Lacrimal Gland Choristoma of the Iris

Michael Klüppel, MD; Wolfram Müller, MD; Rainer Sundmacher, MD, FRCOphth

Intraocular lacrimal gland tissue is an extremely rare choristoma. A newborn girl was seen with a fleshy, vascular tumor arising from the peripheral iris and the anterior chamber angle. The tumor was treated with topical steroids on suspicion of a juvenile xanthogranuloma; later, it grew slightly and a secondary glaucoma developed. Histopathological examination of the resected tumor showed lacrimal gland tissue in the iris. Twelve cases of intraocular lacrimal gland tissue have been reported in the literature.

**REPORT OF A CASE**

A 2-month-old girl was seen in our outpatient department with a yellow-red vascular tumor arising from the nasal iris and the anterior chamber angle (Figure 1). Examination revealed no other abnormalities of the eyes and orbits. Suspecting a somewhat atypical juvenile xanthogranuloma, we applied topical steroids. Examination findings for dermal xanthogranulomata were negative. Five weeks later, the right eye enlarged, with a difference of 1 mm in corneal diameter, due to a rise in intraocular pressure to 33 mm Hg. Steroid therapy was discontinued and dorzolamide eyedrops were applied 3 times a day. During this therapy the intraocular pressure decreased to normal levels. Ultrasound biomicroscopy under general anesthesia showed a very prominent tumor with 2 cystic areas (Figure 2). Considering the unknown identity of the tumor, we performed a careful anterior chamber aspiration; however, neither tumor cells nor other cells could be isolated by cytocentrifugation. Three weeks later we observed a tumor enlargement and a distortion of the pupil. We therefore decided to resect the tumor completely (Figure 3 and Figure 4). Findings on histopathological examination (Figure 5) revealed lacrimal serous gland tissue with regular epithelium and nuclei. Focally, the tubules were ectatic with a flattened epithelium. Postoperative care includes prophylactic patch occlusion of the sound eye and prescription of glasses because of a 2 diopter postoperative astigmatism.

Intraocular tumors should be treated in the most effective but least damaging way. In juvenile xanthogranuloma a conservative treatment with steroids is recommended, but it may take months or years until the tumor disappears and not until then is the diagnosis confirmed. Other tumors will not disappear without surgery and malignant tumors especially must be treated early.

In our patient the biomicroscopic characteristics of the 2 cysts were not typical of a juvenile xanthogranuloma. These cysts may be an important finding leading to the correct diagnosis. Surgery was performed because of tumor enlargement and the in-
creasing intraocular pressure. The latter may be caused by alterations of the anterior chamber angle or by a steroid response. Another reason, which in our opinion is the most probable, may be the secretion of the lacrimal gland. This excretes not only greater or lesser amounts of fluid, but also proteins and enzymes that are able to alter the trabecular network. This could cause progressive damage to important intraocular structures.

We recommend early resection to avoid this damage if such a tumor is of possible lacrimal gland origin and is not too large. The prognosis of our patient’s eye will depend particularly on the development of the intraocular pressure and on consistent prophylactic antiamblyopic treatment.

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Reprints: Michael Klüppel, MD, Department of Ophthalmology, Heinrich-Heine University, Moorenstrasse 3, 40225 Düsseldorf, Germany (e-mail: klueppel@uni-duesseldorf.de).

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