URTICARIA PIGMENTOSA AND DIARRHOEA
(A case report)

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Urticaria pigmentosa is the disease of skin caused by excessive infiltration of Mast cells. This disorder was first described by Nettleship[1] as chronic urticaria leaving brown stains.

Ellis[2] showed that this disease could become systemic and fatal when he presented the autopsy findings of an infant suffering from urticaria pigmentosa.

Till now about 600 cases have been reported in literature but the exact etiology of the disease has not been clearly understood.

The disease has been seen in identical twins. Involvement of many sibs in the same family has been reported (Beatty et al[3] and Shaw et al[4]). On the other hand urticaria pigmentosa discordant in identical twins has also been reported (Gay et al[5] and Selmanowitz et al[6]).

Transmission of this disease has been suggested as autosomal dominant with reduced penetrance. The case presented here, however, did not have any such history in the family but in addition had chronic diarrhoea not responding to common antidiarrhoeal drugs.

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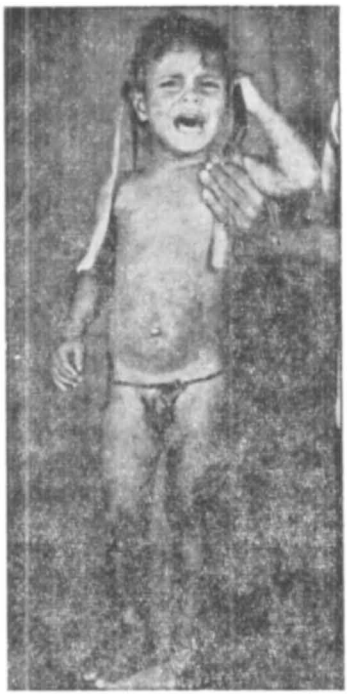
Case Report

A 2½ years male came to the skin O.P.D. of Institute of Medical Sciences, B. H. U., Varanasi, India in the month of October 1971 with history of recurrent itching, erythematous, papulo vesicular lesions appearing since birth. The lesions when heal leave behind hyperpigmented macules.

The parents of the child noticed recurrent erythematous eruptions over the face when the child was 2 months of age. Gradually the number of lesions started increasing and the lesions went on extending on to the trunk and extremities. By the time the child was of 6 months he started having recurrent crops of these lesions on the face, trunk and both upper and lower extremities. The child became restless and he developed severe itching. Itching would produce more wheals and that in turn caused more itching. The child had taken treatment from different practitioners but not to much use. During all this period the child also continued to have diarrhoea for which no other cause could be ascertained. During the neonatal period the child used to have respiratory difficulty and attacks of sneezing. No other member of the family had similar or any other allergic disease.

On examination the child was average built, very irritable and the whole body was studded with hyperpigmented macular lesions. (Fig.) Palm, soles and



Irritable child with erythematous hyperpigmented maculo-papular lesions

mucous membranes, however, were spared. On gentle massage on the back the wheals appeared at the site of hyperpigmented macules and the child started scratching. The size of the lesions varied from 0.5 cm. to 1.5 cm. Dermographism was highly positive. No hepatosplenomegaly or lymphadenopathy could be elicited.

On investigation the child had normal haemogram, urinalysis and stool examination. Skiagram of the chest was normal. Platelet count, bleeding and clotting time were within normal range. Blood histamine levels were very much raised.

Skin biopsy revealed large number of mast cells in the upper part of dermis and around appendages and blood vessels.

The child was put on cyproheptadine hydrochloride 2 mg. three times daily for about 2 weeks. The child improved. Bowels got stabilized and he started taking normal feeds. The child has been on maintenance daily dose of 2 mg. of cyproheptadine HCl and has been followed for about one year. He is now asymptomatic. Hyperpigmented lesions on the trunk, however, are persisting. Moderate mechanical pressure does not now produce wheal formation.

Discussion

The increase in Mast cells in skin is largely responsible for all these symptoms. What causes this increase is exactly not known. Mast cell differentiates from primitive mesenchyme which also forms reticulo-endothelial cells like histiocytes. Whether some mutation occurs during the embryological development which converts larger number of undifferentiated cells into mast cells or whether the mast cells themselves are rapidly proliferating is yet to be ascertained.

As the mast cell is known to produce histamine, heparin and certain other chemical substances in its meta chromatic granules, and the symptoms also result from the release of histamine and heparin, the treatment should remain more on conservative lines like cyproheptadine hydrochloride which has both antihistamine and antiserotonin properties rather than giving cytoxic drugs as suggested by some workers.

Patient had recurrent diarrhoea which was controlled successfully when treatment with cyproheptadine was instituted for urticaria pigmentosa. This indicates that this symptom too might have been due to the infiltration of larger number of mast cells in the gut which makes it irritable, oedematous and congested.

At present the child is completely asymptomatic on a maintenance dose of 2 mg. of cyproheptadine hydrochloride per day.

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FALSE: It has been shown that a diagnostic criterion for a disease, to be classified as mucopolysaccharidosis, one must isolate the AMPS in a purified state and subsequently quantitate the amount present and determine its nature. This is necessary because stains for AMPS even at pH 2.5, stain materials other than AMPS and histochemical quantitation of AMPS even at a crude level is notoriously misleading.