Perilimbal Conjunctival Schwannoma

Schwannomas are primary neurilemmal tumors composed of proliferating Schwann cells. Schwannomas of ophthalmic interest more frequently involve the orbit than the globe, but uveal schwannomas have been reported. Rare among ocular schwannomas is the conjunctival schwannoma. Seven have been described in the English-language literature, only 3 of which were located on the bulbar conjunctiva (Table).

Report of a Case. A 68-year-old white man was referred for excision of a pingueculum at the 3-o’clock position of the left eye. The lesion of interest was 2.2 × 2 mm, well circumscribed, mobile, yellow, and perilimbal (Figure 1). A small pingueculum was also noted at the 9-o’clock position. The remainder of the examination results were unremarkable.

Surgical excision of the lesion at the 3-o’clock position was performed largely to rule out squamous neoplasia. The lesion was easily excised without evidence of scleral extension.

Histologic sections and staining showed a well-circumscribed tumor composed of bland spindle cells with foci suggestive of nuclear palisading. There were regions suggestive of Antoni A and B patterns. There was no connection with the overlying mucosa. There was no atypia, necrosis, or mitotic activity (Figure 2A). Immunohistochemical studies demonstrated immunoreactivity of the tumor cells for S100 protein (Figure 2B) and no reactivity for HMB-45. Reticulin stain showed delicate fibers within the tumor.

Comment. Schwannomas are slow-growing, encapsulated, peripheral nerve sheath tumors that may be found in isolation or in association with von Recklinghausen neurofibromatosis. Overall, schwannomas typically appear in the third to fifth decade of life and demonstrate no sex predilection. Of the 7 previously reported conjunctival schwannomas, however, 6 arose in women with ages ranging from 12 to 72 years (Table).

Schwannoma is not frequently included in the differential diagnosis of nonpigmented conjunctival masses, which consists of pingueculum, nevus, foreign body, neurofibroma, leiomyoma, fibrous histiocytoma, dermoid, squamous and sebaceous cell carcinomas, and amelanotic malignant melanoma. Schwannomas are composed of a pure proliferation of Schwann cells. Two distinct, intermingling histologic patterns are seen: an Antoni A pattern of sheets of palisading spindle cells with spindle-shaped nuclei forming Verocay bodies, and an Antoni B pattern of haphazardly arranged elongated cells in a myxoid stroma. Light-microscopic features of schwannoma are usually characteristic, but immunohistochemistry may be required in some cases.

The tumor presented herein was a bland, well-circumscribed spindle cell tumor with immunohistochemical profile favoring the diagnosis of schwannoma. Antibodies directed against the S100 protein identify melanocytes and neural crest–derived cells, thereby excluding fibrohistiocytic and smooth-muscle actin.

<table>
<thead>
<tr>
<th>Source, y</th>
<th>Patient Age, y/Sex</th>
<th>Location</th>
<th>Size, mm</th>
<th>Histologic Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dabezies and Penner,2 1961</td>
<td>50/F</td>
<td>Upper temporal bulbar conjunctiva (recurrent)</td>
<td>Not described</td>
<td>Nests and whorls; reticulin positive; sharp delineation; palisading of nuclei</td>
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<tr>
<td>Vincent and Cleasby,3 1968</td>
<td>12/F</td>
<td>Lateral to caruncle</td>
<td>10 × 5 × 5</td>
<td>Cells with basophilic nuclei, surrounded by fibrillar matrix; streaming palisades</td>
</tr>
<tr>
<td>Grossniklaus et al,4 1987</td>
<td>61/F</td>
<td>Not described</td>
<td>Not described</td>
<td>Bundles and cords of spindle cells surrounded by fibrous tissue; Antoni A and Antoni B pattern</td>
</tr>
<tr>
<td>LeMarchaudour et al,5 1996</td>
<td>37/M</td>
<td>Nasal bulbar conjunctiva</td>
<td>5 × 4 × 4</td>
<td>Uninodular mass; spindle cells arranged in bundles and fascicles with Antoni type A pattern; S100 positive; HMB-45 negative</td>
</tr>
<tr>
<td>Charles et al,6 1997</td>
<td>19/F</td>
<td>Inferior fornix</td>
<td>15 × 10 × 10</td>
<td>Solid and encapsulated; Antoni A pattern; S100 positive</td>
</tr>
<tr>
<td></td>
<td>26/F</td>
<td>Subconjunctival at 6 o’clock</td>
<td>2 × 2 × 2</td>
<td>Spindle cells in palisades; S100 positive</td>
</tr>
<tr>
<td></td>
<td>72/F</td>
<td>Superior tarsal conjunctiva</td>
<td>10 × 10 × 10</td>
<td>Bland spindle cells in fascicles as well as Antoni B; S100 positive</td>
</tr>
<tr>
<td>Present report</td>
<td>68/M</td>
<td>Perilimbal</td>
<td>2.2 × 2</td>
<td>Nuclear palisading; S100 positive; HMB-45 negative</td>
</tr>
</tbody>
</table>
muscle tumors. HMB-45 antibodies identify cells of melanocytic origin and are a sensitive marker for melanoma and nevi. In addition, the reticulin stain showed abundant fibers that are prominent in most schwannomas.

Fibrous histiocytoma and leiomyoma are often confused with schwannomas because of their spindle cell morphologic characteristics, but can be distinguished by their lack of S100 expression. Neurofibromas are S100-positive Schwann cell tumors, but include fibroblasts and perineural cells, and nuclear palisading is less common. The circumscription, bland cytologic features, and absence of atypia, necrosis, or mitotic activity essentially rule out a malignant tumor.

In summary, we report a rare case of perilimbal conjunctival schwannoma. We suggest this diagnosis be included in the differential diagnosis of amelanotic conjunctival lesions.

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