Some cases of so-called “linear unilateral basal cell nevus with comedo-like lesions” have been described (2). This disorder differs from our reported case due to the unilateral arrangement of the lesions and lack of itching. Histopathological findings are essentially the same. We feel this type of nevoid basal cell lesion is the most closely related to the ours, from the structural and clinical point of view. There is also some affinity between our case and that described by Johnson and Hoockerman (5) as “basal cell hamartoma with follicular differentiation”, but in this latter case there were no cystic comedo-like structures.

In conclusion, we feel justified in considering our reported case as a new variant in the group of disorders called “follicular basal cell nevi”.

REFERENCES

Xanthogranuloma juvenile: A Case Report

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Abstract. A case of juvenile xanthogranuloma in a female newborn infant is reported. Red-brown, flat, well-demarcated lesions were present at birth on her scalp, face and trunk. During the following year of observation the eruptions regressed slowly and spontaneously. Light and electron microscopy revealed histiocytes and Tonton giant cells dominating the dermal cell infiltrate. The Touton cells contained bizzare invaginated nuclei, lysosomes, a granular endoplasmic reticulum, and cholesterol clefts in their cytoplasm. No fat droplets were seen.

Key words: Juvenile xanthogranuloma; Dermal cell infiltrate; Histiocytes; Touton giant cells; Cytoplasmic cholesterol clefts

Juvenile xanthogranuloma is a benign dermatosis of unknown origin, presenting soft, yellow or red-brown flat nodular lesions localized or disseminated over the skin. The disease usually starts in early infancy but is sometimes present at birth. The lesions tend to regress spontaneously during the first 3-4 years of life (1). Involvement of the eyes and mucous membranes has been reported (6), but is not a general trait of the disease.

CASE REPORT

A full-term female infant delivered by Caesarean operation presented red-brown, flat, papular lesions spread over her scalp, face and trunk (Fig. 1). No ocular symptoms, involvement of mucous membranes or internal organs could be demonstrated. No family members had suffered from a similar skin condition. During the following 12 months the skin lesions showed a slow but distinct regression, especially on the face. The involuted areas left a slight atrophic centre with residual annular thickening.

Fig. 1. The patient photographed one week after birth, showing flat, slightly thickened red-brown lesions of juvenile xanthogranuloma of the right lower part of the trunk and lower extremity.

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Pathology

Two-mm punch biopsies were removed from her trunk on the third postnatal day and prepared for light and electron microscopy by routine procedures.

Light microscopy

The dermis contained aggregated cell infiltrates consisting mainly of histiocytes and, among them, Touton giant cells and intermingling lymphocytes and eosinophilic granulocytes. No foam cells were seen. No structural changes were seen in the epidermis.

Electron microscopy

Infiltrating histiocytes contained markedly invaginated nuclei, a granular endoplasmic reticulum and lysosomes (Fig. 2). Touton-type giant cells showed identical cell organelles and nuclei, but a greater abundance of lysosomes and endoplasmic reticula. The nuclei were extremely invaginated, presenting bizarre shapes (Fig. 3).

No fat droplets were present in the cytoplasm, but cholesterol clefts were seen in the cytoplasm (Fig. 3). No Langerhans cell granules could be detected.

DISCUSSION

The finding of histiocytes and Touton-type giant cells indicates that the patient was suffering from juvenile xanthogranuloma, although no fat vacuoles could be demonstrated in the skin biopsy. The disease has been separated from histiocytosis X by the lack of Langerhans cell granules in the infiltrating cells (7). No such granules were found in the present case. In previous electron microscopical studies, fat droplets and lysosomes were reported to be a common finding in the infiltrating histiocytes and Touton cells. Esterly et al. (2) were unable to detect an enclosing bounding membrane around the fat droplets, whereas Kjaerheim et al. (3) and Wolff et al. (7) showed such a structure to be present. None described cholesterol clefts in the ground cytoplasm of the infiltrating cells, such as is reported here. The problem concerning the origin of fat droplets being either metabolic products or lysosomal material, i.e. either non-enclosed or enclosed by a bounding membrane, is still a matter of debate. Cholesterol clefts might indicate that the fat metabolism in the histiocyte has changed in some way. Numerous primary lysosomes may possibly develop into vacuoles of the cytoplasm.

Serial sectioning has demonstrated that multiple nuclei of Touton-type giant cells were the result of cutting the strongly invaginated nuclei (3). Fusion of cells due to the complexity of interdigitating protrusions of the cytoplasm has been suggested (4). In the present study no remarkable cytoplasmic protrusions could be found. Clinically, xanthogranuloma juvenilae may bear some resemblance to urticaria pigmentosa (5) or histiocytosis X (8). Since the disease regresses spontaneously, and malignant transformation is unlikely, no treatment is advisable.

REFERENCES


Leprosy in Vietnamese Refugees: A Case Report

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Abstract. A 41-year-old Vietnamese refugee was found to have tuberculoid leprosy. Dapsone treatment cleared her skin lesion, but did not remove the paraesthesia in...