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Bilateral Follicular Basal Cell Nevus with Comedo-like Lesions

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Abstract. A 12-year-old white boy presented with symmetrically appearing papular, nodular, comedo-like, highly itching lesions involving the skin on his ankles. They had first appeared 3 years earlier. Numerous cutaneous biopsies showed tumoral buds of basalioma cells associated with follicular structures, infundibular cysts and also hypertrophy and hyperplasia of sebaceous glands. To the best of our knowledge, there are no reported cases with the same clinical and histopathological findings as in

those reported here. We believe the lesions represent a "bilateral follicular basal cell nevus with comedones".

Key words: Bilateral follicular basal cell nevus; Linear unilateral basal cell nevus; Basal cell hamartoma with follicular differentiation; Comedo-like lesions; Infundibular follicular cysts

The term "follicular basal cell nevus" denotes a heterogeneous group of histopathological features which are difficult to classify nosographically. In fact they are both related to tumours of follicular origin (3, 6, 7) and to multiple basal cell epithelioma (1) and basal cell nevus syndrome (4).

The case we present seems to belong to the follicular basal cell nevus group, although it differs from the other previously reported cases due to some clinical and histopathological findings.

CLINICAL CASE

A 12-year-old white male came to our attention because of the presence of papular, nodular, crusted and comedo-like lesions localized on the internal and external aspects of the left ankle and the anterior aspect of the right ankle (Fig. 1). These lesions had appeared 3 years before.

The individual lesions consisted of numerous papular, nodular, pink-whitish, dome-shaped or flattened, solitary or bridged, slightly scaling elements, varying in size from 3 mm to 2 cm. Many of them were crusted; furthermore there was a large tumorous plaque on the right leg. Multiple clusters of comedones were present bilaterally. There were, moreover, some areas of slightly scaling "cigarette paper" atrophy.

Subjectively the patient felt intense itching on the affected areas.



Fig. 1. Clinical picture.

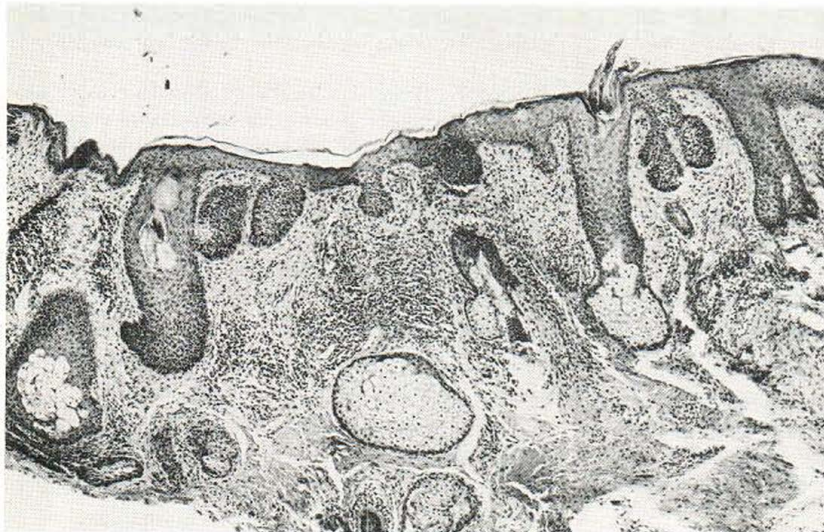


Fig. 2. Multiple buds of basalioma cells in connection with follicular structures (HE, 10 \times).

Neither parent had suffered from skin diseases. The boy's history was normal.

On examination he appeared normally developed, 169 cm in height, weighing 63 kg.

Physical and laboratory investigations failed to reveal any abnormalities. The ECG result was within normal limits and the X-rays of the pelvis were unremarkable.

Multiple biopsy specimens of the lesions were taken and stained with hematoxylin-eosin (HE), PAS and Feulgen. They showed similar histological features: numerous buds of proliferating basal cell epithelioma that projected downwards into the dermis, associated with follicular structures (Fig. 2). The strands of basal cell epithelioma consisted of cellular elements characterized by sparse cytoplasm, hyperchromic nuclei containing small grains of uniformly dispersed chromatin, a limited number of mito-

tic figures and a peripheral palisade arrangement. Moreover, microscopy revealed follicular plugs and follicular cystic cavities filled with keratin-like material (Fig. 3). We also noted a proliferation and hypertrophy of sebaceous glands, a moderately intense chronic inflammatory infiltrate of lymphocytes, histiocytes and plasma cells and a basophilic degeneration of collagen bundles of the dermis.

DISCUSSION

Basal cell nevi with follicular differentiation are a group of related disorders differing one from the other in certain details. The case we report here also presents some peculiar aspects.



Fig. 3. Infundibular cysts plugged by keratinous material (HE, 16 \times).

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 Hooekerman (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".

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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-demar-

cated 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 Touton giant cells dominating the dermal cell infiltrate. The Touton cells contained bizarre 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.

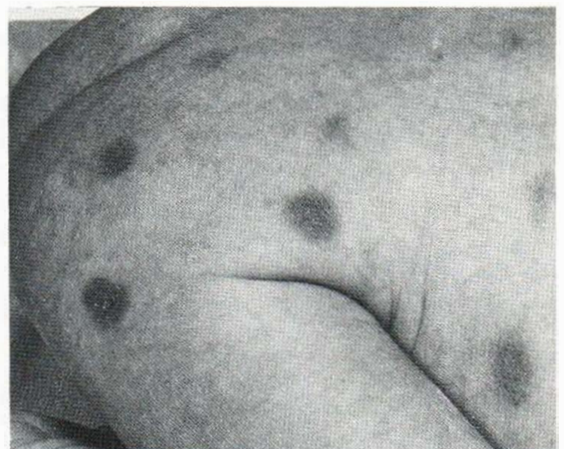


Fig. 1. The patient photographed one week after birth, showing flat, slightly thickened red-brown lesions of juvenile xanthogranuloma of the right lower part of the trunk and lower extremity.