveal any radiological abnormality. Autopsy could not be carried out.

Discussion

Lamellar Ichthyosis of the Newborn is a severe form of dermatoses which is inherited as an autosomal recessive characteristic. Therefore neither parent manifests the disease. The disorder is usually present at birth. At the time of delivery the thick stratum corneum is completely hydrated, soft and flexible as a result of its immersion in amniotic fluid. As it becomes dehydrated after birth, it assumes parchment like appearance which has led to the name of Collodion baby for infants born with lamellar ichthyosis. The dry scaly mantle cracks and is shed in sheets. The rigidity and tautness of the skin of the face may cause eclabion and ectropion. Feeding problem and death primarily from systemic infections have been reported.

In mild varieties of the condition, throughout the neonatal period and adulthood, large coarse quadrangular scales frequently adherent at the centres with slightly raised edges cover most of the body. Although severe scaling is usually not noticeable on the face, palms or soles, considerable hyperkeratosis is present in these locations which gives the skin a thickened appearance. In some cases the helices of the ears are partially adherent to the scalp. This may be the consequence of scarring following infection which is common in this region during childhood. The greatly thickened horny layer of the skin provides a suitable medium for bacterial growth and resultant intercurrent infections.

Following desquamation of the membrane, this condition gradually evolves into a chronic dermatoses the nature of which varies from case to case. The eventual outcome may lie within a spectrum which ranges from complete clearing, through a localised scaling hyperkeratosis, to one of much greater severity involving the entire integument. The case being reported here, falls in the latter category.

We feel that lamellar exfoliation of the newborn is not as rare an occurrence as one would tend to believe from the reports in the literature. The disease may remain undiagnosed particularly in patients with minor involvement which clears within a short time. The severely affected foetus is either stillborn or dies in the early neonatal period before a correct diagnoses can be established.

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