Hairless Scalp Lesion

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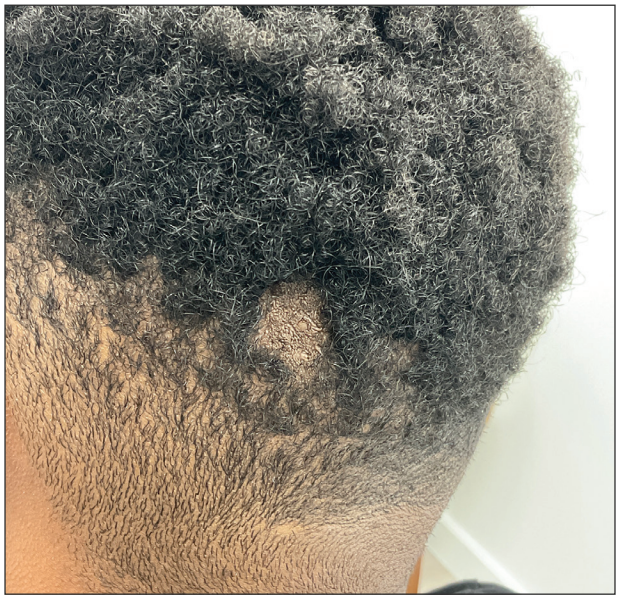

A 23-year-old man presented to the dermatology clinic with hair loss on the scalp of several years' duration. The patient reported persistent pigmented bumps on the back of the scalp. He denied any pruritus or pain and had no systemic symptoms or comorbidities. Physical examination revealed a 1×1.5-cm, yellow-brown, hairless plaque on the left parietal scalp.

WHAT'S YOUR **DIAGNOSIS?**

a. epidermal nevus
b. hypertrophic lichen planus
c. Langerhans cell histiocytosis
d. nevus sebaceus of Jadassohn
e. seborrheic keratosis

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THE **DIAGNOSIS:** Nevus Sebaceus of Jadassohn

The diagnosis of nevus sebaceus of Jadassohn was made clinically based on the lesion's appearance and presence since birth as well as the absence of systemic symptoms. Clinically, nevus sebaceus of Jadassohn typically manifests as a well-demarcated, yellow-brown plaque often located on the scalp, as was seen in our patient. The lack of pruritus and pain further supported the diagnosis in our patient. No biopsy was performed, as the presentation was considered classic for this condition. Our patient opted to forgo surgery and will be routinely monitored for any changes, as nevus sebaceus has a potential risk, albeit low, for malignant transformation later in life. No changes have been observed since the initial presentation, and regular follow-ups are planned to monitor for future developments.

Nevus sebaceus of Jadassohn is a hamartomatous lesion involving the pilosebaceous follicle and adjacent adnexal structures.[1-3] It most commonly forms on the scalp (59.3%) and is accompanied by partial or total alopecia.[3,4] It is seen less often on the face, periauricular area, or neck[1,4]; thorax or limbs[5]; and oral or genital mucosae.[6] Nevus sebaceus of Jadassohn affects approximately 0.3% of newborns,[1] usually as a solitary lesion that can form an extensive plaque. The male-to-female occurrence ratio has been reported as equal to slightly more predominant in females; all races and ethnicities are affected.[1,5]

Nevus sebaceus of Jadassohn follows 3 stages of clinical development: infantile, adolescent, and adulthood. It manifests at birth or shortly afterward as a smooth hairless patch or plaque that is yellowish and can be hyperpigmented in Black patients.[5] It may have an oval or linear configuration, typically is asymptomatic, and often arises along the Blaschko lines when it occurs as multiple lesions (a rare manifestation).[1] During puberty, hormonal changes cause accelerated growth, sebaceous gland maturation, and epidermal hyperplasia.[7] Nevus sebaceus of Jadassohn often is not identified until this stage, when its classic wartlike appearance has fully developed.[1]

Patients with nevus sebaceus of Jadassohn have a 10% to 20% risk for tumor development in adulthood.[2,7] Trichoblastoma and syringocystadenoma papilliferum are the most frequently described neoplasms.[8] Basal cell carcinoma is the most common malignant secondary neoplasm with an occurrence rate of 0.8%.[6,9] However, basal cell carcinoma and trichoblastoma may share histopathologic features, which may lead to misdiagnosis and a higher reported incidence of basal cell carcinoma in adults than is accurate.[2]

Early prophylactic surgical removal of nevus sebaceus of Jadassohn has been recommended; however, surgical management is controversial because the risk for a benign secondary neoplasm remains relatively high while the risk for malignancy is much lower.[2,7] Surgical excision remains an acceptable option once the patient is mature enough to tolerate the procedure.[1] However, patient education regarding watchful waiting vs a surgical approach—and the risks of each—is critical to ensure shared decision-making and a management plan tailored to the individual.

