Solitary hairless lesion on scalp

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CLINICAL FINDINGS

An 8-year-old male child presented to our clinic with hairless lesion noted on the scalp in early childhood, it started as small lesion until the last year, when it increased in size and patient started complaining of disfigurement. The patient didn't receive any treatment for it. There was no past history of similar condition or other skin lesions. The patient had no systemic complaints and there was no family history of similar lesions.

Local examination of the scalp revealed solitary hairless, slightly raised, yellowish plaque, with somewhat verrucous surface. The lesion was about 4 cm in length. [Fig. 1]. Skin, nail and mucous membranes were not affected and showed no significant abnormalities.

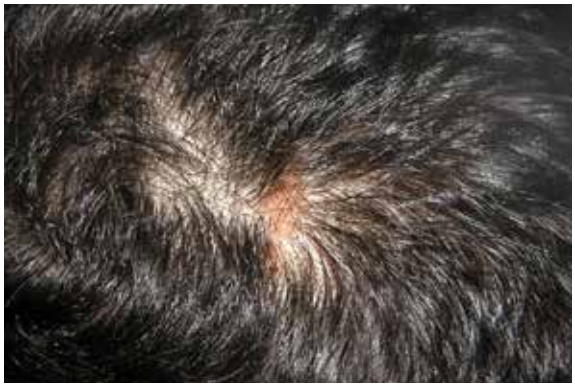

Fig. 1 Solitary hairless, slightly verrucous surface, yellowish plaque in frontal region of scalp.

What is your clinical differential diagnosis?

Aplasia Cutis Congenita
Nevus sebaceous
Wart
Seborrheic Keratosis
Verrucous melanocytic nevus

PATHOLOGICAL FINDINGS

The epidermis shows hyperkeratosis, acanthosis and papillomatosis. The dermis shows numerous mature sebaceous glands attached to hair follicles. Superficial perivascular inflammatory infiltrate formed of lymphohistiocytic cells admixed with melanophages [Fig 2, 3].

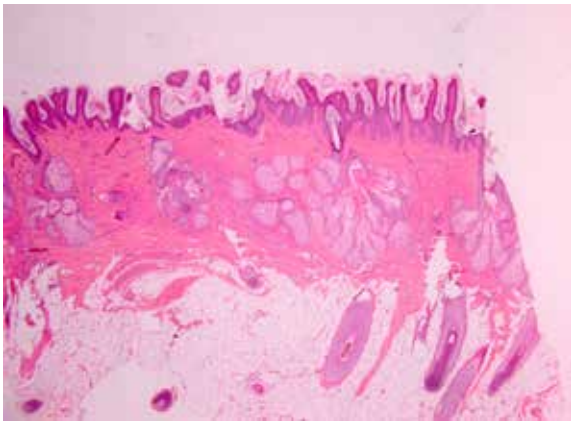

Fig. 2 Hyperkeratosis, acanthosis, papillomatosis and numerous sebaceous glands.

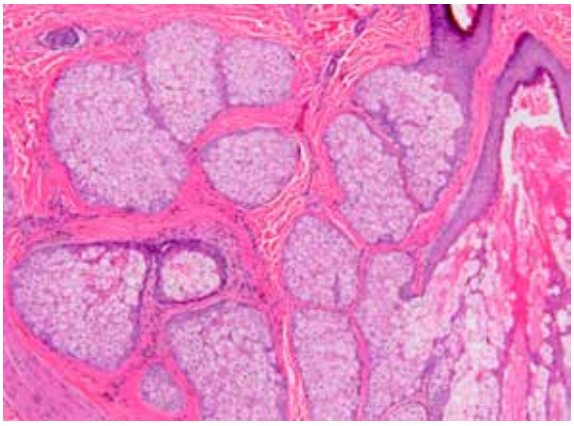

Fig. 3 Mature sebaceous glands attached to hair follicles.

