

A CASE OF WIDESPREAD URTICARIA PIGMENTOSA WITH EPISODES OF FLUSHING

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Urticaria pigmentosa was first described by Nettleship in 1869. Since then more than 500 cases have been reported, amongst which flushing was noted in only 19 cases—14 of widespread cutaneous urticaria pigmentosa and 5 of solitary mastocytoma. It is indeed peculiar that so few cases of vasomotor disturbances have been reported when one considers that urticaria pigmentosa is a mast-cell reticulosis¹ and that mast cells are known to liberate powerful vaso-active substances, particularly histamine and 5-hydroxytryptamine.

Marshall² has seen 2 further patients with solitary mastocytoma associated with flushing and also a woman with scattered pigmented lesions and vasomotor disturbances so severe that they frighten her children. The case of widespread urticaria pigmentosa with flushing described by Birt and Nickerson³ and the 2 cases reported by Neill⁴ are the most recent to be recorded.

The following case is recorded in view of the extreme rarity of the phenomenon of flushing with any of the mastocytoses:

CASE REPORT

A.W., a European boy aged 16 months, was apparently normal at birth. There was no family history of any skin disease.

When he was 2 weeks old blisters began to appear on his scalp; these dried and formed brown crusts, which fell off leaving light-brown marks. About 2 weeks later, brown spots and patches appeared in large numbers on the rest of the body, blisters only forming on those which were subjected to mild trauma like bumping or rubbing.

At the age of 2 months the first flush appeared, the whole skin except that on the lower legs being suffused with a uniform bright redness which, however, did not distress the baby in any way. Thereafter, flushing occurred at intervals of about 1 month until he was 13 months old, when there were no further episodes until 2 days before I saw him at the age of 16 months. This attack occurred on his waking early in the morning. The flushes lasted about three-quarters of an hour and then rapidly subsided. They seemed to arise quite spontaneously and did not follow trauma to the skin, or urticaria or blistering of the lesions, or the ingestion of food.

When the baby was 5 months old he lay on a cotton reel and a small raised nodule developed at the site of contact on his back. Blisters formed on the nodule from time to time but this did not coincide with the episodes of flushing. The first flush actually occurred 3 months before the nodule appeared on the back.

The normal skin between the pigmented lesions did not exhibit demographism.

The baby was otherwise very healthy and was never in any distress from pruritus during the flushing attacks or during the intervening periods, although he often rubbed one large brown macule in his groin whenever his napkin was removed. He never suffered from dyspnoea or gastro-intestinal upsets.

Examination. The patient was a fine healthy boy. His whole skin from scalp to feet was practically covered with well-defined, discrete, brown macules, irregular in shape and size varying in colour from pale beige on the exposed parts to dark sepia on the trunk. This difference in colour was very striking and looked as though exposure to light might be having some beneficial effect on the pigmentation. Only one raised nodular lesion was seen, the one described as following trauma to the back from a cotton reel. This nodule was oval, about 1 cm. long, and of a peculiar pale lilac colour which did not fade on diascopic pressure; the surface showed *peau d'orange*. Rubbing of this nodule, and indeed of several of the macules, produced immediate local erythema which was followed by whealing and the fairly rapid formation of a bulla, although no general flush was provoked by these measures. The normal intervening skin did not urticate on rubbing.

The urine was examined for 5-hydroxy-indole acetic acid and none was found; the specimen was not taken during a flushing episode. Unfortunately, urinary histamine estimations were not possible in the laboratory to which the urine was sent.

Histology. The nodule on the back was excised and histological sections showed numerous spindle-shaped cells in the corium and upper dermis. These cells contained granules which stained metachromatically with toluidine blue. The infiltrate extended right up to the epidermal basal layer. Disruption of the collagen was evident and the tissue infiltrate was associated with a fine reticular structure. There were no significant epidermal changes. The histological picture was consistent with that of mastocytoma.

I wish to thank Dr. W. Mukheiber for his cooperation and interest in this case.

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