Juvenile Xanthogranuloma: An Analysis of 45 Cases by Clinical Follow-up, Light- and Electron Microscopy

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Juvenile xanthogranuloma is a benign, nevoid histiocytosis not related to histiocytosis X. The lesions wrinkle, then flatten out and in 40% of the cases they leave no trace without any treatment. It is more frequent in boys. One patient also had granulomatous lesions of the eye, 8 had café-au-lait spots and 3 epilepsy. (Received July 17, 1984.)

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Juvenile xanthogranuloma (JXG) is a self-limited, benign disease of the newborn and infants, but cases in adults have also been observed (1). The name was given by Helwig & Hackney (2) in 1954, who collected 53 cases. The ancient term "nevoxanthoendothelioma" was suggested by McDonagh (3), who believed that Touton's giant-cells were of endothelial origin.

MATERIAL AND METHOD

45 children were followed up and analysed clinically, the regression, concomitant disease and eventual organ manifestations were observed. In 19 of the 45 cases biopsies were made for light- and electron microscopy.

RESULTS

In two-thirds of JXG cases the onset occurs immediately after birth to 6 months. Red, orange-coloured, later brown nodules are scattered axially on the body (scalp, face, neck, trunk) increasing in size and number for months, or even years. Old and new lesions exist at the same time.

Male patients predominate. They display solitary, multiple and generalized lesions (Table I). The nodules later regress to leave pigmentation, scar-like atrophy, or anetoderma-like areas while some disappear without any trace (Table II). Only skin manifestations were observed except in one patient who had a granulomatous infiltration in his eye. We found a relevant association between JXG, café-au-lait spots and epilepsy (Table I).

Light microscopy: the subepidermal granuloma was composed of closely packed histiocytes, foamy and multinucleated giant cells. Fibroblasts and lymphocytes were also

Table I. *Type of lesions and extracutaneous symptoms of 45 JXG patients*

n=45	Female 17	Male 28
Solitary	5	5
Multiple	8	19
Generalized	4	6
Organ involvement (eye)		1
Café-au-lait spots	2	6
Epilepsy	1	2
Other disease	1	3

Table II. *Regression of JXG lesions*

Residual lesions	Female (n=17)	Male (n=28)
No trace	2	16
Pigmentation	6	7
Atrophy	6	3
Total excision	3	2

present. Electron microscopy: foamy histiocytes containing dense lysosomes, lipid droplets and cholesterol crystals dominate the dermal infiltrate. The coalescing lipid droplets formed huge vacuoles; these had no limiting membrane but showed a dense peripheral ring also. In the pericytes of the dermal capillaries and in the smooth muscle cells of the arteries and of the arrectores pilorum there were lipid droplets.

DISCUSSION

We found organ infiltrations to be rare. There are, however, reports of consecutive glaucoma, blindness, involvement of muscles, periosteum, pericardium, pancreas, omentum, lymph nodes, lungs, liver, testicles and the CNS (4). The organ manifestations regress, too, and involvement of the iris is only of pathological interest.

Electron-microscopically not only the foamy cells are characteristic (5, 6) but also the lipid and cholesterol contained in dermal pericytes and smooth muscle cells. There is a remarkable connection between JXG and von Recklinghausen's disease (7, 8, 9): some patients have relatives with neurofibromatosis and 8 out of 45 of our patients had numerous café-au-lait spots. Surgical excision is not necessary, because the natural involution yields satisfactory final results.

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