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DIAGNOSIS

Nevus sebaceous of Jadassohn (NSJ)

COMMENT

Sebaceous Nevus or Nevus sebaceous of Jadassohn is a well circumscribed hamartomatous lesion comprised predominantly of sebaceous glands. Most of the sebaceous nevi occur sporadically but some familial cases have been reported.[1] The lesion usually starts as an isolated plaque at birth or may develop later during childhood and remains unchanged until puberty, when it becomes thickened and more elevated by the effect of sex hormones.[2] These lesions have a predilection for the scalp and characteristically appear early in life as a hairless patch or small plaque. Often, they do not cause problems until the patient reaches adolescence, as hormonal factors induce a verrucous or nodular change and the lesion grows in size then patient complains about its cosmetic disfigurement. Recently, it has been shown nevus sebaceous is caused by post zygotic mosaic mutations in the HRAS and KHAS genes.[3] Perhaps reflecting inheritance of an unstable premutation, but attributed by Happle and Konig to predominant inheritance.[4] Because sebaceous nevi have a tendency to develop tumors, various tumor genes have been implicated in their etiology. Constitutive activation of the patched-hedgehog signaling pathway (involved in Gorlin's syndrome and basal cell carcinomas) was suggested by Xin et al.[5] Who found loss of heterozygosity at the PTCH locus 9q22.3 in sebaceous nevi. However, this finding was described by Takata et al.[6] Who also found lack of expression of Gli-1, another gene in the patched-hedgehog signaling pathway expressed in basal cell carcinomas. Mutations in the oncogene PIC have recently been reported in epidermal nevi.[7]

Such nevi are present at birth, or early childhood, affecting males and females of all races equally.[8] The condition was first described by Josef Jadassohn, a German dermatologist, and now bears his name.[9] Nevus sebaceous occurs in approximately 0.3% of all newborn infants.[10]

NSJ usually affects the face and the scalp.[11] The natural tendency of NSJ is to evolve through 3 stages that were first described in 1965 by Mehregan and Pinkus.[12] In the infantile stage, the lesion presents as a characteristic bright yellow hairless plaque. Histologically, a paucity of underdeveloped sebaceous glands and hair follicles is noticed. At the puberty stage, the growth of the lesion is accelerated, and it becomes verrucous with yellow lobular structures. The final stage is characterized by the appearance of nodules or tumors, with the presence of thin telangiectasias in lesions of longer evolution.[13]

A variety of appendageal tumors, sometimes multiple, may develop within sebaceous nevi.[14-31] The most commonly reported are syringocystadenoma papilliferum and trichoblastoma;[14] less common tumors include nodular hidradenoma,[15] apocrine cystadenoma,[16] syringoma, infundibuloma and trichoadenoma[17] and trichilemmoma and pilomatricoma.[18] Locally invasive and malignant tumours include keratoacanthoma,[19] proliferating trichilemmal cyst,[20] and basal cell,[21-24] sebaceous, apocrine,[25] eccrine[26] and squamous carcinomas[17,23] and malignant melanoma.[27] Leiomyosarcoma occurred with trichoblastoma and syringocystadenoma papilliferum in one case.[28]

The true incidence of malignancy in sebaceous nevi is difficult to determine for several reasons, but mainly because different criteria were used to gather cases in each of the larger series. Further-

Diagnosis	Clinical	Histological
Aplasia cutis congenita	• Presents with a localized loss of skin of variable thickness and is rarely associated with underlying skeletal abnormalities • Findings can include an open ulceration or a healed atrophic scar.	• The epidermis may be absent or thin. • The underlying dermis is usually thin and composed of loosely arranged connective tissue in which there is some disarray of collagen fibers. • The dermis may resemble a scar • Elastic fibers may be reduced, increased, or fragmented. • Appendages are absent or rudimentary • The subcutis is usually thin.
Wart	• They are characterized as flat or slightly elevated flesh-colored papules that may be smooth or slightly hyperkeratotic • They range from 1-5 mm or more, and numbers range from a few to hundreds of lesions that may become grouped or confluent	• Showing the characteristic features hyperkeratosis, acanthosis • Hypergranulosis, rete ridge elongation, and large blood vessels at the dermoepidermal junction
Seborrheic keratosis	• Single or multiple, sharply demarcated pigmented lesion that protrudes above surface of skin • Appears to be stuck to skin • Soft, tan-black, "greasy" surface • Few millimeters to several centimeters in size • Commonly on trunk	• Acanthotic proliferations of small cuboidal keratinocytes without cytologic atypia • Usually has “horn pseudocysts” (round intralesional cysts of loose keratin)
Verrucous Melanocytic nevus	• Dark brown verrocus lesion with raised surface since childhood	• Characterized by a diffuse infiltration of nevus cells extending around blood vessels, nerves, salivary ducts as well as between collagen bundles

more, basaloid proliferation has almost certainly been misinterpreted as basal cell carcinoma, the commonest malignancy reported in sebaceous naevi. Takata et al. found clear molecular differences between the basal cell carcinoma-like trichoblastomas arising in sebaceous nevi and true basal cell carcinomas.[6]

