Numerous large nodules on scalp

The patient told us that his father had "cysts" on his body, too. This familial connection provided a clue to the diagnosis.

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A 31-YEAR-OLD HISPANIC MAN presented for evaluation of numerous disfiguring growths on his scalp. They first appeared when he was 19 years old. A review of his family history revealed that his father had 2 "cysts" on his body.

The patient had 10 nodules on his scalp and upper back (**FIGURES 1A AND 1B**). The ones on his scalp lacked puncta and appeared in a "turban tumor" configuration. The lesions were pink, smooth, and semisoft, and ranged in size from 1 to 6 cm.

Six years earlier, the patient had been seen for evaluation of 20 protuberant nodules. At the time, he had been referred to plastic surgery, where 15 lesions were excised. No other treatment was reported by the patient during the 6-year gap between exams.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE 1
Large nodules on the front (**A**) and back (**B**) of the patient's scalp

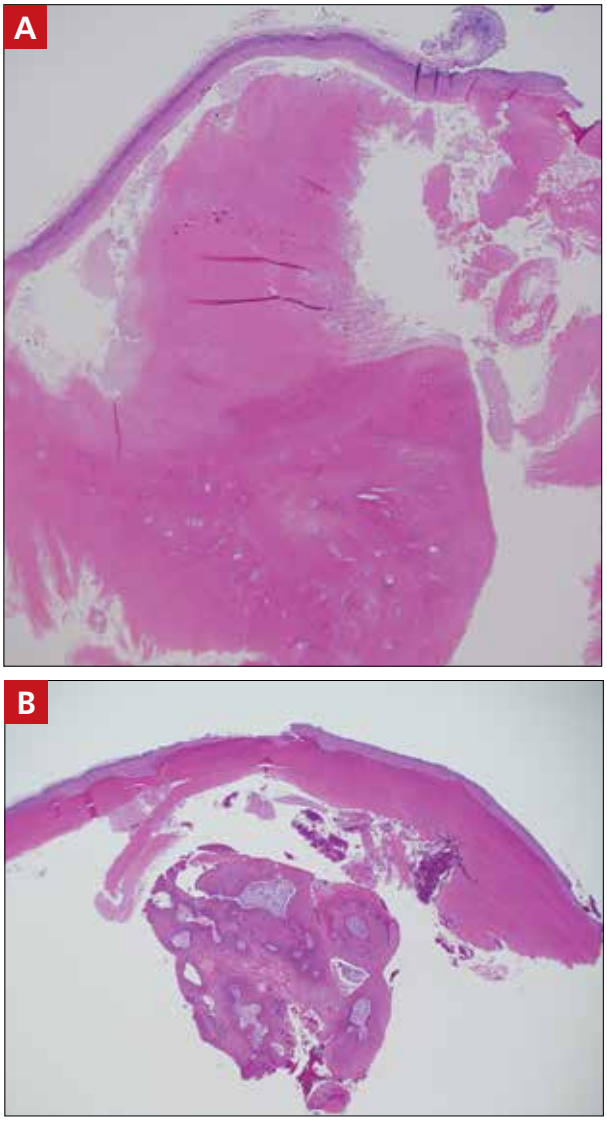
IMAGES COURTESY OF MARGARET E. BROWN, MD

Diagnosis: Pilar cysts

Pilar cysts (PC), also known as *trichilemma cysts*, *wen,* or *isthmus-catagen cysts*, are benign cysts that manifest as smooth, firm, well-circumscribed, pink nodules. PCs originate from the follicular isthmus of the hair's external root sheath[1] and are found in 5% to 10% of the US population.[2] Possible sites of appearance include the face, neck, trunk, and extremities, although 90% of PCs develop on the scalp.[1] They tend to have an autosomal dominant pattern of inheritance with linkages to the short arm of chromosome 3.[3] PCs can occasionally become inflamed following infection or trauma.

FIGURE 2

Nonproliferating and proliferating cysts excised from patient's scalp



(**A**) Shows a nonproliferating pilar cyst, as demonstrated by trichilemmal keratinization, while (**B**) shows a proliferating pilar cyst, as exhibited by multiple foci of squamous cell proliferation within the dermis. (H&E, ×20)

Characteristic histology of PCs demonstrates semisolid, keratin-filled, subepidermal cysts lined by stratified epithelium without a granular layer (trichilemmal keratinization). Lesions excised from this patient's scalp showed 2 subtypes of PCs: nonproliferating (**FIGURE 2A**) and proliferating (**FIGURE 2B**). Subtypes appear similar on exam but can be differentiated on histology.

With gradual growth, proliferating PCs can reach up to 25 cm in diameter.[1] Rapid growth, size > 5 cm, infiltration, or a non-scalp location may indicate malignancy.[4]

Differential diagnosis includes lipomas

The differential diagnosis for a lesion such as this includes epidermal inclusion cysts, dermoid cysts, and lipomas. Epidermal inclusion cysts have a punctum, whereas PCs do not. Dermoid cysts are single congenital lesions that manifest much earlier than PCs. Lipomas are easily movable rubbery bulges that appear more frequently in lipid-dense areas of the body.

For this patient, the striking turban tumor–like presentation, with numerous large cysts on the scalp, initially inspired a differential diagnosis including several genetic tumor syndromes. However, unlike the association between Gardner syndrome and numerous epidermoid cysts or Brooke-Spiegler syndrome and spiradenomas, no syndromes have been linked to numerous trichilemmal cysts.

Excision is effective

Excision is the treatment of choice for both proliferating and nonproliferating PCs.[5] The local recurrence rate of proliferating PCs is 3.7% with a rare likelihood of transformation to trichilemmal carcinoma.[6]

Our patient continues to be followed in clinic for monitoring and periodic excision of bothersome cysts. JFP

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