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URTICARIA PIGMENTOSA

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A case of urticaria pigmentosa with bullous lesions in a 2-year-old male is presented. No systemic manifestation of mastocytosis was found.

Key words : Urticaria pigmentosa, Mastocytosis.

Mastocytosis means local and systemic accumulations of mast cells, while the term urticaria pigmentosa is used for cutaneous disease only. Nettleship[1] first described the disease in 1869, and Sangster[2] coined the term urticaria pigmentosa. Unna[3] demonstrated the relationship between mast cells and urticaria pigmentosa. Since then, over 600 cases have been reported. It may occur at any age from birth to middle age. Approximately 10-20% of the cases appear between 1-2 years and puberty. In 25-45% of patients, it starts in adult age while half the cases have their onset before six months of age.

Cutaneous lesions may consist of macules, papules, nodules, verrucae, vesicles or bullae, petechiae or ecchymoses, and plaques and telangiectasia. Symmetry and monomorphism are unusual. The most diagnostic sign is Darier's sign i.e. when lesions are firmly stroked or rubbed, these develop urticaria with surrounding erythematous flare. Bullous lesions are more frequent in infants and children because large collections of metabolically active mast cells are more commonly found in babies, and infantile skin blisters more readily. In some cases of mastocytosis, the systemic complications overshadow the significance of the cutaneous involvement.[4,5] However, in erythrodermic mastocytosis, it is not necessary that systemic disease should also occur.

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Caplan[6] described three types of the disease : (1) Solitary lesion (10% cases); (2) Multiple lesions with onset in infancy or childhood that tends to clear up with adolescence (65% cases), and (3) Multiple lesions that develop in adults (25% cases).

A rare case of urticaria pigmentosa with bullous lesions is being reported.

Case Report

A 2-year-old boy was having itching all over the body with flushing, bullous lesions and ulcers for the last 6 months. Urticarial wheals would develop over the body after mechanical trauma. The patient was born normal and of non-consanguinous parents. His mile-stones were normal. There was no history of gastro-intestinal disturbances, mucosal ulcerations or bleeding tendencies. The patient was found to have flushed skin with multiple yellow to dull brown macular, papulo-nodular, vesicular and bullous lesions, scattered all over the body, but especially on the abdomen, back and neck. The bullous lesions were 6-8 in number, 2-3 cm in size, surrounded by erythema and soft but not easily ruptured. Ulcerated lesions were present on the right side of neck and back. Limbs and face were comparatively spared. Urticarial wheals were also present at a few places, and Darier's sign was strongly positive. Dermographism of clinically uninvolved skin could also be demonstrated. Mucous membranes were not involved. Hair and nails were normal.

There was no lymphadenopathy, hepatosplenomegaly, abdominal tenderness, or joint or bone pain on movements.

Routine urinalysis, haemogram, platelet count, reticulocyte count and bleeding or clotting times were normal. No abnormal or immature cells were seen in the peripheral smear. Liver function tests and serum calcium were normal. VDRL test on blood was negative. X-rays of skull, pelvic girdle, shoulder girdle, hands and feet did not show any abnormality. Urinary porphyrins were negative.

Histopathology of a lesion showed a dense aggregate of large mononuclear cells with an abundant basophilic cytoplasm. Metachromatic granules were seen on toluidine blue staining. The bulla was sub-epidermal.

Treatment with cyproheptadine (6 mg daily for 3 weeks),cimetidine(400 mg daily for 3 weeks), terbutaline (4.5 mg daily for 3 weeks), prednisolone (15 mg daily for 3 weeks and then gradually tapered in 10 days period), and erythromycin (100 mg 4 times a day for 10 days) induced complete remission. Patient on follow up treatment was given cyproheptadine 4 mg per day. Patient came regularly for 6 months for check up and was in complete remission with 2 mg cyproheptadine only.

Comments

In our patient, there was no evidence of systemic mastocytosis, only the cutaneous lesions were present. The prognosis in such cases is good. About half of the cases clear completely, while the others improve considerably. Some may persist indefinitely.

Bullous urticaria pigmentosa is a rare entity. Fox[7] first reported bullous urticaria pigmentosa in 1883. Robinson et al[8] reported 5 cases of bullous urticaria pigmentosa while Miller et al[9] reported 2 infants with only bullous lesions. The patients reported by Joy et al[10] and Haribhakti et al[5] did not have bullous lesions. The diagnosis can be confirmed histopathologically.

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