Development of an exophytic nodule on a nevus sebaceous usually represents a benign appendageal tumour or viral wart.[31] Rapid, circumscribed enlargement or ulceration should arouse suspicion of malignant transformation. This usually occurs in middle age, but can undoubtedly occur in adolescence, or even in childhood.[19] The lifetime risk of malignant transformation is probably less than 5%, but is difficult to establish with any precision because the major studies have varied considerably in methods of selection and in ages of the patients.[29,31] The most common malignancy is basal cell carcinoma, but the incidence of this tumor has been overestimated because of misinterpretation of trichoblastoma[14] and basaloid proliferation as

basal cell carcinoma. Other malignant tumors reported include squamous,[17,23] sebaceous,[32] tricholemmal[33] and apocrine[25] carcinomas, and malignant melanoma.[27] Leiomyosarcoma occurred with trichoblastoma and syringocystadenoma papilliferum in one case.[28] Despite occasionally aggressive histopathological features, most of these tumors are of low-grade malignancy. Nevertheless, local recurrence after excision, metastasis and a lethal outcome have all been reported.[29] It is not unusual to find several different types of tumour arising from nevus sebaceous.[17,18,28,33]

Associated abnormalities occur in a small proportion of cases. These are considered under the heading of 'epidermal nevus syndrome'.

Treatment include removal during childhood may be necessary for cosmetic reasons but is difficult to justify on grounds of risk of malignancy.[34] Excision of scalp lesions with primary closure gives an excellent cosmetic result, but larger lesions may require tissue expansion. Simple excision is generally adequate even in the presence of histological malignancy. Superficial removal by dermabrasion or carbon dioxide laser is likely to be followed by partial recurrence but may be helpful where excision is not feasible.[35] An extensive nevus sebaceous of face and scalp in a 38-year-old woman was cleared with 13 sessions of photodynamic therapy, with no recurrence 16 months later.[36]

REFERENCES

1. Sahl WJ: Familial nevus sebaceous of Jadassohn: occurrence in three generations. J Am Acad Dermatol 1990; 22:853-54.
2. Hamilton KS, Johnson S, Smoller BR: The role of androgen receptors in the clinical course of nevus sebaceous of Jadassohn.
3. Happle R; Nevus sebaceus is a mosaic RASopathy. J Invest Dermatol, 2013; 133(3):597-600
4. Happle R, Konig A. Familial nevus sebaceus may be explained by paradominant transmission. Br J Dermatol 1999; 141:377.
5. Xin H, Matt D, Qin JZ et al. The sebaceous nevus: a nevus with deletions of the PTCH gene. Cancer Res 1999; 59:1834-36.
6. Takata M, Tojo M, Hatta N et al. No evidence of deregulated patched-hedgehog signaling pathway in trichoblastomas and other tumors arising within nevus sebaceus. J Invest Dermatol 2001; 117:1666-70.
7. 10 Hafner C, López-Knowles E, Luis NM et al. Oncogenic PIK3CA mutations occur in epidermal nevi and seborrheic keratoses with a characteristic mutation pattern. Proc Natl Acad Sci USA 2007; 104:13450-54.
8. Teng, Joyce M.C. Nevus sebaceous, University of Wisconsin Hospitals and Clinics Authority, last updated 16 November 2007.
9. Saedi T, Cetas J, Chang R, Krol A, Selden NR; Newborn with sebaceous nevus of jadassohn presenting as exophytic scalp lesion. PediatrNeurosurg, 2008; 44(2):144-47.
10. Terenzi V, Indrizzi E, Buonaccorsi S, Leonardi A, Pellacchia V, Fini G; Nevus sebaceus of Jadassohn. J Craniofac Surg, 2006; 17(6):1234-39.
11. Pereira LB, Gontijo B, Silva CM: Dermatoses neonatais. A Bras Dermatol 2001; 76:505-37.
12. Mehregan A, Pinkus H: Life history of organoid nevi. Special reference to nevus sebaceus of Jadassohn. Arch Dermatol 1965; 91:574-88.
13. Kim NH, Zell DS, Kolm I, Oliviero M, Rabinovitz HS: The dermoscopic differential diagnosis of yellow lobularlike structures. Arch Dermatol 2008; 144:962. 23.
14. Jaqueti G, Requena L, Sanchez-Yus E. Trichoblastoma is the most common neo- plasm developed in nevus sebaceus of Jadassohn. Am J Dermatopathol 2000; 22:108-18.
15. Ioannides G, Simonson L. Nodular hidradenoma in nevus sebaceus of Jadas- sohn. Arch Dermatol 1964; 89:250-52.
16. Campbell JP, Solomon AR, Woo TY. Apocrine cystadenoma arising in a nevus sebaceus of Jadassohn. Cutis 1984; 34:510-12.
17. Miller CJ, Ioffreda MD, Billingsley EM. Sebaceous carcinoma, basal cell carci- noma, trichoadenoma,

