

## URTICARIA PIGMENTOSA ASSOCIATED WITH ANETODERMA

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**Abstract.** A case of urticaria pigmentosa with associated anetoderma is reported. Although these two conditions may have been etiologically related, there was no evidence of anything other than a coincidental association.

To my knowledge, the association of anetoderma and urticaria pigmentosa has not been reported previously. A report of one such case follows.

### CASE REPORT

A 13-year-old Italian boy was examined first in July, 1969 for a generalized asymptomatic dermatosis which had been present since he was 8 months old. The process began around the neck and extended over the rest of his integument during the following 2 year period. Few new lesions had appeared since then. The parents believed that the pigmented, macular, papular, and nodular lesions preceded the scar-like lesions and that the latter lesions evolved from the former. However, they were not sure of this. The patient enjoyed excellent general health. One of his cousins had typical disseminated urticaria pigmentosa.

Physical examination disclosed a healthy-appearing adolescent boy. Many red-brown macules, papules, and nodules ranging in size from 0.3 cm to 2 cm were present over his trunk, neck, face, arms, and thighs. These lesions urticated when stroked; however, there was also milder urtication of the normal-appearing skin when it was stroked. Yellow-brown, thickened, nodular plaques were present in both axillae (Fig. 1), the ear canals, the angles of the ala nasae, the left inguinal crease, and perianally. Darier's sign was positive in these lesions too. Scattered over the chest, back, shoulders and neck were numerous, round, atrophic, scar-like lesions, most of which were macular (Fig. 2). Several were depressed beneath the surrounding skin surface level, and a few protruded into soft pseudotumors which were inverted easily upon palpation. Palpation of all the scar-like lesions disclosed the sharply marginated herniation through the dermis typically associated with anetodermatous lesions. There was equivocal, mild urtication of these lesions when they were stroked. There was no organo-

megaly, lymphadenopathy or other abnormalities in the remainder of the physical examination.

Skin biopsies were taken from 1) the thickened plaque in the right axilla, 2) an anetodermatous lesion on the back, and 3) an area of normal-appearing skin on the left buttock where dermatographism was present. Routine H & E stained slides on the tissue from the right axilla revealed myriads of deeply stained cells in strands and nests throughout the dermis. Giemsa (Fig. 3) and acridine orange fluorescent stains confirmed that these



*Fig. 1.* Large plaque and several small macular mastocytomas plus several anetodermatous lesions in the right axilla.

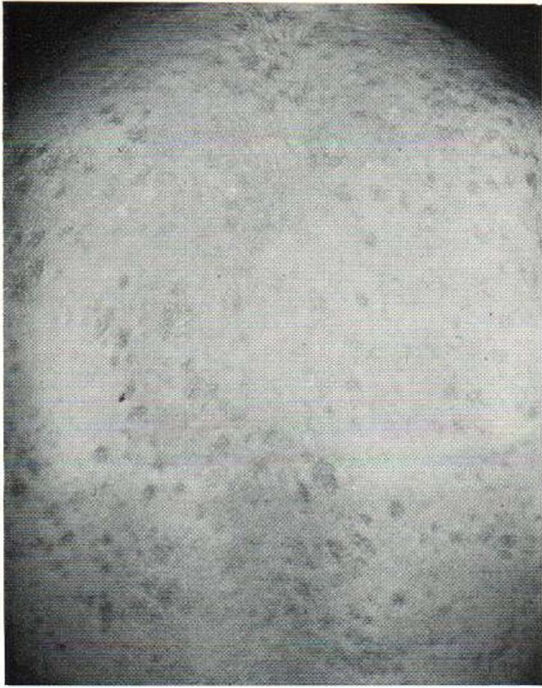


Fig. 2. Mastocytomas and anetodermas on the back.

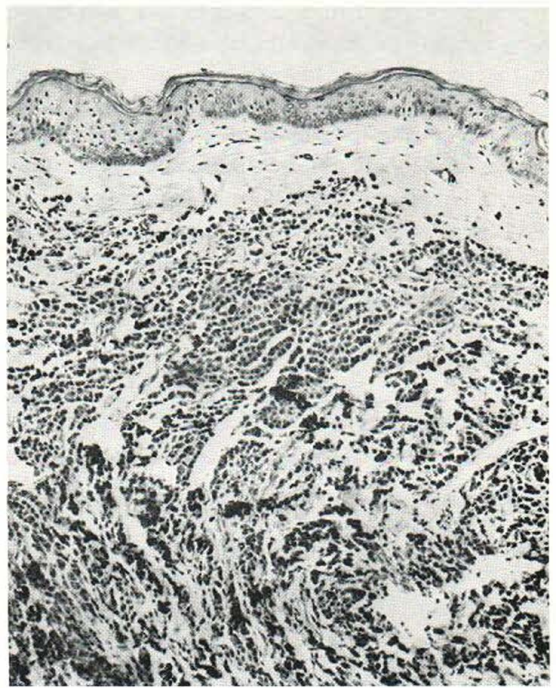


Fig. 3. Photomicrograph of tissue from axillary mastocytoma showing a dense infiltrate of mast cells in the dermis ( $\times 94$ ).

cells were all mastocytes. The biopsy tissue from the anetodermatous lesion showed some looseness in the dermal collagen and a few of the same cells seen in the first biopsy scattered around dermal blood vessels and lying in strands in the deep dermis (Fig. 4). Giemsa stain confirmed these cells' identity with the mastocytes seen in the tissue from the mastocytoma. Verhoeff elastic stain showed a virtually complete absence of elastic tissue in the dermis of this anetodermatous lesion. The biopsy tissue from the area of normal-appearing skin also revealed nests and strands of mast cells when stained with H & E and Giemsa stains.

Complete blood count, urinalysis, serum protein electrophoresis, and radiologic survey of the long bones were unremarkable.

#### COMMENT

I was unable to find any reports of urticaria pigmentosa and anetoderma in the same patient. In their book on mastocytosis, Sagher and Evan-Paz (2) state they have seen mastocytomas resembling skin tags and quote Nobe as reporting mastocytomas resembling soft fibromata which could "sometimes be indented with the fingers". Montgomery (1) states that in 1884 Pellizzari described a type of macular atrophy which started with urticarial lesions.



Fig. 4. Photomicrograph of tissue from an anetodermatous lesion showing loose dermal collagen and nests of mast cells in the dermis ( $\times 66$ ).

Although the urticaria pigmentosa may have been causally related to the anetodermatous areas in the present case, there is no evidence to suggest anything other than a coincidental association. Histologically, mast cells were present in increased numbers not only in the anetodermatous lesions but also in the clinically uninvolved skin.

If there were a causal relationship between the mastocytomas and the anetodermatous lesions, it would be difficult to understand the pathogenetic mechanisms involved. Neither heparin nor histamine—the two substances known to be present in human mast cell granules—are thought to cause any degenerative changes in dermal collagen or elastic tissue.

The thick, yellowish, xanthoma-like plaques of urticaria pigmentosa in the patient's intertriginous areas also are quite unusual and apparently are most commonly seen in the diffuse and erythrodermic forms of cutaneous mastocytosis rather than the more common maculopapular form. This patient's lesions had the clinical appearance of the latter form, but histologic evidence of diffuse mastocytic involvement with increased numbers of mast cells present in normal-appearing skin as well as in the clinically abnormal areas.

#### ACKNOWLEDGEMENT

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#### REFERENCES

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2. Sagher, F. & Evan-Paz, Z.: *Mastocytosis and the Mast Cell*, chap. 6. Year Book Medical Publ. Inc., Chicago, 1967.

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