and D). A similar lesion was seen on the inferotemporal conjunctiva of the left eye. Funduscopic examination disclosed Bergmeister papilla and retinal vascular tortuosity bilaterally. The only facial cutaneous capillary malformations were on the lips and periorally. Lymphatic malformations had not been identified elsewhere in the body.

Following incisional biopsy of the lymphangiectasia in the right eye, the entire conjunctival surface flattened, presumably because the lymphatic channels were interconnected. Histopathological examination showed conjunctiva with focal epithelial thinning and numerous endothelium-lined, dilated channels filled with serous material in the substantia propria (Figure 2A). Capillaries were seen intermixed with the dilated lymphatic channels. Immunohistochemical examination revealed positive staining for CD34 in the endothelial cells of the blood vessels and negative staining for CD34 in the endothelial lining of the dilated, blood-free channels, consistent with lymphatic vessels. Immunofluorescence with the lymphatic marker D2-40 was positive in the dilated channels (Figure 2B).

Six weeks postoperatively, the lesion again was elevated in all areas except for the site from which the biopsy specimen was taken.

Comment. Lymphangiectasia of the conjunctiva is a localized or diffuse enlargement of the lymphatics that appears as chemosis or a freely movable cyst or series of cysts (“string of pearls”). The localized form is often seen secondary to conjunctival pathological findings such as pinguecula or scar. The diffuse form is less common and often there is no attributable cause, but it has been reported in a patient with Turner syndrome and hereditary lymphedema (Milroy disease). When a connection exists between a blood vessel and the dilated lymphatics, intermittent filling with blood occurs and the lesion is known as lymphangiectasia hemorrhagica conjunctivae.

Lymphatic abnormalities are among the congenital vascular malformations seen in KTWS and are predominantly located in the extremities and adjacent parts of the trunk (shoulder, pelvis). Other reported sites include the genitourinary system, lower gastrointestinal tract, liver, lung, and spine, with facial involvement being exceedingly rare. The bilateral conjunctival lymphangiectasia in this patient represents another manifestation of this poorly understood syndrome. Although lymphangiectasia is benign in nature, excision for cosmesis or diagnosis may be required.

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Soft-Tissue Perineurioma of the Bulbar Conjunctiva

Perineuriomas are rare peripheral nerve sheath tumors composed exclusively of neoplastic perineurial cells and showing distinctive morphologic, ultrastructural, and immunophenotypic features that distinguish them from other nerve sheath tumors. Perineuriomas can be broadly divided into 2 histological categories: an intraneu-
ral group and a more common extraneural or soft-tissue group. Most of these neoplasms are benign, but perineuriomas of low-grade malignant potential and malignant form have also been reported. Soft-tissue perineurioma clinically manifests as a painless nodule, occurs mostly in superficial soft tissue, and only infrequently affects deep soft tissue of the extremities or trunk. Rare examples arising at visceral locations have been reported. There are no reported cases in the English-language literature of perineuriomas affecting the conjunctiva.

**Report of a Case.** A 47-year-old man had a painless, slowly growing mass in the right bulbar conjunctiva since 3 months before admission. He underwent surgical removal of a 1.0-cm nodule. Grossly, the specimen consisted of tissue fragments with aggregated dimensions of $1.0 \times 0.8 \times 0.3$ cm that were brownish and firm. Microscopical examination revealed a nonencapsulated subepithelial spindle cell proliferation composed of elongated cells with wavy nuclei and long, slender, eosinophilic cytoplasmic processes. The tumor cells showed an interweaving fascicular growth pattern and focal storiform arrangement, with hypercellular areas and mild cytologic atypia. There were 7 mitotic figures per 10 high-power fields and no necrosis. (Figure 1). Immunohistochemical studies showed reactivity for epithelial membrane antigen (EMA) (titer, 1:150 [Dako Corp, Glostrup, Denmark]; antigen retrieval titer, 1:20 [Trilogy; Cell Marque Corp, Rocklin, California] for 40 minutes with heating at 95°C), glucose transporter protein 1 (GLUT-1) (titer, 1:200 [Cell Marque Corp]), and protein gene product 9.5 (titer, 1:100 [Neo-Markers Inc, Fremont, California]); showed reactivity focally for claudin-1 (prediluted [Dako Corp]) and collagen IV (titer, 1:100 [Biogenex, Andhra Pradesh, India]); showed negative results for AE1/3 (titer, 1:50 [Cell Marque Corp]), actin (titer, 1:30 [Cell Marque Corp], CD34 (titer, 1:100 [Dako Corp]), and S-100 protein (titer, 1:4000 [Bio SB, Inc, Santa Barbara, California]). Ki-67 (titer, 1:25 [Cell Marque Corp]) labeled approximately 10% of the neoplastic cells (Figure 2). Based on the morpho-

