Brief Report

Clinicopathologic analysis of 21 cases of nevus sebaceus: A retrospective study

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ABSTRACT

Background: Nevus sebaceus (NS), otherwise designated as 'organoid nevus', involves proliferative changes of the sebaceous glands, sweat glands, and the hair follicles. It displays a range of appearances, depending on the lesion's age. **Aims:** To study the histopathological features of NS and correlate these with clinical findings. **Methods:** All skin biopsy specimens over a 12-year period from 1995 to 2007 which had a diagnosis of NS were included. Clinical data with follow-up notes and histopathology were reviewed. **Results:** Half of the cases had a verrucous clinical appearance, while the rest presented as papules, plaques, or patches. All the cases showed immature hair follicles, and 24% of cases showed immature sebaceous glands. Normal terminal hair follicles were characteristically absent in the lesion. Nineteen percent of the cases showed dilated apocrine glands, and 14% showed hyperplasia of eccrine glands. Epidermal changes in the form of acanthosis, papillomatosis, and hyperkeratosis were seen in 86% of cases. Dilated keratin-filled infundibula were observed in 24% of cases. One case was associated with a squamous cell carcinoma. **Conclusions:** Nevus sebaceus is a cutaneous hamartoma, consisting of various elements indigenous to the organ. Normal terminal hair follicles are characteristically absent in the lesion although the same may be seen in rest of the epidermis, a feature of diagnostic importance, not usually highlighted in literature. The divergent differentiation observed in NS is consistent with the common embryologic origin of the folliculosebaceous-apocrine unit and should not mislead the pathologist.

Key Words: Differentiation, Hamartoma, Nevus sebaceus

INTRODUCTION

In 1895, Jadassohn first described nevus sebaceus (NS), a circumscribed hamartomatous lesion predominantly composed of sebaceous glands.[1] However, Pinkus designated this disease 'organoid nevus', as the changes are not confined only to the sebaceous glands but also involve proliferative changes of sweat glands and hair follicles.[2]

The lesion is interesting because its morphologic appearances vary with the age of the lesion.[3] As a consequence, there is a great likelihood of missing the diagnosis if only the classical findings of a well-developed NS are looked for. It is also associated with a range of secondary tumors, both benign and malignant. The incidence of these tumors increases with age, particularly after puberty. They include benign adnexal tumors such as trichoblastoma, syringocystadenoma papilliferum, trichilemmoma, sebaceoma, nodular hidradenoma, hidrocystoma, and eccrine poroma. Malignant cutaneous neoplasms are less commonly seen and include basal cell carcinoma and, to a lesser extent, squamous cell carcinoma, trichilemmal carcinoma, sebaceous carcinoma, porocarcinoma, and apocrine carcinoma.[4,5] In India, barring case reports and studies on treatment, there are no data documenting the clinicopathologic spectrum of NS, which prompted us to undertake this retrospective clinicopathologic study.[6,7]

METHODS

All skin biopsy specimens received at the Department of Pathology over a 12-year period from July 1995 to July 2007 which had a histologic diagnosis of nevus sebaceus were

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included. The clinical details, treatment modalities, and follow-up data pertaining to these patients were retrieved from the medical records of the hospital. The slides were reviewed by 3 pathologists independently.

RESULTS

A total of 21 patients with NS were included. Their age ranged from 8 years to 68 years. Maximum prevalence was seen in those aged between 21 and 30 years. Among the 21 patients, 11 were females and 10 were males. The location of the lesions is depicted in Table 1, scalp being the commonest site. Most (90%) of the cases were solitary. Forty-eight percent presented as verrucous plaques [Table 2].

Microscopy

The microscopic findings of the 21 cases are displayed in Table 3. All of them showed abortive/immature hair follicles characterized by cords of undifferentiated, basaloid cells resembling the embryonic stages of hair follicle. Germ and papilla formation was seen [Figure 1]. Twenty-four percent of cases showed immature sebaceous glands characterized by cells with larger nuclei, eosinophilic cytoplasm, and indistinct cell borders. The sebaceous glands also showed hyperplasia, crowding. Nineteen percent of the cases showed dilated, prominent apocrine glands, and 14% showed prominent, dilated eccrine glands. Epidermal changes in the form of acanthosis, papillomatosis, and hyperkeratosis were seen in 86%. Dilated keratin-filled infundibula were seen in 24% of the cases. One of the cases showed squamous cell carcinoma of the overlying epidermis; 2 lesions showed associated keratinous cysts.

The most striking feature was the absence of normal terminal hair follicles within the confines of the lesion, sharply demarcated from the adjacent skin, which was seen in all 21 cases [Figure 2].

Treatment and follow-up

All the lesions were treated by local excision. Follow-up data was available for 15 cases, none of which showed recurrence. The mean follow-up period was 3 years. The only patient with associated squamous cell carcinoma underwent wide excision and is currently disease-free.

DISCUSSION

Nevus sebaceus is usually present at birth; and in its most common location, viz., the scalp, it is manifested by a well-defined area of alopecia with smooth surface and yellow-to-orange discoloration.[8,9] In the infantile stage, the epidermis is only slightly acanthotic and may be hyperpigmented.[7] The hair follicles are small, incompletely formed, and are often represented by solid cords of undifferentiated basaloid cells. The sebaceous glands are not prominent.

