Dermoscopic patterns of nevus sebaceous – captured at different stages of evolution

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Abstract

The nevus sebaceous (NS) is a hamartoma of the skin and its adnexa pertaining to the group of organoid nevi. NS presents at birth and grows proportionally with the growth of the child. During adulthood, secondary neoplasms may arise from NS. Though the diagnosis of NS is mainly clinical, dermoscopy can be used as a diagnostic aid. Dermoscopy characteristically shows yellow globules, white, grey lobules arranged in clusters in a yellow background in a NS during childhood. Dermoscopic assessment will assist in diagnosis; and also, in assessing and monitoring the evolution of NS. It is also helpful in detecting malignant transformation of NS early, to arrange timely surgical excision in dermatological practice especially in resource poor setting.

Key words: nevus sebaceous, dermoscopy, secondary neoplasms

Introduction

The nevus sebaceous (NS) is a hamartoma of the skin and its adnexa pertaining to the group of organoid nevi¹. It was first described by a world-renowned German dermatologist, Josef Jadassohn in 1895. NS contains cells of epithelial and non-epithelial origin, characterized by epidermal, follicular, sebaceous, and apocrine gland abnormalities². It is a congenital lesion occurring in 0.3% of neonates and the incidence is equal between male and female. Lesions commonly occur on the scalp and face, but the involvement of limbs and chest also have been reported in the literature. Though the lesions are often solitary, multiple lesions can occur, sometimes forming extensive plaques along the lines of Blaschko. Scalp lesion may present as a patch of alopecia which can mimic aplasia cutis or cicatricial alopecia at birth.

NS presents at birth and grows proportionally with the growth of the child. Initially present as solitary smooth yellow well-circumscribed patch or plaque with oval/linear configuration. During puberty, lesions grow faster and become more prominent with verrucous or nodular appearance of the surface, secondary to the hormonal effects. During adulthood, secondary neoplasms such as trichoblastoma, syringocystadenoma papilliferum, trichilemmoma,

sebaceous adenoma, apocrine adenoma and basal cell carcinoma can arise in NS².

Etiology of NS is unknown. Though NS is congenital, it is not an inherited condition. It is thought to be due to mosaic postzygotic mutation. In some patients, an abnormality of the PTCH gene has been identified. NS may also present as a feature of certain genetic syndromes including nevus sebaceous syndrome.

Diagnosis of NS is mainly clinical. Patients present at their different ages with different growth stages of NS. In case of clinical suspicion, biopsy and histopathological diagnosis may be needed to confirm the diagnosis.

Dermoscopy which is a non-invasive bedside imaging technique for diagnosing various skin lesions, can be used in case of diagnosis of NS as it may clinically mimic other conditions like cicatricial alopecia, filiform wart, and neoplastic overgrowth.

Herein, we present 4 cases of NS that presented at different stages of growth from birth to adulthood; and describe their dermoscopic features.

Case 1

A two-day old neonate was found to have a single linear patch of alopecia over the frontal scalp during postnatal assessment. On examination, a well-defined linear patch of alopecia with yellowish hue was noted (Figure 1). Rest of the clinical examination was normal. Dermoscopic examination of the lesion revealed prominent yellow background with few yellow aggregated globules in the lesion (Figure 2).

Case 2

A one-year-old baby girl presented with a linear yellowish thin plaque over her left preauricular region with the history of slow increase in size since birth (Figure 3). She had no other extra cutaneous manifestations. Dermoscopically yellowish globules aggregated in clusters observed with yellow-orange background (Figure 4).

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Figure 1. Linear circumscribed patch of alopecia with yellowish hue.

Figure 2. Prominent yellow background with few yellow aggregated globules in the lesion.

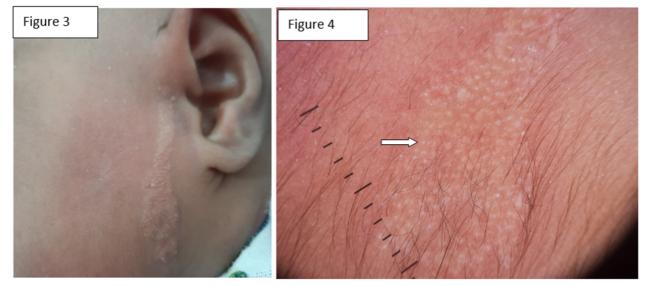


Figure 3. Linear yellowish thin plaque over her left preauricular region.