trichoblastoma, and syringocystadenomapapilliferum arising within a nevus sebaceus. Dermatol Surg 2004; 30:1546-49.
18. Miyake H, Hara H, Shimojima H, Suzuki H. Follicular hybrid cyst (trichilemmal cyst and pilomatricoma) arising within a nevus sebaceus. Am J Dermatopathol 2004; 26:390-93.
19. Ujiie H, Kato N, Natsuga K, Tomita Y. Keratoacanthoma developing on nevus sebaceous in a child. J Am Acad Dermatol 2007; 56 (2 Suppl.):S57-58.
20. Rahbari H, Mehregan A. Development of proliferating trichilemmal cyst in an organoid nevus. J Am Acad Dermatol 1986; 14:123-26.
21. Goldstein GD, Whitaker DC, Argenyi ZB et al. Basal cell carcinoma arising in a sebaceous nevus during childhood. J Am Acad Dermatol 1988; 18:429-30.
22. Westfried M, Mikhail GR. Multifocal basal cell carcinomas in a nevus sebaceus of Jadassohn. J Dermatol Surg Oncol 1981; 7:420-22.
23. Ball EA, Hussain M, Moss AL. Squamous cell carcinoma and basal cell carcinoma arising in a naevus sebaceous of Jadassohn: case report and literature review. Clin Exp Dermatol 2005; 30:259-60.
24. Winer LH, Levin GH. Pigmented basal cell carcinoma in verrucous nevi. Arch Dermatol 1961; 83:960-64.
25. Dalle S, Skowron F, Balme B, Perrot H. Apocrine carcinoma developed in nevus sebaceus of Jadassohn. Eur J Dermatol 2003; 13:487-89.
26. Tarkhan II, Domingo J. Metastasizing eccrine porocarcinoma developing in a sebaceous nevus of Jadassohn: report of a case. Arch Dermatol 1985; 121:413-15.
27. Abe S, Yamamoto Y, Uno S et al. Malignant melanoma arising in a sebaceousnevus of the scalp. Br J Plast Surg 2003; 56:171-73.
28. Premalata CS, Kumar RV, Malathi M et al. Cutaneous leiomyosarcoma, tricho- blastoma, and syringocystadenomapapilliferum arising from nevus sebaceus. Int J Dermatol 2007; 46:306-08.
29. Domingo J, Helwig EB. Malignant neoplasms associated with nevus sebaceus of Jadassohn. J Am Acad Dermatol 1979; 1:545-56.
30. Weschler HL, Fisher ER. A combined polymorphic epidermal and adnexal tumor in nevus uniuslateris. Dermatologica 1965; 130:158-64.
31. Cribier B, Scrivener Y, Grosshans E. Tumors arising in nevus sebaceus: a study of 596 cases. J Am Acad Dermatol 2000; 42:263-68.
32. Matsuda K, Doi T, Kosaka H et al. Sebaceous carcinoma arising in nevus seba- ceus. J Dermatol 2005; 32:641-44.
33. Misago N, Narisawa Y. Tricholemmal carcinoma in continuity with trichoblas- toma within nevus sebaceus. Am J Dermatopathol 2002; 24:149-55.
34. 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; 14:658-60.
35. Ashinoff R. Linear nevus sebaceus of Jadassohn treated with the carbon dioxide laser. Pediatr Dermatol 1993; 10:189-91.
36. Dierickx CC, Goldenhersh M, Dwyer P et al. Photodynamic therapy for nevus sebaceus with topical δ-aminolevulinic acid. Arch Dermatol 1999; 135:637-40.