

Chronic Bullous Disease of Childhood

A 9-year-old boy presented with itching and bulla formation since the last 3 years. The lesions commenced over the abdomen, scalp and then became generalized. Past perinatal and drug history were uneventful. On examination, tense bullae (0.5 cm to 3.5 cm in diameter) were present all over the body (**Fig. 1**). Bullae arranged in an annular pattern were seen in perineal region (string of pearl's sign) (**Fig. 2**). Bulla spread and Nickolsky sign were negative. Tzanck smear was negative for acantholytic cells. Based on above findings, a differential diagnosis of chronic bullous diseases of childhood, childhood bullous pemphigoid and dermatitis herpetiformis were considered. Skin biopsy revealed

bulla formation between basal layer of epidermis and dermis, papillary dermal edema, dilated blood vessels with mild perivascular inflammatory infiltrate comprising of lymphocytes, neutrophils and eosinophils. Direct immunofluorescence test (for which patient was not affording) is confirmatory for Chronic bullous disease of childhood, demonstrating linear deposition of IgA at basement membrane zone. While this disease has been defined on the basis of its unique immunopathology and occurs in both adults and children, however, in children the cutaneous features may be clinically unique. The disease responds to dapsone at less than 0.5 mg/kg/day, with regular monitoring of hemoglobin levels. Spontaneous remission occurs after 3-6years.

RASHMI MAHAJAN, PRIYANK SHAH AND SHEELA BHARANI
*Department of Dermatology, SBKS Medical College and
 Research Centre, Gujarat, India.
 rsoodmahajan@gmail.com*



FIG.1 Bullae all over body.



FIG.2 String of pearl sign.