A 75-year-old woman presented with an enlarging plaque on the scalp of 5 years’ duration. Physical examination revealed a 5.6 × 2.9-cm, tan-colored, verrucous plaque with an overlying pink friable nodule on the left occipital scalp. The lesion was not painful or pruritic, and the patient did not have any constitutional symptoms such as fever, night sweats, or weight loss. The patient denied prior tanning bed use and reported intermittent sun exposure over her lifetime. She denied any prior surgical intervention. There was no family history of similar lesions.

WHAT’S THE DIAGNOSIS?

a. adnexal neoplasm arising in a nevus sebaceous
b. atypical fibroxanthoma
c. basal cell carcinoma
d. cylindroma
e. metastatic renal cell carcinoma

PLEASE TURN TO PAGE E20 FOR THE DIAGNOSIS
Biopsy of the lesion showed a proliferation of basaloid-appearing cells with focal ductal differentiation and ulceration consistent with poroma (Figure 1). Due to the superficial nature of the biopsy, the pathologist recommended excision to ensure complete removal and to rule out a well-differentiated porocarcinoma. Excision of the lesion showed large basaloid aggregates with a hypercellular stroma and a surrounding papillomatous epidermis with well-developed sebaceous lobules consistent with a trichoblastoma and a nevus sebaceous, respectively (Figure 2). There also was evidence of poroma; however, there were no findings concerning for porocarcinoma, which could lead to metastasis (Figure 3).

Nevus sebaceus is a benign, hamartomatous, congenital growth that occurs in approximately 1% of patients presenting to dermatology offices. It usually presents as a single asymptomatic plaque on the scalp (62.5%) or face (24.5%) that changes in morphology over its lifetime. In children, a nevus manifests as a yellowish, smooth, waxy skin lesion. As the sebaceous glands become more developed during adolescence, the lesion takes on more of a verrucous appearance and also can darken.

Although nevus sebaceus is benign, it may give rise to both benign and malignant neoplasms. In a 2014 study of 707 cases of nevus sebaceus, 21.4% developed secondary neoplasms, 88% of which were benign. The origins of these neoplasms can be epithelial, sebaceous, apocrine, and/or follicular. The 3 most common secondary neoplasms found in nevus sebaceus are trichoblastoma (34.7%), syringocystadenoma papilliferum (24.7%), and apocrine/eccrine adenoma (10%), all of which are benign. Trichoblastomas represent a type of hair follicle tumor. Malignant lesions manifest in approximately 2.5% of cases, with basal cell carcinoma (BCC) being the most common (5.3% of all neoplasms), followed by squamous cell carcinoma (2.7% of all neoplasms). Differentiating BCC from trichoblastoma can be difficult, but histologically BCCs usually have tumor stromal clefting while trichoblastomas do not. The incidence of secondary tumors in nevus sebaceus displays a strong correlation with age; thus, the highest proportion of neoplasms occur in adults.

Treatment of nevus sebaceus depends on the patient’s age. In children, because of the low probability of secondary neoplasms, observation in lieu of surgical excision is a common approach. In adults, the approach typically is surgical excision or close follow-up, as there is a concern for secondary neoplasm and the potential for malignant degeneration.

A nevus sebaceus leading to 2 or more tumors within the same lesion is rare (seen in only 4.2% of lesions).

**FIGURE 1.** A and B, Scalp biopsy showed poroid-appearing cells and ductal differentiation (H&E, original magnifications ×100 and ×100).

**FIGURE 2.** A and B, Scalp biopsy showed basaloid cells and a hypercellular stroma (H&E, original magnifications ×100 and ×200).
The most common combination is trichoblastoma with syringocystadenoma papilliferum (0.6% of all cases).\(^2\) Poromas represent sweat gland tumors that usually appear on the soles (65%) or palms (10%).\(^4\) It is uncommon for these neoplasms to manifest on the scalp or within a nevus sebaceus. Three independent studies (N=596; N=707; N=450) did not report any occurrences of eccrine poroma.\(^1,2,5\) Eccrine poroma in conjunction with nodular trichoblastoma arising in a nevus sebaceus is unusual, and definitive excision should be strongly considered because of the possibility to develop a porocarcinoma.\(^6\)

Atypical fibroxanthoma presents on sun-exposed areas as an exophytic nodule or plaque that frequently ulcerates. Pathology of this tumor shows a spindled cell proliferation that can stain positively for CD10 and procollagen 1. Basal cell carcinoma presents as a pearly papule or nodule displaying basaloïd-appearing aggregates with tumor stromal clefting and can stain with Ber-EP4. Cylindromas typically present on the scalp as large rubbery-appearing plaques and nodules. Cylindromas usually present as a solitary tumor, but in the familial form there can be clusters of multiple nodules. Metastatic renal cell carcinoma frequently appears as a bleeding nodule on the scalp in patients with known renal cell cancer or as the initial presentation.

REFERENCES