Yellow-Orange Hairless Plaque on the Scalp

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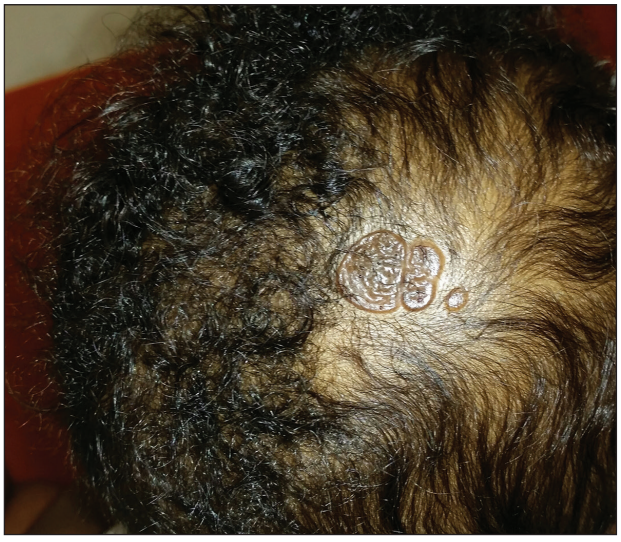

An otherwise healthy 13-month-old boy presented with a well-circumscribed, 3×4-cm, yellow-orange plaque with a verrucous velvety surface on the right side of the posterior scalp. The patient was born at 33 weeks' gestation and had an uneventful perinatal course with a normal head ultrasound at 4 days of age. The lesion had been present since birth and initially was comprised of waxy, yellow-orange, hairless plaques that became more thickened and noticeable over time. The mother recalled that the surface of the plaque initially was flat and smooth but gradually became bumpier and greasier in consistency in the months prior to presentation. The patient was otherwise asymptomatic.

WHAT'S THE **DIAGNOSIS?**

a. apocrine carcinoma
b. basal cell carcinoma
c. nevus sebaceous
d. squamous cell carcinoma
e. tinea capitis

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THE **DIAGNOSIS:** Nevus Sebaceous

The patient presented with a typical solitary scalp lesion characteristic of nevus sebaceous (NS). The lesion was present at birth as a flat and smooth hairless plaque; however, over time it became more thickened and noticeable, which prompted the parents to seek medical advice.

Nevus sebaceous, also known as NS of Jadassohn, is a benign congenital hamartoma of the sebaceous gland that usually is present at birth and frequently involves the scalp and/or the face. The classic NS lesion is solitary and appears as a well-circumscribed, waxy, yellow-orange or tan, hairless plaque. Despite the presence of these lesions at birth, they may not be noted until early childhood or rarely until adulthood. Generally, the lesion tends to thicken and become more verrucous and velvety over time, particularly around the time of reaching puberty.[1] Clinically, NS lesions vary in size from 1 cm to several centimeters. Lesions initially tend to grow proportionately with the child until puberty when they become notably thicker, greasier, and verrucous or nodular under hormonal influences. The yellow discoloration of the lesion is due to sebaceous gland secretion, and the characteristic color usually becomes less evident with age.

Nevus sebaceous occurs in approximately 0.3% of newborns and tends to be sporadic in nature; however, rare familial forms have been reported.[2,3] Nevus sebaceous can present as multiple nevi that tend to be extensive and distributed along the Blaschko lines, and they usually are associated with neurologic, ocular, or skeletal defects. Involvement of the central nervous system frequently is associated with large sebaceous nevi located on the face or scalp. This association has been termed *NS syndrome*.[4] Neurologic abnormalities associated with NS syndrome include seizures, mental retardation, and hemimegalencephaly.[5] Ocular findings most communally associated with the syndrome are choristomas and colobomas.[6-8]

There are several benign and malignant epithelial neoplasms that may develop within sebaceous nevi. Benign tumors include trichoblastoma, syringocystadenoma papilliferum, trichilemmoma, sebaceoma, nodular hidradenoma, and hidrocystoma.[1,8,9] Malignant neoplasms include basal cell carcinoma (BCC), apocrine carcinoma, sebaceous carcinoma, and squamous cell carcinoma. The lifetime risk of malignancy in NS is unknown. In an extensive literature review by Moody et al[10] of 4923 cases of NS for the development of secondary benign and malignant neoplasms, 16% developed benign tumors while 8% developed malignant tumors such as BCC. However, subsequent studies suggested that the incidence of BCC may have been overestimated due to misinterpretation of trichoblastoma and may be less than 1%.[11-13]

Usually the diagnosis of NS is made clinically and rarely a biopsy for histopathologic confirmation may be needed when the diagnosis is uncertain. Typically, these histopathologic findings include immature hair follicles, hyperplastic immature sebaceous glands, dilated apocrine glands, and epidermal hyperplasia.[9] For patients with suspected NS syndrome, additional neurologic and ophthalmologic evaluations should be performed including neuroimaging studies, skeletal radiography, and analysis of liver and renal function.[14]

The current standard of care in treating NS is full-thickness excision. However, the decision should be individualized based on patient age, extension and location of the lesion, concerns about the cosmetic appearance, and the risk for malignancy.

The 2 main reasons to excise NS include concern about malignancy and undesirable cosmetic appearance. Once a malignant lesion develops within NS, it generally is agreed that the tumor and the entire nevus should be removed; however, recommendations vary for excising NS prophylactically to decrease the risk for malignant growths. Because the risk for malignant transformation seems to be lower than previously thought, observation can be a reasonable choice for lesions that are not associated with cosmetic concern.[12,13]

Photodynamic therapy, CO_2 laser resurfacing, and dermabrasion have been reported as alternative therapeutic approaches. However, there is a growing concern on how effective these treatment modalities are in completely removing the lesion and whether the risk for recurrence and potential for neoplasm development remains.[1,9]

This patient was healthy with normal development and growth and no signs of neurologic or ocular involvement. The parents were counseled about the risk for malignancy and the long-term cosmetic appearance of the lesion. They opted for surgical excision of the lesion at 18 months of age.

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