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

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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...
**Short reports**

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

<table>
<thead>
<tr>
<th>Type of lesions</th>
<th>Female (n=17)</th>
<th>Male (n=28)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Solitary</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Multiple</td>
<td>8</td>
<td>19</td>
</tr>
<tr>
<td>Generalized</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>Organ involvement (eye)</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>Café-au-lait spots</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Other disease</td>
<td>1</td>
<td>3</td>
</tr>
</tbody>
</table>

Table II. *Regression of JXG lesions*

<table>
<thead>
<tr>
<th>Residual lesions</th>
<th>Female (n=17)</th>
<th>Male (n=28)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No trace</td>
<td>2</td>
<td>16</td>
</tr>
<tr>
<td>Pigmentation</td>
<td>6</td>
<td>7</td>
</tr>
<tr>
<td>Atrophy</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>Total excision</td>
<td>3</td>
<td>2</td>
</tr>
</tbody>
</table>

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, peristomeum, 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.

**REFERENCES**

Plasma 5-S-Cysteinyldopa Concentrations in Oculocutaneous Albinism

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5-S-cysteinyldopa concentrations were determined by high-pressure liquid chromatography and electrochemical detection in plasma from normally pigmented patients and patients with oculocutaneous albinism, both tyrosinase-positive and tyrosinase-negative. The plasma 5-S-cysteinyldopa concentrations were similar in all three groups, suggesting that 5-S-cysteinyldopa can be produced by mechanisms which do not involve tyrosinase.

Key words: Tyrosinase; Melanin.

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Human oculocutaneous albinism is of two main types: tyrosinase-positive and tyrosinase-negative albinism. These can be distinguished on the basis of both clinical features, and presence or absence of tyrosinase activity in the hairbulbs (1).

Tyrosinase is thought to be essential for the first two steps in melanin synthesis. These are the hydroxylation of tyrosine to form dihydroxyphenylalanine (dopa) and the subsequent oxidation of dopa to form the highly reactive intermediate dopaquinone. The pathway leading to formation of the red/yellow phaeomelansins involves the spontaneous coupling of dopaquinone with cysteine to form a number of different cysteinyldopas of which quantitatively the major component is 5-S-cysteinyldopa (5-S-CD) (2).

5-S-CD can be detected in the urine of normal healthy individuals, regardless of skin or hair colour (3). Patients lacking demonstrable tyrosinase activity would be expected to be incapable of synthesising melanin or its precursors, including 5-S-CD.

We report the results of a study in which we measured plasma 5-S-CD concentrations in both tyrosinase-positive and tyrosinase-negative patients with oculocutaneous albinism to see if it was possible to distinguish between the two groups on the basis of this measurement.

MATERIALS AND METHODS

There were twenty patients in the study, all with oculocutaneous albinism. They were divided into ten tyrosinase-positive and ten tyrosinase-negative albinos, on the basis of clinical assessment either on its own or, in a few cases, accompanied by a hairbulb tyrosinase test (4).

Controls were ten healthy volunteers of both sexes with a wide range of hair colour.