

Case 14

A 30 year-old Thai woman from Bangkok

Chief complaint: An asymptomatic nodule on the scalp for 8 months



Present illness:

An asymptomatic hairless, yellowish lesion on her scalp has been observed since she was born. Eight months earlier, a red nodule arose within the lesion and gradually increased in size. There was no history of itching, pain, or bleeding. She is otherwise healthy.

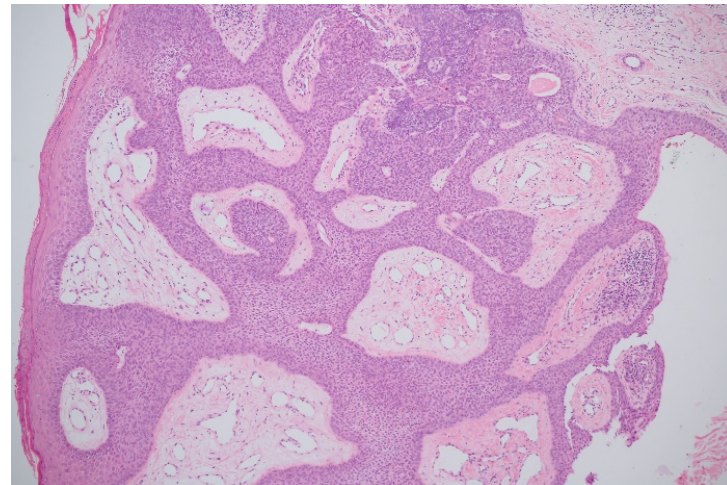
Past history:

- She has no major illnesses including dermatologic diseases.
- None of her family members has similar cutaneous lesions or dermatologic diseases.

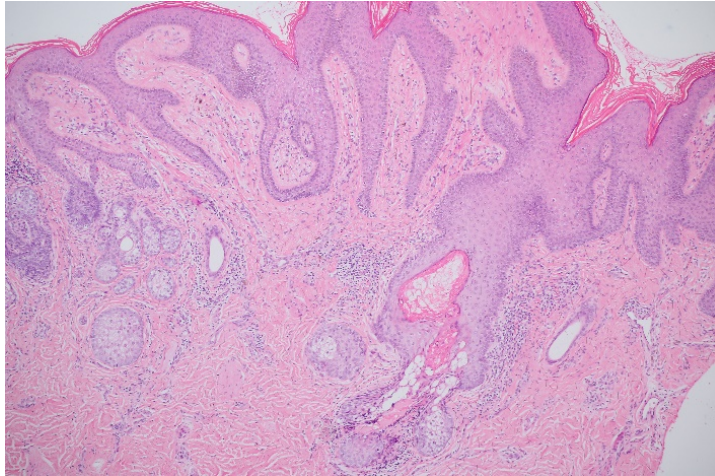
Skin examination:

- A slightly verrucous, dome-shaped, erythematous nodule with minimal scales and crusts on a well-demarcated, hairless, verrucous, yellowish plaque measuring 3x6 cm, on the left frontoparietal site of the scalp. There was no enlargement of cervical, submandibular, and occipital lymph nodes. Other systems were unremarkable.

Histopathology: (S16-5403A, S16-5403B, scalp)



- Anastomosing epithelial bands emanating from the epidermis
- Epithelial bands composed of uniformly cuboidal cells (poroid cells), some with ductal lumina
- Fibrovascular stroma



- Hyperkeratosis and papillae epidermal hyperplasia
- Malformed pilosebaceous units composed of abortive hair follicles and premature sebaceous glands

Diagnosis: Eccrine poroma arising within nevus sebaceous

Treatment: Wide surgical excision by plastic surgery

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Discussion:

Nevus sebaceous, otherwise known as nevus sebaceous of Jadassohn or organoid nevus, is a congenital hamartoma originating from cutaneous structures including both surface epithelial and adnexal components.^{1, 2} It is typically noticed since birth or early childhood as either a separate entity or in association with neurologic, ophthalmic, or other defects (epidermal nevus

syndrome), particularly when lesions are multiple.³ Nevertheless, it is mostly found in otherwise healthy children.⁴ Clinically, it presents as a well-demarcated, oval or linear, hairless, yellowish, waxy, and verrucous plaque, and mainly affects the head and neck area, most commonly the scalp.²⁻⁴ According to Mehregan and Pinkus, there are three stages of nevus sebaceous including infancy, puberty, and adulthood.⁵ At or shortly after birth, it is usually seen as a well-demarcated, hairless, waxy, yellowish red plaque. Due to the androgen effects on sebaceous glands during puberty, the lesion usually completes its characteristic pathology as becoming thicker and verrucous. The third stage may occur when associated benign or malignant tumors develop within the primary lesions.

