Junctional Epidermolysis Bullosa in a Neonate

A term male newborn presented with extensive skin peeling with multiple areas of blister formation since first day of life. There was extensive peeling of the skin affecting about 40% of body surface area. The oral mucosa was also involved. There were large areas of flaccid bullae formation (***Fig.* 1**). Initial differentials considered were: bullous impetigo, staphylococcal scalded skin syndrome and epidermolysis bullosa (EB). He was treated with intravenous cefotaxime, amikacin and cloxacillin; cultures from blood and wound were subsequently reported as sterile. The sepsis markers were also negative. Skin biopsy revealed blister formation at dermo-epidermal junction with no inflammatory cells in the blister, suggestive of junctional epidermolysis bullosa (***Fig.* 2**). Minimal handling and daily dressing with vaseline gauze were done. Child died on 40th day of life due to extensive involvement and secondary sepsis.

The differential diagnosis for a neonate presenting with blisters are: bullous impetigo, staphylococcal scalded skin syndrome, epidermolysis bullosa and

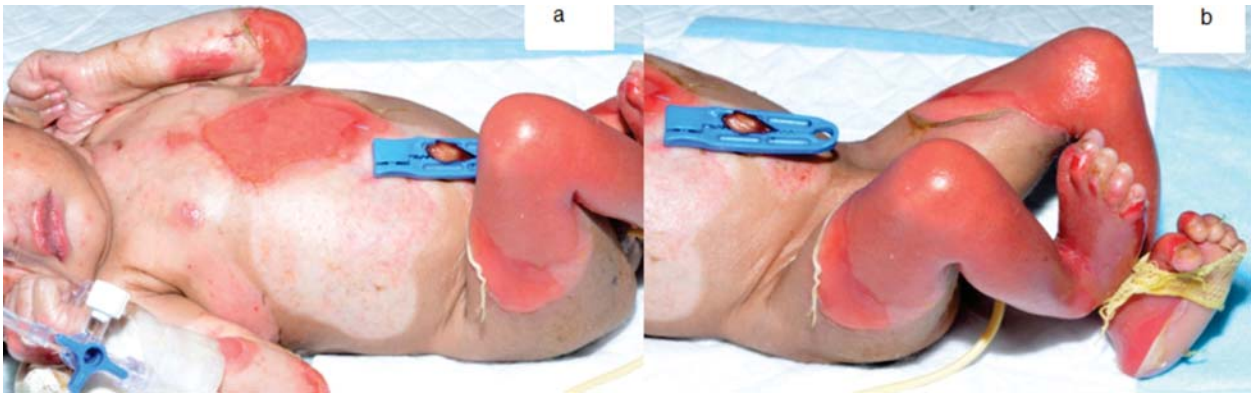


FIG. 1 *Neonate showing extensive blistering with erosions over the face, trunk and extremities.*

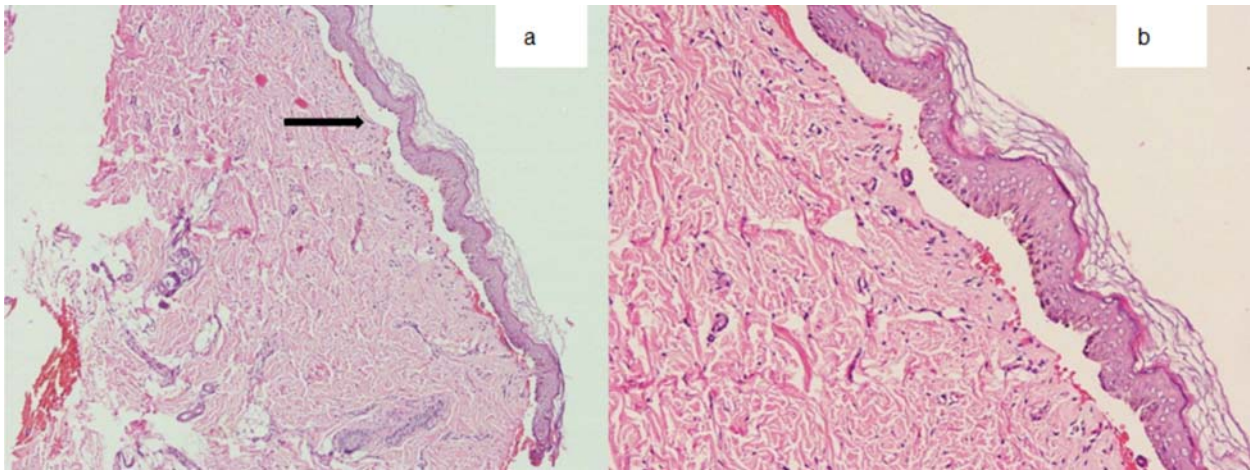
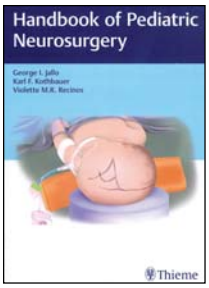


