Hemangiopericytoma (HPCT) rarely originates in the lacrimal sac; 7 cases have been reported previously and only 1 contained an ultrastructural study. In this article we report an additional case and review the previous reports. While the initial biopsy specimen showed nonspecific cytologic abnormalities, light and electron microscopic studies on the subsequently excised tumor demonstrated that it had a structure characteristic of HPCT. The onset of lacrimal sac HPCT occurs in a younger age group than that of HPCT of other orbital locations. The tumor may recur locally but, to our knowledge, never has been reported to metastasize from a sac location. The treatment goal is complete surgical excision.

Masses in the lacrimal sac are found infrequently in ophthalmic practice. While most are inflammatory, a variety of neoplasms may occur, three fourths being of epithelial origin. Uncommon nonepithelial neoplasms include mesenchymal tumors (fibrous histiocytoma, hemangiopericytoma, and lipoma), lymphoid tumors, melanoma, granulocytic sarcoma, and neural tumors (neurofibroma and neurilemoma).1

REPORT OF A CASE

A 31-year-old man noted a slowly enlarging, painless lump in the right medial canthus with epiphora, which he had for 6 months. No bloody reflux or blood tinged tears had been observed. Findings from an ophthalmic examination were unremarkable except for a ballotable, non-tender lacrimal sac mass without exophthalmos. Lower punctal irrigation produced only a clear reflux through the upper punctum. Computed tomographic scan showed a well-circumscribed, nonhomogeneous tumor confined to a slightly enlarged nasolacrimal fossa and extending above the medial canthal tendon, without bony destruction (Figure 1).

Fine needle aspiration yielded only blood, without tumor cells. An open biopsy had been interpreted elsewhere as showing a “benign fibrohistiocytic tumor.” Histopathologic review showed tumor cells with irregularly shaped nuclei, small nucleoli, and bubbly cytoplasm alternating with areas of fibrosis (Figure 2). Immunohistochemical staining showed tumor cell positivity for vimentin and negativity for desmin, ulex, KP1, actin, α-antichymotrypsin, cytokeratin/AE-1, and S100.

A right dacryocystectomy was performed; the surgeon estimated total tumor removal. Healing was normal, and in the 3½ years that have elapsed since the removal, the tumor has not recurred.

PATHOLOGIC FINDINGS

Gross examination showed a gray ovoid mass measuring 12 × 12 × 10 mm. Histopathologic examination showed a hypocellular tumor surrounded by a fibrous pseudcapsule. The tumor consisted of innumerable ovoid-to-spindle–shaped nuclei, which—unlike the original biopsy specimen—were clearly arranged within a network of small blood vessels, frequently in a “staghorn” configuration (Figure 3). There were 1 to 2 mitotic figures per 40× high-power field. Periodic acid–Schiff and reticulin stains demonstrated abundant basal lamina enveloping vessels and individual cells within a rich vascular network. Immunoreagent...
CD34 stained only endothelial cells (Figure 4).

Electron microscopy showed collapsed thin-walled vessels surrounded by abundant basement membrane material and elongated cells with long cytoplasmic processes. Some cells showed surface basement membrane and, rarely, a cilium. Nuclei contained clumped chromatin and a prominent nucleolus (Figure 5 and Figure 6).
An uncommon tumor in the field of ophthalmic pathology, HPCT is usually encountered in the orbit,2 but may originate in conjunctiva,3,4 choroid,5 optic nerve,6 or medial canthal skin.7 Its occurrence in the lacrimal sac is rare (found in about 1% of sac tumors); only 7 cases have been reported1,8-13 (Table). While orbital HPCT usually appears at the median and mean age of approximately 42 years,2 the earlier onset of lacrimal sac HPCT (third to fifth decades of life)1,8-13 probably relates to its relatively noticeable, symptomatic periorcular location when small.

Classical HPCT microanatomy resembles that of the final excision in histopathologic terms. Occasionally, as in the initial biopsy specimen, diagnosis may be obscured by areas of myxoid, fibroid, or hyaline degenerative or inflammatory changes. Immunohistochemistry, which is invariably positive for vimentin, and electron microscopy may be valuable in such instances.

While most pathologists agree that a good case can be made for the existence of HPCT as a pathologic and morphologic entity, controversy derives from “hemangiopericytomatous” focal growth patterns present in such diverse entities as mesenchymal chondrosarcoma, malignant fibrous histiocytoma, solitary fibrous tumor, and malignant peripheral nerve sheath tumors.14 Such tumors can usually be separated from “true” HPCT by dissimilar subcellular and clinical features detected by specialized pathologic analyses. For example, HPCT reacts with vimentin, with or without CD34, but lacks other immunodeterminants of epithelial, neural, and myogenous differentiation (except for rare focal actin and desmin positivity).15

Ultrastructurally, HPCT cells resemble pericytes, having few cytoplasmic organelles. Subpopulations of HPCT cells may, however, show thin actin filaments with fusiform densities (indicating smooth muscle differentiation and correlating with positive actin stains). Continuous or interrupted basement membrane may envelop the cell.14

While orbital HPCTs may show a spectrum from benign to frankly malignant cytologic patterns, the subsequent biological behavior of the tumor cannot be accurately predicted from cytologic features alone. Me-
tastasis has been documented in some tumors that appeared cytologically benign. To our knowledge, no lacrimal sac HPCT has been reported to have metastasized to date.

Although optimal therapy for lacrimal sac HPCT is not established because of its rarity, treatment should follow advocated regimens for HPCT’s occurrence in other locations. Total local excision is recommended. As bony invasion is unusual in orbital locations, stripping the periorbit usually results in complete excision. Patients should be monitored for local recurrence, which has been documented in 3 of 8 lacrimal sac tumors. In cases of incomplete excision, local recurrence, or malignant cytologic features, ocular function should be assessed and additional surgery, radiation therapy, or chemotherapy should be considered. To date, however, no consensus exists on whether either of the latter 2 treatment modalities are effective.

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REFERENCES


A look at the past . . .

Within recent years much has been written by the pathologists about a type of tumor variously designated by the names “myoblastic myoma,” “myoblastoma,” “rhabdomyoma,” and “granular cell myoblastoma.” . . . The characteristic microscopic features may be summarized as follows: (1) large polyhedral cells, 20 to 60 microns in diameter, constitute a nodular accumulation of neoplastic cells; (2) the cytoplasm stains only faintly with eosin and contains many coarse neutrophilic granules; (3) cross or longitudinal striations are rarely seen. Granular cell myoblastoma is a relatively common tumor and widely distributed throughout the body. Since its occurrence in the orbit is rare, 2 cases are reported. The tumors in these cases presented no characteristic clinical picture, and the differential diagnosis was purely a histologic one. The tumor is more commonly benign, but in 1 of the present 2 cases it was malignant.