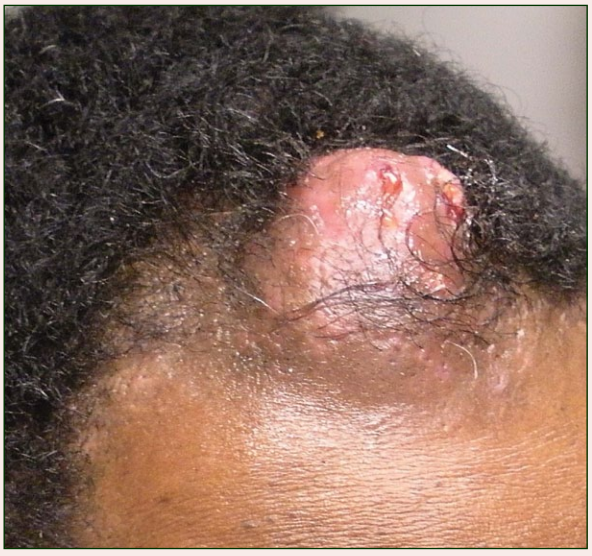
Solitary Nodular Lesion on the Scalp

What's the diagnosis?



An otherwise healthy 40-year-old man presented for examination of a solitary nodular lesion on the frontal aspect of the scalp of 1 year's duration. The lesion had rapidly increased in size in the 2 weeks prior to presentation. He presented to the emergency department after he noted pain and drainage from the lesion. Biopsy of the lesion revealed islands of pale eosinophilic shadow cells with an intense dermal infiltrate consisting of lymphocytes, histiocytes, plasma cells, and neutrophils.

Sotonye Imadojemu, MD, MBE; Douglas J. Pugliese, MD, MPH; Carrie L. Kovarik, MD

From the Perelman School of Medicine, University of Pennsylvania, Philadelphia. Dr. Pugliese is from the Department of Dermatology, and Drs. Imadojemu and Kovarik are from the Departments of Dermatology and Medicine.
The authors report no conflict of interest.
Correspondence: Carrie L. Kovarik, MD, University of Pennsylvania, Department of Dermatology, 2 Maloney Bldg, 3600 Spruce St, Philadelphia, PA 19104 (carrie.kovarik@uphs.upenn.edu).

The Diagnosis: Pilomatricoma

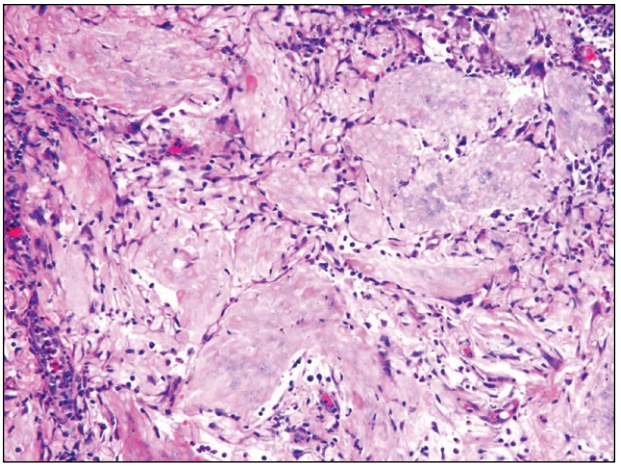
Pilomatricoma, first described by Malherbe and Chenantais[1] in 1880, is a benign appendageal tumor derived from hair follicle matrix cells. It classically manifests as a solitary, asymptomatic, firm dermal nodule with a normal overlying epidermis. Less common morphologic variants include perforating, lymphagiectatic, keratoacanthomalike, pigmented, and anetodermalike surface changes.[2] Inflammation and erosion through the skin surface are observed in the rare perforating variant, as seen in our patient. The average size is 1 cm, and it rarely exceeds 3 cm in diameter.[3] The tumors predominantly occur on the head, neck, and upper extremities, with only 9.5% on the scalp.[2] It may occur at any age, though it has a bimodal distribution with peaks in childhood and in adults older than 60 years. A slight preponderance in females has been observed with a female to male ratio of 1.5 to 1.[2] Although our patient is black, most reported cases have occurred in individuals of European descent. Because cases of pilomatricoma are not systematically reported, it is uncertain if this finding represents a publication bias or if race is an actual risk factor. Multiple pilomatricomas and familial cases have been described in association with myotonic dystrophy, Turner syndrome, Gardner syndrome, Rubinstein-Taybi syndrome, polyfactorial coagulopathy, trisomy 9, xeroderma pigmentosum, and basal cell nevus syndrome.[2,4]

