CONGENITAL XANTHOMA TUBEROSUM

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Giant-cell xanthomas have been described by various writers dealing with xanthoma tuberosum multiplex (1–5). Touton (1) distinguished xanthoma giant cells from other giant cells by the fact that in the former the nuclei are arranged in regular circles around a cloudy center, and are in turn enclosed by a broad border of protoplasm (Figs. 2 and 3). Giant cells, however, are most predominant in the solitary xanthomas occurring principally in tendons and tendon sheaths. In some of these cases xanthoma cells have been present in only small numbers or have even been entirely absent (Coenen, 6; Landois and Reid, 7; Unna, 8). Unna, describing a case of solitary giant-cell xanthoma, says: "There was not a trace of ordinary xanthomatous material, not even so much as may be found even in alcoholic preparations of ordinary xanthoma of the eyelid."

Solitary xanthoma in the newborn is extremely rare. Jorge and Brachetto-Brian (9) reported two cases involving the lower gum in day-old infants, and a third case, in the upper gum of an infant of six days. Corten believes that immature or not fully differentiated endothelial or connective-tissue cells are most likely to be changed into xanthoma cells. Chauffard (10) and Pollitzer and Wile (2) conclude that xanthoma tuberosum represents an irritation connective-tissue hyperplasia in which the extravasation of cholesterol fatty-acid
esters, present in excess in the blood, serves as a stimulant. Cases of xanthoma have been reported, in which the blood cholesterol was normal or even subnormal (Rosenthal and Braunisch, 11). Anitschkow (12) produced xanthoma cells in the connective tissue of rabbits by feeding them large amounts of cholesterol, and Weidman (13) obtained xanthoma in a dog by
similar measures. Gruenfeld and Seelig (14) describe the histologic structure of the solitary xanthomatous giant-cell tumor as resembling that of a sarcoma of the epulis type. Unna considers this tumor as constituting a distinct class.

The influence of trauma has been stressed by many authors. In a case reported by Chauffard (10) a xanthoma developed at the site of an injection of sodium cacodylate, while Ochs (15) described a case arising at the site of injections for congenital syphilis. Barker (16) concludes that the tumor affects by choice those areas which are predisposed to trauma. In some instances xanthoma tuberosum has been found to have a familial incidence. Wile and Duemling (17) collected fourteen such instances in the literature up to 1930.

CASE REPORT

J. L., a white, male infant, nine months old, was admitted to the Brooklyn Cancer Institute Sept. 5, 1936, for radiation therapy, following excision of a tumor in the right subclavicular region, diagnosed as "cancer."

Shortly after the child's birth the mother had noticed a swelling below the right clavicle, about 5 mm. in diameter, which she attributed to the trauma incident to forceps delivery. When, however, the other abrasions and contusions from this cause disappeared, the growth below the clavicle remained and became progressively larger. At six months it had a diameter of 2 cm. Subsequently the mother noticed that the tumor was "bleeding under the skin," and consulted a local physician, who excised the growth.

The family history reveals frequent pyogenic infections in both parents. The paternal grandfather died from cancer of the rectum, the paternal aunt from cancer of the uterus, the maternal grandfather from diabetes. There was no history of xanthoma in the family.

The child was well nourished and in good general condition. At the site of the original lesion, about 1 cm. below the right clavicle, was a healed scar 5 cm. long (Fig. 1). No adenopathy was demonstrable, and there was no local recurrence. A roentgenogram of the chest was negative. The blood chemistry of mother and child was as follows:

<table>
<thead>
<tr>
<th></th>
<th>mg. per 100 c.c. blood</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mother</td>
<td></td>
</tr>
<tr>
<td>Sugar</td>
<td>110.</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>202.02</td>
</tr>
<tr>
<td>Cholesterol ester</td>
<td>111.11</td>
</tr>
<tr>
<td>Free cholesterol</td>
<td>90.91</td>
</tr>
<tr>
<td>Child</td>
<td></td>
</tr>
<tr>
<td>Sugar</td>
<td>98.</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>238.1</td>
</tr>
<tr>
<td>Cholesterol ester</td>
<td>83.3</td>
</tr>
<tr>
<td>Free cholesterol</td>
<td>154.8</td>
</tr>
</tbody>
</table>

Histologic examination revealed a giant-cell xanthoma.

This case is reported because of its rarity and many unusual features, the age of the patient, the evidence of trauma, the absence of cholesterolemia, the predominance of giant cells, and the absence of xanthoma cells.

NOTE: The writer wishes to express his thanks to Dr. R. W. Gadbois for furnishing him with a specimen of the tumor.

REFERENCES