Eyelid schwannomas with associated neoplasms: A report of 2 cases

Zoe Brown-Joel, MD, a Neda Esmaili, MD, b Sang Hong, MD, b Kara Young, MD, a and Karolyn Wanat, MD a

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INTRODUCTION

Schwannomas are neural tumors that typically present on the extremities, head, and neck, and can be seen in association with neurofibromatosis type 2. We present 2 cases of cutaneous schwannomas of the eyelid due to their unusual location and association with additional neoplasms in a 71-year-old patient and in a 35-year-old patient.

Schwannomas presenting on eyelids are unusual tumors. In the literature, there are 22 reported cases. These cases are unique not only in their location but also their association with other neoplasms, which we hypothesize, may be related to a local increase in growth factors. Presence of these tumors on the eyelid is an unusual and rarely reported finding.

CASE REPORT

Case 1

A 71-year-old female presented for evaluation of a slowly growing, right lower eyelid lesion of at least 5 years duration. She was referred to oculoplastics for further evaluation. Clinically, the area was described as an 11 × 4 mm pink to skin-colored papule on the lower eyelid margin with madarosis, central ulceration, and a raised telangiectatic border (Fig 1, A). Upon incisional biopsy, a separate firm, well-circumscribed mass was discovered immediately underneath the eyelid margin papule. Histopathology displayed a circumscribed spindled mass composed primarily of Antoni A tissue, consistent with a schwannoma, as well as an adjacent infiltrative basal cell carcinoma (Fig 1, B-D). The patient then underwent Mohs resection as a definitive treatment for the basal cell carcinoma.

Case 2

A 35-year-old female presented for evaluation of an asymptomatic left lower eyelid lesion of 3 years duration with gradual growth. Clinically, the area was described as a 4.3 mm cystic papule on the left inferior central lid margin with slight eyelash displacement (Fig 2, A). The lesion was excised by oculoplastics and displayed a similar Antoni A predominant schwannoma with an overlying intradermal nevus (Fig 2, B-D). Mart-1 staining was localized to the overlying nevus and SOX-10 was positive in both areas (Fig 2, E and F).

DISCUSSION

Schwannomas of the eyelid are rarely reported. Within the literature, most cases have been reported in adults; however, the entity has been reported in children, the youngest of which was 8 years old.1 Due to its non-specific clinical appearance, the clinical differential is broad and includes entities such as an epidermal inclusion cyst, amelanotic nevus, chalazion, hidrocystoma, basal cell carcinoma, and molluscum contagiosum.2,3 Eyelid schwannomas are theorized to arise from the supraorbital, supratrochlear, and infraorbital nerves.4,5 Clinically, they have been reported to ulcerate, however, in most cases the lesions are asymptomatic.6 Malignant transformation of eyelid schwannomas has not been reported, however, complete removal is recommended for diagnosis and to prevent recurrence.6
**Fig 1. A-D.** (from left to right). **A,** clinical image: small papule on the lower eyelid. **B,** superficial portion of the excision with angulated basaloid islands extending from the epidermis, consistent with BCC, 100×. **C,** deeper portion of the histologic specimen with well-circumscribed spindled neoplasm, 40×. **D,** spindled neoplasm at 100× composed of cellular, palisaded type Antoni A tissue.

**Fig 2. A-F.** (from left to right). **A,** clinical photograph: lower eyelid papule with eyelash displacement. **B,** superficial portion of excision showing a dermal proliferation of well-nested melanocytes, 40×. **C,** deeper portion with well-circumscribed spindled neoplasm, 40×. **D,** higher magnification demonstrating with Antoni A changes, 100×. **E,** Sox10 stain highlighting both the nevus and underlying schwannoma, 100× (nevus not pictured). **F,** Mart-1 stain highlights only overlying nevus, 200×.
Histologically, schwannomas stain strongly positive for S100, with accentuation of S100 within Antoni A portions. They typically occur as single lesions, although multiple schwannomas can develop in the setting of neurofibromatosis type 2. The presence of additional neoplasms in proximity to the schwannomas, as described in these cases, is a curious finding. Helbing et al proposed that schwannomas may preferentially develop in areas prone to neural injury, as a failure of the normal nerve regeneration process, akin to a chronic neural wound. Changes in cell signaling within the tumor microenvironment such as inflammatory signals from macrophages and vascular growth signals from endothelial cells may contribute to schwannoma development. Interestingly, both neoplasms in case 2 share neural crest derivation. While for both cases it may be coincidental, the presence of adjacent neoplasms does raise the question of developmental timing, as perhaps these neoplasms altered the growth environment, leading to development of the eyelid schwannomas.

Conflicts of interest
None disclosed.

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