It has been shown that the proliferating cells of pilomatricomas stain with antibodies directed against Lef1 (lymphoid enhancer binding factor 1), a marker from hair matrix cells, providing biochemical evidence for the morphologic appearance of these neoplasms.[5] Pilomatricomas have been associated with B-cell/chronic lymphocytic leukemia lymphoma 2 gene, *BCL2*, expression, a proto-oncogene that suppresses apoptosis in benign and malignant neoplasms, which may contribute to the pathogenesis of these tumors.[6] Pilomatricomas also have been associated with β-catenin mutation, expression of Bmp2 (bone morphogenetic protein 2), and human hair keratin basic 1.[7-9]

Definitive diagnosis is obtained through biopsy, looking for characteristic histopathologic findings. The lesion usually is found in the lower dermis and subcutaneous fat. However, in the perforating variant, the lesion is more superficial, located in the papillary and mid dermis, as seen in our patient.[10] Pilomatricomas are sharply demarcated, often surrounded by a connective-tissue capsule. Histopathologic analysis reveals islands of epithelial cells comprised of 3 subtypes: basophilic cells with scant cytoplasm, shadow cells with a central pallor (Figure), and transitional cells between the former 2 cellular types.[11] The number of basophilic and transitional cells is inversely related to the number of shadow cells. In older lesions, the shadow cells predominate, while the basophilic cells are few in number or absent. Calcium deposits are seen in 80% of lesions with von Kossa staining.[12]

Transformation into malignancy, known as pilomatrical carcinoma, is rare. These malignant neoplasms are characterized by aggressive biologic behavior such as recurrence, diffuse spread, or metastasis, or by cytologic abnormalities such as poor cellular organization, squamous differentiation, and conspicuous mitotic activity.[13] The recent growth of the long-standing lesion in our patient might be interpreted as a sign of malignant transformation. However, this observation may be related to the intense inflammatory reaction supported by the histopathology.

Pilomatricomas are not associated with mortality. Pilomatrical carcinomas are uncommon but are locally invasive and can cause visceral metastases and death. Spontaneous regression has never been observed and medical treatment is ineffective. The treatment of choice is incision and curettage or surgical excision.[14] Although recurrence has only been reported in 2.6% of cases from a large case series (N=228), patients should be monitored after surgical excision.[12]



Islands of pale eosinophilic shadow cells with an intense dermal infiltrate consisting of lymphocytes, histiocytes, plasma cells, and neutrophils (H&E, original magnification ×400).

REFERENCES

1. Malherbe A, Chenantais J. Note sur l'epithelioma calcifie des glandes sebacees. *Prog Med*. 1880;8:826-828.
2. Julian CG, Bowers PW. A clinical review of 209 pilomatricomas. *J Am Acad Dermatol*. 1998;39(2, pt 1):191-195.
3. Lozzi GP, Soyer HP, Fruehauf J, et al. Giant pilomatricoma. *Am J Dermatopathol*. 2007;29:286-289.
4. Hubbard VG, Whittaker SJ. Multiple familial pilomatricomas: an unusual case. *J Cutan Pathol*. 2004;31:281-283.
5. Kizawa K, Toyoda M, Ito M, et al. Aberrantly differentiated cells in benign pilomatrixoma reflect the normal hair follicle: immunohistochemical analysis of Ca-binding S100A2, S100A3 and S100A6 proteins. *Br J Dermatol*. 2005;152:314-320.
6. Farrier S, Morgan M. bcl-2 expression in pilomatricoma. *Am J Dermatopathol*. 1997;19:254-257.
7. Park SW, Suh KS, Wang HY, et al. Beta-catenin expression in the transitional cell zone of pilomatricoma. *Br J Dermatol*. 2001;145:624-629.
8. Kurokawa I, Kusumoto K, Bessho K, et al. Immunohistochemical expression of bone morphogenetic protein-2 in pilomatricoma. *Br J Dermatol*. 2000;143:754-758.
9. Cribier B, Asch PH, Regnier C, et al. Expression of human hair keratin basic 1 in pilomatrixoma: a study of 128 cases. *Br J Dermatol*. 1999;140:600-604.
10. Bayle P, Bazex J, Lamant L, et al. Multiple perforating and non perforating pilomatricomas in a patient with Churg-Strauss syndrome and Rubinstein-Taybi syndrome. *J Eur Acad Dermatol Venereol*. 2004;18:607-610.
11. Elder D, Elenitsas R, Ragsdale BD. Pilomatricoma. In: Elder D, Elenitsas R, Jaworsky C, et al, eds. *Histopathology of the Skin*. 8th ed. Philadelphia, PA: Lippincott-Raven; 1997:757-759.
12. Forbis R Jr, Helwig EB. Pilomatrixoma (calcifying epithelioma). *Arch Dermatol*. 1961;83:606-618.
13. Wood MG, Parhizgar B, Beerman H. Malignant pilomatricoma. *Arch Dermatol*. 1984;120:770-773.
14. Thomas RW, Perkins JA, Ruegemer JL, et al. Surgical excision of pilomatrixoma of the head and neck: a retrospective review of 26 cases. *Ear Nose Throat J*. 1999;78:541, 544-546, 548.