The second stage in the life history of the organoid nevus occurs during adolescence and is characterized by an increase in the thickness of the lesion, which may then show smooth surface nodularities or verrucous hyperkeratosis.[8,10] Most of the patients in the current study presented during adolescence or early adulthood. At this stage, histologic findings include verrucous epidermal hyperplasia, which was the commonest clinical appearance in this study. This overlaps

Table 1: Site of lesions in nevus sebaceus

Site of the lesion	No of cases (%)
Scalp	12 (57)
Cheek	3 (14)
Neck	1 (5)
Nose	1 (5)
Eyebrow	1 (5)
Unknown	3 (14)
Total	21 (100)

Table 2: Clinical appearance of lesions of nevus sebaceus

Appearance of the lesion	No of cases (%)
Verrucous	10 (48)
Papule	4 (19)
Plaque	1 (5)
Patch	2 (10)
Filiform	1(5)
Unknown	3 (14)
Total	21 (100)

Table 3: Histologic features in nevus sebaceus

Microscopy	No. of cases (%) (Total= 21)
Immature/abortive hair follicles	21(100)
Immature sebaceous glands	5 (24)
Hyperplastic eccrine glands	3 (14)
Hyperplastic apocrine glands	4 (19)
Basaloid epidermal proliferation	5 (24)
Papillomatous epidermal hyperplasia	18 (86)
Absence of normal terminal hair follicles	21 (100)
Dilated infundibula	5 (24)
Inverted hair follicles	1 (5)
Inflammation	6 (29)
Calcification	2 (10)
Associated squamous cell carcinoma	1 (5)
Associated epidermal cyst	2 (10)

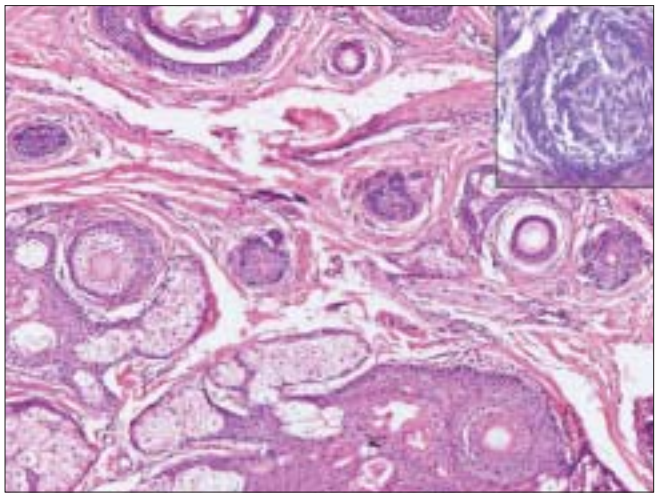

Figure 1: Nevus sebaceus showing immature hair follicles and hyperplastic sebaceous lobules. Immature hair germ in the inset (H and E, ×100; H and E, ×400)

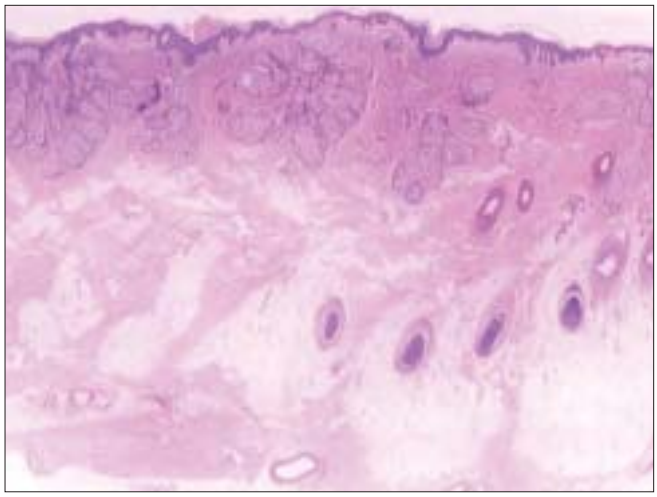

Figure 2: Nevus sebaceus showing absence of terminal hair follicles in the lesion (H and E, ×20)

with the histologic picture of epidermal nevus and can be differentiated from it by the presence of malformations of the dermis, most prominently hyperplasia and malpositioning of the sebaceous glands.[11] The hair follicles remain small and primordial. The sebaceous glands are now numerous and hyperplastic. Many of the lesions may also exhibit apocrine glands with dilated lumina as well as hyperplasia of eccrine glands, features seen in 19% and 14% of cases respectively in this series. These divergent features are in keeping with a common embryological origin of the pilosebaceous-apocrine unit, and any of them may predominate over the other. This should not mislead the pathologist.

One of the characteristic findings observed in the current series was the absence of normal terminal hair follicles within the lesion although the same may be seen in the rest of the epidermis, a feature of diagnostic importance described by Ackerman but not usually highlighted in literature. This feature was present in all our 21 cases and serves as a vital clue, particularly on scanner view. In our opinion, this feature may also be useful to assess if the biopsy is representative.

The third stage occurs during the adult life when epidermal hyperplasia, large sebaceous glands, and ectopic apocrine glands dominate the histologic findings. The hair follicles remain primordial.[12] The third stage is also distinguished by development of a variety of benign and malignant adnexal tumors. Benign tumors are seen in about 13.6%, while malignancies are met with in 1% of the cases of NS.[5] Trichoblastoma and syringocystadenoma papilliferum are the commonest benign tumors encountered in NS.[8] Malignant cutaneous neoplasms are less common, inclusive of basal cell carcinoma and, to a lesser extent, squamous cell carcinoma.[3,4]

In conclusion, NS is an organoid nevus, i.e., a hamartoma consisting of various elements indigenous to the organ and not merely the sebaceous units. Normal terminal hair follicles are characteristically absent in the lesion although the same may be seen in rest of the epidermis, a feature of diagnostic importance, not usually highlighted in literature. Though malignancy is uncommon, a cautious histologic analysis is mandated, especially if there are clinical changes in a lesion.

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