The differential diagnosis includes hypertrophic lichen planus, Langerhans cell histiocytosis (Letterer-Siwe disease type), epidermal nevus, and seborrheic keratosis. Hypertrophic lichen planus often occurs symmetrically on the dorsal feet and shins with thick, scaly, and extremely pruritic plaques. The lesions often persist for an average of 6 years and may lead to multiple keratoacanthomas or follicular base squamous cell carcinomas. Langerhans cell histiocytosis (Letterer-Siwe disease type) manifests with acute, disseminated, visceral, and cutaneous lesions before 2 years of age. These lesions appear as 1- to 2-mm, pink, seborrheic papules, pustules, or vesicles on the scalp, flexural neck, axilla, perineum, and trunk; they often are associated with petechiae, purpura, scale, crust, erosion, impetiginization, and tender fissures. Epidermal nevus occurs within the first year of life and is a hamartoma of the epidermis and papillary dermis. It manifests as papillomatous pigmented linear lines along the Blaschko lines. Seborrheic keratosis manifests as well-demarcated, waxy/verrucous, brown papules with a "stuck on" appearance on hair-bearing skin sparing the mucosae. They are common benign lesions associated with sun exposure and often manifest in the fourth decade of life.[10]

REFERENCES

1. Baigrie D, Troxell T, Cook C. Nevus sebaceus. *StatPearls [Internet]*. Updated August 16, 2023. Accessed September 12, 2024. https://www.ncbi.nlm.nih.gov/books/NBK482493/
2. Terenzi V, Indrizzi E, Buonaccorsi S, et al. Nevus sebaceus of Jadassohn. *J Craniofac Surg*. 2006;17:1234-1239. doi:10.1097/01.scs.0000221531.56529.cc
3. Kelati A, Baybay H, Gallouj S, et al. Dermoscopic analysis of nevus sebaceus of Jadassohn: a study of 13 cases. *Skin Appendage Disord*. 2017;3:83-91. doi:10.1159/000460258
4. Ugras N, Ozgun G, Adim SB, et al. Nevus sebaceous at unusual location: a rare presentation. *Indian J Pathol Microbiol*. 2012;55:419-420. doi:10.4103/0377-4929.101768
5. Serpas de Lopez RM, Hernandez-Perez E. Jadassohn's sebaceous nevus. *J Dermatol Surg Oncol*. 1985;11:68-72. doi:10.1111/j.1524-4725.1985.tb02893.x
6. Cribier B, Scrivener Y, Grosshans E. Tumors arising in nevus sebaceus: a study of 596 cases. *J Am Acad Dermatol*. 2000;42(2 pt 1):263-268. doi:10.1016/S0190-9622(00)90136-1

7. Santibanez-Gallerani A, Marshall D, Duarte AM, et al. Should nevus sebaceus of Jadassohn in children be excised? a study of 757 cases, and literature review. *J Craniofac Surg*. 2003;14:658-660. doi:10.1097/00001665-200309000-00010
8. Chahboun F, Eljazouly M, Elomari M, et al. Trichoblastoma arising from the nevus sebaceus of Jadassohn. *Cureus*. 2021;13:E15325. doi:10.7759/cureus.15325
9. Cazzato G, Cimmino A, Colagrande A, et al. The multiple faces of nodular trichoblastoma: review of the literature with case presentation. *Dermatopathology (Basel)*. 2021;8:265-270. doi:10.3390/dermatopathology8030032
10. Dandekar MN, Gandhi RK. Neoplastic dermatology. In: Alikhan A, Hocker TLH (eds). *Review of Dermatology*. Elsevier; 2016: 321-366.