Figure 4. Yellowish globules (\rightarrow) aggregated in clusters observed with yellow-orange background.

Case 3

A 8-year-old girl presented with a progressively enlarging asymptomatic linear lesion over her forehead, which was there since birth. A linear yellowish thin plaque with cobblestone like surface was found on clinical examination (Figure 5). Dermoscopy showed brown to white lobules admixed with yellow globules arranged in a cluster on a yellow background over the lesion (Figure 6).

Case 4

A 22-year-old male presented with a rapidly enlarging lesion over his birthmark on his scalp for last 6-month duration. On clinical examination, greyish verrucous lesion was found over a yellow plaque with cobblestone surface on vertex (Figure 7). On dermoscopic examination, grayish papillary and filiform appearance was observed with yellow–white clusters of lobules and globules with few arborescent vascularization (Figure 8). But there was no blue–grey ovoid nest, cystic appearance or surface ulceration noted dermoscopically.

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Figure 5. Linear circumscribed yellowish thin plaque with cobblestone like surface over forehead.

Figure 6. Brown (\star) to white (Δ) lobules admixed with yellow globules (\Diamond) arranged in a cluster on a yellow background.



Figure 7. Greyish verrucous lesion over the yellow plaque with cobblestone surface on vertex scalp.



Figure 8. Greyish papillary and filiform appearance (()) noted with yellow-white lobules and globules clusters and few arborescent vascularization (→).

We confirmed the clinical diagnosis of nevus sebaceous with dermoscopic findings in all cases. Out of the 4 cases, the patient with filiform and cobblestone lesions was referred for complete surgical excision, while others were followed up in the skin clinic with regular clinical and dermoscopic assessments.

Discussion

NS is a type of epidermal nevus predominantly with sebaceous hamartoma, presenting at birth. Natural history of NS is to evolve through 3 stages. In the infantile stage, the lesion presents as a characteristic yellowish patch of alopecia or thin plaque. Dermoscopy shows few yellowish globules with bright yellow background, representing underdeveloped sebaceous gland and hair follicles, that are histologically viewed¹.

During puberty, the growth of the lesion is accelerated; and the lesion becomes verrucous. So, dermoscopy exhibits yellow, white, and grayish lobules arranged in clusters over a yellow background during adolescent stage. These dermoscopic features represent the developing and mature sebaceous glands, sweat glands and hair follicles within the hamartoma^{1,3}.

The final stage is characterized by appearance of nodules and tumors that may occur during adulthood. At the adulthood stage, dermoscopic examination may show papillary overgrowth in addition to yellow, gray and white clusters. Importantly, arborescent vessels may also be observed, representing the beginning of secondary neoplastic overgrowth within the lesion. In the third/neoplastic stage, that is in the late adulthood, dermoscopic features of secondary neoplasm could be identified. Micro/macro ulcerations, prominent arborescent vessels with bleeding spots, yellowish homogenous nodules and blue - grey ovoid nests will correlate clinical and histopathological type of secondary neoplasm^{1,3-5}.

Though benign tumors are the most common neoplasms known to arise on NS, malignant degeneration can occur with a lifetime risk of 5-20%. For this reason, NS should be managed with surgery before or during puberty. As most of the parents are anxious and concerned about the surgical excision of the child's birthmark during early childhood and permanent scarring, monitoring the NS with clinical

and dermoscopic assessments would be the best follow up practice.

Though dermoscopy was initially intended for the assessment of pigmented lesions, in current dermatology practice usage of dermoscopy has revolutionized diagnostic accuracy of skin lesions with diverse pathology, especially in a resource poor setting, where histological diagnosis not possible.

Dermoscopy is a useful tool in differentiating NS at its different stages of growth, from other clinical differential diagnosis. Dermoscopy may also give a clue about the type of neoplastic transformation, for example blue-white ovoid nest over the lesion suggestive of basal cell carcinoma.

Although histopathology of the lesion gives definite diagnosis of NS and its secondary neoplastic transformation, dermoscopic assessment will help the dermatologist to be more confident to make diagnosis clinically.

Conclusion

In this case series, we describe the dermoscopic pattern analysis of NS at its different stages of growth evolution. Dermoscopic assessment will assist in diagnosis; and assessing and monitoring the evolution of NS; and also, in detecting malignant transformation to arrange timely surgical excisions.

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