SUBCORNEAL PUSTULAR DERMATOSIS IN A CHILD WITH LEUKEMIA

K Pavithran

A 3½-year-old male child with acute lymphatic leukemia developed pustular eruptions. The clinical course, histopathological study and therapeutic response to dapsone suggested a diagnosis of subcorneal pustular dermatosis.

Key words : Subcorneal pustular dermatosis, Sneddon-Wilkinson disease, Acute lymphatic leukemia.

Subcorneal pustular dermatosis, originally described by Sneddon and Wilkinson[1] in 1956, is characterized by chronic recurrent pustular eruptions occurring most frequently in middle aged women. Rarely, it may develop in children also.[2-3] The youngest patients reported were 3-year-old when the disease was recognized.[3] Here, we report a 3½-year-old male child who developed subcorneal pustular dermatosis simultaneously with the early signs and symptoms of acute lymphatic leukemia.

Case Report

A 3½-year-old male child was seen in the paediatric department of this hospital for haematuria and an asymptomatic pustular eruption of one month duration. Examination revealed pallor, generalised lymphadenopathy and hepatosplenomegaly. There were multiple, bilateral, superficial, oval, flaccid pustules on the abdomen, axillae and groins. The pustules on the abdomen remained discrete, while those on the axillae and groins coalesced to form annular and serpiginous crusted lesions. Nikolsky sign was absent. The face, limbs and mucous membranes remained unaffected.

Blood haemoglobin was 8 gm%, ESR 58 mm and total leucocyte count 18,000 cells per cubic mm. More than 80% of the cells in the peripheral smear were lymphoblasts. Urine showed presence of red blood cells and albumin. Blood urea was 80 mg%. X-ray of the chest and long bones did not show any abnormality. Repeated cultures of the pustules were sterile and cytology did not show acantholytic cells. Histopathological study of the skin lesions showed subcorneal blisters filled with neutrophils and serous fluid. There were no leukemic cells in the blister fluid. Dapsone given in a dose of 50 mg twice a day for 7 days caused complete subsidence of the skin lesions. Meanwhile, the condition of the patient deteriorated and dapsone was discontinued resulting in a relapse of the pustular eruptions. Before receiving any specific treatment for leukemia, the child died of cerebral haemorrhage.

From the Department of Dermato-Venereology, Medical College Hospital, Kottayam-686 008, India.

Comment

The morphology and evolution of the skin lesions, their histopathologic features and therapeutic response to dapsone suggested a diagnosis of subcorneal pustular dermatosis (SCPD). Our patient was 3½-year-old, male while, Johnson and Cripps[3] reported cases of SCPD in two, 3-year-old children, a boy and a girl. Beck et al[4] in 1961 reported a case of SCPD in a 29-year-old woman who gave history of onset of her skin disease at the age of three months. The cause of this dermatosis is unknown. The role of trigger mechanisms such as preceding or concomitant infections though repeatedly discussed, has remained speculative. Recently, immunologic mechanisms have been implicated in its pathogenesis. Krogh and Tonder[5] could detect anti-stratum corneum antibodies in two cases. Tagami et al[6] observed IgA deposits at the upper epidermis of the paralesional skin. Wallach et al[7] in 1982, observed circulating

monoclonal IgA KAPPA immunoglobulin in a patient with SCPD and speculated that IgA deposition might be implicated in its pathogenesis.

The occasional association of this dermatosis with certain other diseases may represent more than a mere coincidence. Some of the diseases reported in association with SCPD include systemic lupus erythematosus, pustular psoriasis, rheumatoid like crippling arthritis, erythema multiforme, multiple IgA myeloma, and pyoderma gangrenosum.[8-13] Whether or not the coexistence of these conditions reflects common pathogenic mechanism remains to be clarified. In the present case the development of SCPD simultaneously with the early signs and symptoms of acute lymphatic leukemia appears to be more than fortuitous. Either the concomitant leukemia in this patient has acted as a triggering factor for inducing SCPD or some immunologic abnormality associated with leukemia has played a pathogenic role in its development.

References

1. Sneddon IB and Wilkinson DS : Subcorneal pustular dermatosis, Brit J Dermatol, 1956; 68 : 385-394.
2. Bedi TR and Bhutani LK : Subcorneal pustular dermatosis in a male child, Castellania, 1975; 3 : 35.
3. Johnson SAM and Cripps DJ : Subcorneal pustular dermatosis in children, Arch Dermatol, 1974; 109 : 73-77.
4. Beck AL, Kipping HC and Crissey JJ : Subcorneal pustular dermatosis : Report of a case, Arch Dermatol, 1961; 83 : 629-630.
5. Krogh HK and Tonder O : Subcorneal pustular dermatosis : Pathogenetic aspects, Brit J Dermatol, 1970; 83 : 429-434.
6. Tagami H, Iwatsuki K, Iwase Y et al : Subcorneal pustular dermatosis with vesiculo-bullous-eruption : Demonstration of subcorneal IgA deposits and a leukocyte chemotactic factor, Brit J Dermatol, 1983; 109 : 581-587.
7. Wallach D, Cottenot F, Pelbois G et al : Subcorneal pustular dermatosis and monoclonal IgA, Brit J Dermatol, 1982; 107 : 229-234.
8. Sanchez NP, Perry HO, Muiler SA et al : Subcorneal pustular dermatosis and pustular psoriasis : A clinicopathological correlation, Arch Dermatol, 1983; 119 : 715-721.
9. Saulsbury FT and Kesler RW : Subcorneal pustular dermatosis and systemic lupus erythematosus, Intern J Dermatol, 1984; 23 : 63-64.
10. Olsen TG, Wright RC and Lester AI : Subcorneal pustular dermatosis and crippling arthritis, Arch Dermatol, 1979; 115 : 185-188.
11. Sneddon IB : Subcorneal pustules in erythema multiforme, Brit J Dermatol, 1973; 88 : 605-607.
12. Cream JJ, Grimes SM and Robert PD : Subcorneal pustular dermatosis and IgA myelomatosis, Brit Med J, 1977; 1 : 550.
13. Wolff K : Subcorneal pustular dermatosis (Sneddon-Wilkinson): Pyoderma gangrenosum mit IgA—Paraproteinemia, Dermatol Monatschr, 1971; 157 : 842.