

# Denouement and Discussion

## Eyelid Pilomatricoma

**B**oth lesions underwent surgical excision (**Figure 2**), and histopathologic examination revealed well-demarcated tumors with distinct regions of darkly stained basophilic cells and pale-staining anuclear cells in both cases. There were also focal calcium deposits within the tumors.

The images depict pilomatricomas involving the eyelid. Pilomatricomas, also referred to as pilomatrixomas and calcifying epitheliomas of Malherbe, represent benign tumors of hair follicle origin. They were first described in 1880 by Malherbe and Chenantais.<sup>1</sup> Although they can occur in any age group, they are most common in children and adolescents.<sup>2</sup> They typically appear as solitary, firm, painless dermal papules or nodules on the head and neck. Pilomatricomas may have a bluish coloration or may have central ulceration revealing underlying calcification (**Figure 3**). When palpated, they often exhibit the so-called teeter-totter sign, as pressure on one edge of the lesion causes elevation of the opposite side. When excised, these lesions are often larger than they appear on the surface.

Pilomatricomas uncommonly manifest on the eyelid areas, and when they do, they are often mistaken clinically for chalazions, granuloma annulare, epithelial cysts or dermoid cysts when they are bluish in color,<sup>3</sup> or kera-

toacanthomas or other malignant tumors if they have central ulceration.<sup>4</sup>

The appearance of multiple pilomatricomas has been occasionally associated with myotonic dystrophy,<sup>5</sup> Gardner syndrome,<sup>6</sup> Steiner disease,<sup>7</sup> Turner syndrome,<sup>8</sup> and sarcoidosis.<sup>9</sup> At least 75% of human pilomatricomas possess activating mutations in  $\beta$ -catenin, suggesting that  $\beta$ -catenin plays a role in the tumorigenesis of pilomatricomas.<sup>10</sup>  $\beta$ -Catenin is a member of the Wnt signaling pathway and plays an important role in the morphogenesis of hair follicles.<sup>11</sup> Although typically benign, malignant pilomatricomas have rarely been reported.<sup>12</sup> Treatment consists of surgical excision, and recurrence is rare after complete resection.

Accepted for Publication: May 6, 2009.

Correspondence: Albert C. Yan, MD, Section of Dermatology, Children's Hospital of Philadelphia, 3550 Market St, 2nd Floor, Philadelphia, PA 19104 (yana@email.chop.edu).

Author Contributions: Study concept and design: Castelo-Soccio, J. A. Katowitz, and Yan. Acquisition of data: Castelo-Soccio, W. R. Katowitz, Shah, and Yan. Analysis and interpretation of data: Castelo-Soccio, W. R. Katowitz, Shah, Treat, and Yan. Drafting of the manuscript: Castelo-Soccio and Yan. Critical revision of the manuscript for important intellectual content: W. R. Katowitz, J. A. Katowitz, Shah, Treat, and Yan. Administrative, technical, and material support: Castelo-Soccio, W. R. Katowitz, J. A. Katowitz, Shah, Treat, and Yan. Study supervision: Yan. Financial Disclosure: None reported.



**Figure 2.** Patient 1 (A) and patient 2 (B) following oculoplastic excision and repair.



**Figure 3.** Calcified pilomatricoma.

## REFERENCES

1. Malherbe A, Chenantais J. Note sur l'épithéliome calcifié des glandes sébacées. *Prog Med (Paris)*. 1880;8:826-837.
2. Yench MW. Head and neck pilomatricoma in the pediatric age group. *Int J Pediatr Otorhinolaryngol*. 2001;57(2):123-128.
3. Levy J, Ilisar M, Deckel Y, Maly A, Anteby I, Pe'er J. Eyelid pilomatrixoma. *Surv Ophthalmol*. 2008;53(5):526-535.
4. Lan MY, Lan MC, Ho CY, Li WY, Lin CZ. Pilomatricoma of the head and neck. *Arch Otolaryngol Head Neck Surg*. 2003;129(12):1327-1330.
5. Hino S, Kondo S, Sekiya H, et al. Molecular mechanisms responsible for aberrant splicing of SERCA1 in myotonic dystrophy type 1. *Hum Mol Genet*. 2007;16(23):2834-2843.
6. Pujol RM, Casanova JM, Egido R, Pujol J, de Moragas JM. Multiple familial pilomatricomas: a cutaneous marker for Gardner syndrome? *Pediatr Dermatol*. 1995;12(4):331-335.
7. Barberio E, Nino M, Dente V, Delfino M. Guess what! multiple pilomatricomas and Steiner disease. *Eur J Dermatol*. 2002;12(3):293-294.
8. Wood S, Nguyen D, Hutton K, Dickson W. Pilomatricomas in Turner syndrome. *Pediatr Dermatol*. 2008;25(4):449-451.
9. Julian CG, Bowers PW. A clinical review of 209 pilomatricomas. *J Am Acad Dermatol*. 1998;39(2, pt 1):191-195.
10. Park SW, Suh KS, Wang HY, Kim ST, Sung HS. Beta-catenin expression in the transitional zone of pilomatricoma. *Br J Dermatol*. 2001;145(4):624-629.
11. Moreno-Bueno G, Gamallo C, Pérez-Gallego L, Contrero F, Palacios J. Beta-catenin expression in pilomatrixomas. *Br J Dermatol*. 2001;145(4):576-581.
12. Bassarova A, Nesland JM, Sedloev T, Danielsen H, Christova S. Pilomatrix carcinoma with lymph node metastases. *J Cutan Pathol*. 2004;31(4):330-335.