Solitary xanthoma of the limbus

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The purpose of this report is to present the findings in a case of solitary xanthoma of the limbus. Previous reports of ocular xanthomata have been associated with some other clinical or metabolic dysfunction, but in the case presented no other associated abnormalities could be found.

Case report

An 11-year-old Caucasian boy was seen at the Indiana University Medical Center, Department of Ophthalmology, because of a yellow limbal lesion of the left eye which had been first noted 2 years previously, and which had been progressively enlarging. The family history was not contributory. The child was premature, but developed normally with no history of significant systemic or ocular disease. There was no history of ocular trauma. The general physical examination was within normal limits. Specifically, no dermatological sign of xanthoma formation was found and hepato-splenomegaly was not present. The visual acuity was 20/20 in the right eye and 20/70 in the left.

The yellowish, raised mass measured 6×7 mm. in diameter at the medial limbus and encroached upon the pupillary area (Fig. 1). Several large trunk vessels penetrated the mass. Thin needle-like, multicoloured crystals were noted in the mid-stroma of the cornea bordering the advancing edge of the lesion.

FIG. 1 *Yellow raised limbal xanthoma*

Removal of the tumour was performed, and a conjunctival flap was pulled over the area of excision and sutured into place. There were no postoperative complications, and the patient showed no evidence of recurrence when seen 7 years postoperatively.

Laboratory investigations Urine analysis normal; cholesterol 172 mg. per cent. (normal 50 to 250); cholesterol esters 124 mg. per cent. (normal 100 to 150); neutral fat (triglycerides) 79 mg. per cent. (normal 150 to 180); blood urea nitrogen 18 mg. per cent.; uric acid (blood) 5.2 mg. per cent.; electrolytes normal.

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X rays of skull, chest, long bones, and pelvis normal.

Follow-up serum lipid studies, performed 7 years postoperatively, revealed: cholesterol 167 mg. per cent.; cholesterol esters 136 mg. per cent.; triglycerides 69 mg. per cent.

Pathological findings Microscopic examination (Figs 2 and 3) revealed dense infiltration of the corneal stroma by macrophages containing foamy cytoplasm. A few lymphocytes were scattered in each field. The pathological diagnosis was "xanthoma".

FIG. 2 Infiltration of cornea with lipid-laden macrophages. Haematoxylin and eosin. $\times 50$

FIG. 3 High-power view of corneal infiltration. Haematoxylin and eosin. $\times 430$

Comment

A xanthoma is a nodular mass formed by lipid-laden histiocytes and is usually a manifestation of a group of diseases which are collectively referred to as the xanthomatoses. Thannhauser (1958) has classified the xanthomatoses into the hypercholesterolaemic, hyperlipaemic, and normocholesterolaemic.

Hypercholesterolaemic There is an increase in the total serum cholesterol while the neutral fat remains normal or slightly elevated. This type of lipid abnormality is found in familial hypercholesterolaemia, hypothyroidism, and liver disease. The most common lesions associated with this type of lipid abnormality are tuberous xanthomata, plain xanthomata, and xanthelasma formation.

Hyperlipaemic These are associated with an increase in serum neutral fat, but the total lipids may be normal. Hyperlipaemia is found in a familial form, in severe untreated diabetes, chronic pancreatitis, glycogen storage disease, and lipid nephrosis. The clinical picture is that of secondary eruptive xanthoma formation.

Normocholesterolaemic These are found in patients with normal lipid values. Duke-Elder and Leigh (1965) mentioned that normocholesterolaemic xanthomatoses affected the outer eye in only two conditions—xanthoma disseminatum and Hand-Schüller-Christian disease. Xanthoma formation without apparent metabolic disturbance is also found in eosinophilic granuloma and Letterer-Siwe disease (Liebman, Crocker, and Geiser, 1966; Kara, 1957), in Neiman-Pick and Gaucher's disease (Albert and Smith, 1968), in juvenile xanthogranuloma (Sanders, 1962; Zimmerman, 1965), and in postinflammatory sclerosing lipogranuloma. The above mentioned examples of normocholesterolaemic xanthomatosis have typical findings on either physical or radiological examination and were excluded in our patient.

Summary

A unique case of solitary xanthoma of the limbus is presented. No association with trauma, inflammation, lipid abnormality, or systemic xanthomatosis could be found.

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