**Figure 1.** Histological section stained with hematoxylin-eosin. A, Squamous epithelium with subepithelial spindle cell proliferation with an interweaving fascicular pattern (original magnification $\times 40$). B, Hypercellular areas and mild cellular atypia (original magnification $\times 200$). C, The nuclei of neoplastic cells have an elongated, spindle-shaped, wavy profile with long, slender, bipolar, eosinophilic cytoplasmic processes (original magnification $\times 400$). D, Mitotic figure (arrow) (original magnification $\times 400$).
logic and immunohistochemical findings, the diagnosis of a perineuroma with atypical histological features in the bulbar conjunctiva was made. After the resection, the patient was lost to follow-up.

Comment. Although perineurial cell proliferation may be suspected histologically with routine hematoxylin-eosin staining, a definite identification needs the demonstration of perineurial cell features using immunohistochemical studies. The morphologic criteria include spindle cells with curved or wavy thin nuclei and thin, elongated cytoplasmic processes, arranged in lamellae, and a storiform growth pattern forming loose whorls and bundles. By immunohistochemistry, perineurial cells stain positive for EMA, GLUT-1, and claudin-1; may stain positive for protein gene product 9.5 and CD34; and stain negative for S-100 protein, CD57, and neurofilaments. According to Hornick and Fletcher, cases with atypical and malignant examples have been reported.1,3,4

Perineuromas are benign soft-tissue neoplasms, but atypical and malignant examples have been reported.1,3,4 According to Hornick and Fletcher, cases with atypical pleomorphic cytologic results that show abrupt transition from typical cytomorphicologic findings and storiform architecture to markedly hypercellular areas with a fascicular growth pattern and diffuse infiltration of adjacent skeletal muscle have been classified as atypical perineuromas. Hornick and Fletcher also state that the mitotic count is variable. These histological features have no clinical significance and may be comparable to the degenerative changes observed in ancient schwannoma. In benign and atypical forms of perineuroma, surgical resection with free margins is curative; only 5% of cases have shown recurrence, and there are no reported cases with metastases.1,4,5

The differential diagnosis of soft-tissue perineuroma includes cellular schwannoma (S-100 protein +; CD34 +/−; EMA −), low-grade fibromyxoid sarcoma of Evans (S-100 protein −; EMA +/−; CD34 +), solitary fibrous tumor (CD3 +; S-100 protein −; EMA −), and benign fibrous histiocytoma or low-grade malignant fibrous histiocytoma (CD3 −; S-100 protein −; EMA −; CD68 +). Immunohistochemical studies will help separate these 4 entities.1,4,6

To our knowledge, this is the first case of atypical perineuroma arising in the bulbar conjunctiva reported in the English-language literature.

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EVIDENCE THAT ANTERIOR EPISCLERAL NERVE SHEATH TUMORS ARISE FROM THE AXENFELD NERVE LOOP

T he occurrence of solitary episcleral neurofibroma has been described previously in 3 case reports1-4; episcleral schwannomas have been reported 4 times.5-8 The origin of these rare tumors is unknown. However, topographic analysis from our case and those in the literature provides evidence that most anterior scleral nerve sheath tumors arise from intrascleral nerve loops.

Report of Cases. A 45-year-old woman had a mildly tender, white nodule of a few weeks’ duration that was adherent to the sclera, measuring 3.5 × 3.5 mm in surface dimension and located approximately 4 mm from the limbus in the inferotemporal quadrant. Neither prior eye operations nor traumatic injury occurred in this region. Microscopic examination of histologic sections revealed a spindle cell tumor with features of a nerve sheath tumor (Figure). The tumor did appear encapsulated.

Cases culled from the literature of episcleral nerve sheath tumors are summarized in the Table. Under the assumption that benign nerve sheath tumors enlarge in a symmetric fashion, the distance of the tumor from the limbus (d) was calculated from the photograph by the following equation: d = (11.7 × r)/c, where c indicates the mean corneal diameter in millimeters measured in the photograph and 11.7 represents the corneal diameter in millimeters.9

The key characteristics of episcleral peripheral nerve sheath tumors are shown in the Table. The mean age of