FIG. 2 *Photomicrograph of skin biopsy (haematoxylin and eosin stain: 10 X magnification) showing clefting and blister formation at dermo-epidermal junction (arrow) (a), with no acantholytic cells or inflammatory cells within the blister (b).*

bullous pemphigoid. Junctional epidermolysis bullosa (JEB) is a rare autosomal recessive disorder due to mutations in the gene coding for Laminin 332 causing blisters in Lamina lucida. Mechanical fragility at birth is the hallmark of the disease characterized by extensive blistering of the skin associated with crusting and erosions. Skin biopsy is the investigation of choice for the suspicion of bullous disorders in the neonatal period. Light microscopy helps to exclude the other infective bullous disorders; immunofluorescence helps to diagnose the immune-mediated bullous disorders. Electron microscopy helps in identifying the level of blister formation within the dermo-epidermal junction. Finally genetic testing helps in confirmation of the diagnosis. Treatment includes proper wound care, prevention of secondary bacterial infections, adequate nutrition and prevention of dehydration. This case probably had Herlitz type JEB, presenting in neonatal period with extensive blistering and a lethal course.

SANTHIYA SRINIVASAN, INDAR KUMAR SHARAWAT AND LOKESH SAINI*
Pediatric Neurology Unit, Department of Pediatrics, PGIMER, Chandigarh, India.
**drlokeshsaini@gmail.com*

BOOK REVIEW



Handbook of Pediatric Neurosurgery

GEORGE I JALLO, KARL F KOTHBAUER AND VIOLETTE MR RECINOS

Thieme Publisher, Germany.
Pages: 514; Price: Not mentioned.

This book contains 11 sections and 56 subsections. All put together, there are more than 300 chapters contributed by galaxy of Neurosurgeons and allied Neuro-scientists, numbering over 80. The book covers various aspects of Pediatric Neurosurgery, including basic aspects like intracranial pressure (ICP), pain management, imaging, intervention and chemo- and radio-therapy. It also covers pediatric Neuro-trauma, including spinal, peripheral nerve and brachial pexius injury.

First two sections deal with ICP monitoring and pain management. Knowledge of ICP, its pathophysiology, monitoring and management is like ABC of Neurosurgery. The radiology and imaging aspects are dealt to the points, and will help the readers. Authors of this book have emphasized on the value of patient examination and Clinical neurosurgery, which is amply exemplified by the chapters.

The section of tumors is extensively dealing with various intracranial and spinal tumors having 11 chapters. All important pediatric Neurosurgical tumors are discussed optimally with up-to-date relevant literature. I am sure that by reading this portion, a student or a practicing pediatric-neurosurgeon will not look for anything else. Chapters on vascular abnormalities and malformations are also well written with illustrations and surgical techniques. There is a section on epilepsy surgery and various surgical methods for intractable epilepsy, which also deals with vagal nerve stimulation, deep brain stimulation, various movement disorders and surgical treatment of spasticity.

Overall this book is a landmark publication by eminent authors. The book has larger number of contributors from various parts of the world with plenty of colored illustrations. I am sure this book will have a universal acceptance, and will immensely benefit our friends dealing with pediatric neurosurgical problems.

AK MAHAPATRA
Director, IMS & SUM Hopsital
SOA, Bhubaneswar, Odisha, India.
akmahapatra22000@gmail.com