EPIDERMOLYSIS-BULLOSA

Two Case Reports

By

P. NAGABHUSHANAM,*

Epidermolysis-bullosa is a rare hereditary cutaneous disorder in which adherence of epidermis to dermis is poor. The salient clinical manifestations are appearance of non-inflammatory asymptomatic vesiculobullous lesions at the site of trauma or friction, leaving pigmented or depigmented scars after healing. These appear at the time of birth over the presenting part, in early infancy over the back and knuckles, during crawling age on knees and elbows, and during childhood on feet, palms and soles. E. B. aquisita probably represents a delayed variant of this genetically determined disease during adolescence and old age. Weber-Cockayne syndrome probably may be modified type of E. B. where lesions appear during adolescence and are confined to hands and feet.

Touraine and Leinbrock describes three forms :—

1. E. B. Simplex (Dominant) where bullae heal without scarring. Mucous membrance and nails are rarely affected, and often the individual improves at puberty.

2. E. B. Dystrophica (a) Hyperplastica (Dominant) where bullae heal by mild scarring and few milia may be seen on the scars. Rarely mucous membrance and nails show thickening. (b) Dysplastic (Recessive) Extensive atrophic scarring occurs. milia on scars are numerous. Mucous membrane is involved leading to deformities.

3. E. B. Letalis (Recessive) is lethal to life. Child usually dies below 3 months of age. The bullae show little tendency to heal.

Subepidermal bullae are pathognomonic of the disease but rarely in older lesions of simple and hyperplastic type they may be seen suprabasal and even subcorneal due to early regeneration of epidermis. The absence of elastic tissue is more due to the disease, rather than the cause. The defect is primarily due to vascuolar degeneration of the basal cells.

This disease should be differentiated from Impetigo bullosa, bullous-syphiloderm, juvenile-dermatitisherpetiformis, acrodermatitis-enteropathica during early childhood, while during adolescence should be differentiated clinically from pemphigus vulgaris, dermatitis herpetiformis, erythema multiforme and drug eruption.

As this disease is hereditary, attention should mostly be directed towards prevention of trauma and infection.

Case Report No: I.

Master B. S., a Hindu, vegetarian boy aged 13 years was admitted into the ward of Skin Department on 20-11-66 for vesiculo-bullous lesions all over the body which appear and disappear spontaneously, since the age of 12 days after birth.

*Professor of Dermatology, Gandhi Medical College, Hyderabad. AP.

Received for Publication on 25-10-1968.

According to the patient's parents, the boy was healthy till 12 days after birth, when he developed bullous lesions first over the ankles and legs and later over the buttocks. As the child grew up the bullae started developing all over the body. A week after their eruption, they formed scabs which used to fall off, leaving pigmented and depigmented lesions. The vesicles also appear below the nails of fingers and toes resulting in dystrophy and destruction of nails. There is no itching over the lesions. There are no contractures or any other deformities seen.

The prenatal, natal and post-natal history is not signigicant and non-contributory.

The parents of the patient are living and healthy. No history of similar disease is forthcoming in near relatives either on the paternal or maternal side. The patient is the eldest of the offspings of the parents, with two younger sisters aged 9 and 4 years, living and healthy.

On examination, the boy is moderately nourished with body measurements appropriate to age. No congenital abnormalities observed. Pulse 80/mt., temp. 98.5°F. B. P. 110/70. The teeth are irregular and dystrophic.

The skin over the trunk and extremities shows plenty of pigmented and depigmented areas. At present, bullae are seen over the toes and over the medial aspect of right foot. The nails of the fingers are grossly dystrophic and are elevated from their nail beds. The toe nails are rudimentary and dystrophic. All the other systems are normal.

PHOTOS NO: II & III. Dystrophy of toes and fingers.

The following investigations were carried out:—

Total W. B. C. count	..	10,800 cell/c. mm,
Differential count	..	P57 L32 E 11
Haemoglobin	..	11.5% sahle.
R. B. C. count	..	3.8 millions/c. mm.
E. S. R.	..	35 mm/1st hour
Urine	..	No albumin No Sugar Straw coloured
Urine for prophyrins	..	Negative, both clinically and by spectroscopy.
Stool...NAD	..	VDRL Negative
Serum proteins	..	4.8 G%
Bullae fluid for absolute eosinophil count	..	6 cells/c. mm.

PHOTO NO: IV. Histopathology of vesicle.

Skin biopsy of vesicle-shows sub-epidermal split, with degeneration of basal cell layer. No cellular infiltrate in upper dermis.

Case Report No: II.

Miss V, a christian, Non-vegetarian girl aged 12 years was admitted in the Ward of the Skin Department on 6-5-1968 for recurrent vesiculo-bullous-eruptions all over the body since five years, with delayed healing.

According to the patient's parents, the girl was healthy till she was six years old, when she developed rash all over the body with itching, which subsided after few injections.

Later she developed vesicule-bullous lesions, first on both legs and later on the trunk. The vesicles were taking long time to heal, leaving atrophic scars.

The prenatal, natal and post-natal history is not significant and non-contributory.

The parents of this patient are living and healthy. No history of similar disease is forth-coming in near relatives either on the paternal or maternal side. The patient is the 9th of offsprings of the parents with five elder and one younger sister, and 3 elder brothers. The eldest sister died during the 9th month of her age, and eldest brother had neonatal death. No history of similar disease in other members was elicited.

On examination the girl was undernourished and anemic. No congenatal abnormalities were seen. The teeth were slightly irregular and dystrophic. Functional systolic murmur was heard at base of the heart. Plenty of atrophic depigmented scars mainly on elbows, knees, legs, thighs, ankles, and trunk were present. A chronic ulcer on left lateral malleolus is seen. No mucosal involvement was observed.

The following investigations were carried out:—

Total W.B C. count.	..	3500 per cmm.
Differential count	..	N52 L43 M1 E4
Haemaglobin.	..	7 gram%
R. B. C. count.	..	2.85 million per cmm.
Culture of Bullousfluid.	..	Sterile.
E. S. R.	..	48 mm/first hour.
Serum Proteins.	..	6.5 gm%.
A. C. Ratio:	..	Alb. 3.6 gm. Globulin 2.9 gm.
Urine for Prophyrins.	..	Negative.
Biopsy.	..	Sub-epidermal bulla compatable with Epidermolysis bullosa.

SUMMARY

Two case reports of Epidermolysis-Bullosa have been reported.

The diagnosis is confirmed histo-pathologically. The only lacuna is that similar history in other members of the family is not forth-coming. Most probably the disease may be latent in the family.

REFERENCES

1. Lever-Histopathology of Skin.
2. Pillsbury-Text book of Dermatology.