The various histologic features of nevus sebaceous are described corresponding to the stages of the lesion. In well-formed lesions seen in adults, papillomatous and verrucous epidermal hyperplasia are reported. Lobules of heterotropic/ectopic apocrine glands in the reticular dermis are characteristically seen. The sebaceous glands are numerous and prominent, and directly open into the epidermis.³ In general, the diagnosis of nevus sebaceous can be established based on its typical clinical characteristics. However, histopathologic examination is needed if the clinical diagnosis is uncertain.⁶

As mentioned earlier, several types of secondary tumors developing in nevus sebaceous have been reported with the varied prevalence among published studies, between 8.5 and 22.5%.^{1, 2, 7} In the largest recent study, trichoblastoma was the most common benign tumor arising within nevus sebaceous lesions followed by syringocystadenoma papilliferum, accounting for 7.4% and 5.2%, respectively. Other benign tumors including tricholemmoma, desmoplastic trichilemmoma, sebaceoma, solar keratosis, adenomyoepithelioma, and connective tissue nevus were less commonly reported. Basal cell carcinoma became the most common malignant tumor whereas squamous cell carcinoma ranked

second, representing 1.1% and 0.57%, respectively.¹ Multiple tumors developing within one lesion were also reported.^{1, 7-9} Eccrine poroma is rarely seen as an associated tumor growing in nevus sebaceous. To our knowledge, there are few cases reported in the published literature.⁸⁻¹⁰

In our case, the erythematous nodule of the scalp should be differentiated with benign and malignant tumors frequently occurring on nevus sebaceous including trichoblastoma, syringocystadenoma papilliferum, basal cell carcinoma, and squamous cell carcinoma. In addition, infections such as nontuberculous mycobacterium infection and warts should be also considered.

Eccrine poroma is a benign adnexal tumor of the intraepidermal part of eccrine sweat duct. It was first described by Pinkus et al. in 1956.¹¹ It tends to affect middle-aged to elderly individuals. A solitary, dome-shaped, skin-colored to pink, smooth or verrucous, papule or nodule is a common manifestation of eccrine poroma.^{12, 13} The predilection sites are soles of feet (65%) followed by hands (10%).¹¹ However, it was recently shown that it could involve in other areas including trunk, scalp, forehead, cheek, neck, chin, temple, and auricle.^{12, 14, 15} In comparison with lesions on extremities, the tumors located on head and neck were more likely to be asymptomatic and slow growing whereas patients with lesions on extremities might report clinical presentation such as bleeding, discharge, and rapid growth or symptoms such as pain.^{13, 14} Histologic findings reveal aggregations of proliferative cuboidal or poroid cells that spread from the basal layer of the epidermis into the dermis. It has been indicated that 18% of poromas transform into porocarcinoma (malignant poroma). Lesions with spontaneous bleeding, ulcers, sudden itching or pain, and rapid growth in a short duration should be carefully excluded for malignancy.¹³

The standard treatment for nevus sebaceous is full-thickness surgical excision.⁶ Historically, prophylactic surgery in

children was advocated due to the possibility in developing associated malignant tumors. However, the risk of secondary malignancy developing before puberty is very low according to recent studies.^{1, 2, 4, 7} Mohs Micrographic Surgery can be considered as an option for removal of nevus sebaceous with associated carcinoma.¹⁶ Although treatments such as photodynamic therapy, carbon dioxide laser, and dermabrasion showed unpredictable outcomes, they can be considered as alternative options.¹⁷⁻²¹ Close follow-up for malignancy surveillance should be warranted in all cases whose nevus sebaceous is not totally removed.⁶

Our case was referred to plastic surgery for wide excision. The lesion was completely removed with satisfactory cosmetic result. There was no recurrence at 3-month follow-up.

References

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