

Child With “Distressing” Problem

A “bald spot” is the chief complaint of a 12-year-old girl brought for evaluation by her mother. The lesion in her left parietal scalp has been there since birth, slowly growing but producing no symptoms.

Although the child’s primary care provider has reassured the family that the “birthmark” is benign, they remain concerned. Furthermore, the patient has become increasingly distressed by the hairlessness.

The child is otherwise healthy. There is no history of excessive sun exposure.

The lesion is a roughly oval, uniformly pink, hairless 3.6-cm plaque with a faintly mammillated surface and well-defined margins. It is only visible when the surrounding hair is parted sufficiently to reveal it. Examination of the rest of the patient’s skin is unremarkable.

Given the facts as presented, the most likely diagnostic explanation is

- Nevus sebaceous
- Aplasia cutis congenita
- Epidermal nevus
- Neonatal lupus

ANSWER

The correct answer is nevus sebaceous (choice “a”). This benign hamartomatous lesion is derived



from local tissue and grows at the same rate.

It differs considerably from the other items in the differential, including aplasia cutis congenita (choice “b”). In this condition, a focal area of epidermis simply fails to develop, leaving a permanent hairless scar that contrasts sharply with the raised, mammillated plaque of nevus sebaceous.

Epidermal nevus (choice “c”) is usually a collection of tan to brown superficial nevoid papules that can be linear, agminated, or plaque-like. These lesions lack the

color and mammillated surface of those seen in nevus sebaceous.

Neonatal lupus (choice “d”) can present at birth with hairless, cicatricial inflamed lesions. However, these tend to resolve quickly, often leaving focal scarring alopecia but no plaque formation.

DISCUSSION

Nevus sebaceous (NS), first described by Jadassohn in 1895, has long been recognized as an unusual but by no means rare congenital lesion. Occurring equally in both sexes and comprising



Joe R. Monroe, MPAS, PA, practices at Dawkins Dermatology Clinic in Oklahoma City. He is also the founder of the Society of Dermatology Physician Assistants.

sebaceous glands in a nevoid morphologic context, NS is considered a variant of sebaceous nevi and verrucous epidermal nevi in some circles. All three are derived from overgrowth of local, normal tissues that typically grow at the same rate as surrounding structures.

The vast majority of NS lesions are found in the scalp, although they can also develop on the ear or neck and, rarely, elsewhere on the body. This patient's plaque—with its uniform surface; tiny, smooth, shiny papules; and (perhaps most important) total lack of hair—is typical. Other classic features are congenital onset and permanent nature, which distinguish them from the rest of the differential.

Focal malignant transformation of NS lesions has been re-

ported—in fact, this author has seen two such cases in 30 years. Both were small basal cell carcinomas, although cases of melanoma and other malignancies have been reported.

Such changes are rare enough that most experts consider prophylactic removal to be unwarranted. Watching the lesions for change over the years is certainly reasonable, as is protecting them from sun exposure.

Surgical removal—usually performed by a plastic surgeon—is occasionally necessary for cosmetic reasons. This is particularly so when NS covers a portion of the face, or when the cosmetic implications of having a hairless plaque in the scalp are sufficiently distressing.

This patient and her parents



Nevus sebaceous, variant of epidermal nevus in the scalp.

were educated about the nature of the diagnosis and apprised of their options. **CR**

*Editor's note: For a similar presentation with a very different diagnosis, see the March 2015 *DermaDiagnosis* case (<http://bit.ly/1ye69Ym>).*



Follow us on

Facebook and Twitter!

Stay up-to-date on the latest health care trends from *Clinician Reviews*! Find all of our content in one spot, just for you. The latest dermatology cases, radiology reviews, and ECG challenges are just a click away.



Like us on Facebook at [facebook.com/ClinRev](https://www.facebook.com/ClinRev) and follow us on **Twitter @ClinRev!**