

Dermoscopy of nevus sebaceus: A cross-sectional study of 22 cases

Dear Editor,

Nevus sebaceus of Jadassohn (NS) is a cutaneous hamartoma classically evolving from an alopecic yellowish patch during infantile stage to verrucous in adulthood with yellow lobular appearance, and finally to nodular or tumoral appearance with peripheral telangiectasias. Multiple tumors, both benign and malignant, such as trichoblastoma, syringocystadenoma papilleferum and basal cell carcinoma may develop secondarily over nevus sebaceus . Benign tumours and malignant growth are seen in 13.6% and 1–2.5% of nevus sebaceus cases, respectively. The risk of malignant transformation increases with age. Hence, regular follow-up is needed for early detection of neoplasms. Dermoscopy, a non-invasive modality, can be utilised for its early diagnosis and follow-up. We aim to describe different dermoscopic patterns of nevus sebaceus according to its evolutionary stages.

We conducted a cross-sectional study from Jun 2019 to May 2021 in an Indian tertiary care center. A total of 22 patients with clinico-histopathological diagnosis of nevus sebaceus at various evolutionary stages were enrolled in the study, and the mean age was 27.14 years (range: 3-65 years). There was male preponderance (M:F = 18:4). Nevus sebaceus was located on scalp in 15 (68.1%) and on face in 7 (31.8%) patients. Alopecic patch was present in two (9%) patients, slightly elevated plaque in nine (40.9%), verrucous plaque in eight (36.3%) and verrucous plaque with nodules in three (13.6%). All patients underwent histopathology using 4 mm punch. It was consistent with nevus sebaceus alone in 19 (86.3%) patients, while features suggestive syringocystadenoma papilleferum were reported in 3 (13.8%) patients. It showed three patterns (1) Patch stage revealed normal epidermis with numerous sebaceous glands which were hyperplastic and small primordial hair follicles [Figure 1a]; (2) Plaque stage showed papillomatosis and acanthosis with sebaceous gland hyperplasia, ectopic apocrine glands and primordial hair follicles [Figure 1b]; and (3) nevus sebaceus with syringocystadenoma papilleferum revealed papillomatosis with several cystic invaginations extending into the dermis, forming small cystic areas lined by double layer of cuboidal to columnar epithelial cells with pale eosinophilic cytoplasm. Some of these cells showed decapitation secretion. The stroma contained a dense mononuclear cell infiltrate, comprising predominantly of plasma cells. Hyperplastic sebaceous glands were also noted. [Figure 1c].

Dermoscopic features are shown in Tables 1 and 2. Patch stage (n = 2) showed yellow globules (YG) arranged in clusters on a yellowish to pinkish yellow background [Figure 2a]. Slightly elevated plaque (n = 9) showed two patterns: (1) brown globules (BG) in crateriform pattern in six (66.6%) patients [Figure 2b] and (2) brown and orange globules (OG) in cobblestone pattern in three (33.3%) patients, which was suggestive of transition phase [Figure 2c]. Verrucous plaque with histology of nevus sebaceus alone (n = 8) showed three patterns: (1) only brown globules in cerebriform pattern in two (25%) patients [Figure 3a]; (2) brown globules in cerebriform pattern along with greyish to white exophytic papillary projections (EPP) at places, dotted to polymorphous vessels and yellow to white scales among three (37.5%) patients and crusted erosions among two (25%) patients [Figure 3b]; and (3) brown globules in cerebriform pattern with homogenous yellow globules in three (37.5%) patients with morphology of verrucous plaques with nodules [Figure 3c]. Verrucous plaque of nevus sebaceus in all these eight patients showed evolution from crateriform to cerebriform pattern and to homogenous yellow globules from periphery towards the centre. [Figure 3d].

All three patients with histopathological diagnosis of nevus sebaceus with syringocystadenoma papilleferum showed predominantly greyish white exophytic papillary projections with dotted, linear or polymorphous vessels, erosions or crust and scales (white or yellow) with brown globules in cerebriform pattern at places [Figures 4a and 4b].

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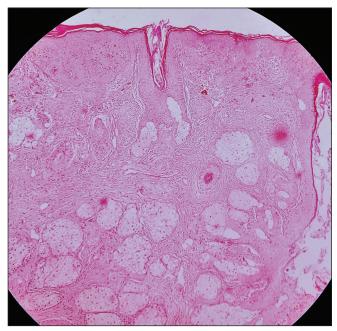


Figure 1a: Patch stage of nevus sebaceus showing normal epidermis with numerous, hyperplastic sebaceous glands and small, primordial hair follicles (H & E, 40x)

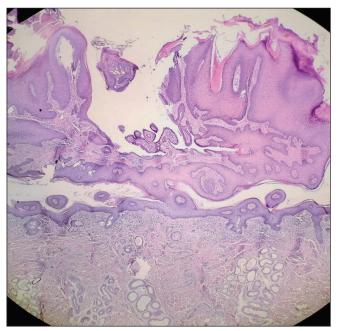


Figure 1c: Histopathology of nevus sebaceus with syringocystadenoma papilleferum showing papillomatosis with several cystic invaginations forming small cystic areas lined by double layer of cuboidal to columnar epithelial cells. Few decapitation secretions seen. Dense mononuclear cell infiltrate in the papillary dermis and numerous ectopic apocrine glands are seen (H & E, 40x)

Discussion

Clinically, it is difficult to differentiate nevus sebaceus from aplasia cutis congenita (ACC) in an infant. The infantile stage of this condition is characterised by clustered yellow globules on a yellow background or bright yellow dots not associated with hair follicles. Dermoscopy in aplasia cutis congenita

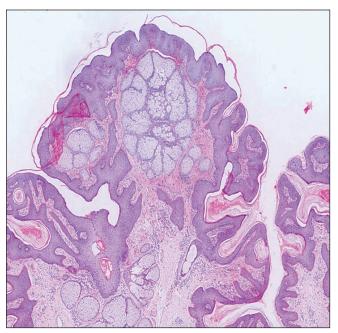


Figure 1b: Plaque stage of nevus sebaceus revealing papillomatosis and epithelial hyperplasia with sebaceous gland hyperplasia, ectopic apocrine glands and primordial hair follicles (H & E, 40x)

reveals absence of follicular ostia at the centre with radially arranged hair follicles at hair bearing margin forming a ring of hypertrichosis or starburst pattern with dark pigmented ends of these elongated hair bulbs visible through the translucent epidermis. Some telangiectasia are visible over the patch.⁴

In the childhood stage, lesions evolve from patches to slightly elevated plaques. Yellow globules in cobblestone pattern are seen corresponding to numerous hyperplastic sebaceous glands.^{3,5} However, we observed unique orange–brown globules in cobblestone pattern suggestive of transition phase in slightly elevated plaques in three (33.3%) patients and brown globules in crateriform pattern in six (66.6%) patients in the elevated plaque lesions. Orange globules represent immature sebaceous glands, while brown globules correspond to hyperplastic, mature sebaceous glands.

In the adult stage with verrucous plaque to nodular lesions, brown gobules in cerebriform pattern with fissures and ridges to crateriform pattern with peripheral striations to whitish yellow lobular or yellow-greyish papillary structures, in verrucous plaque and homogenous yellow appearance in nodular lesions with linear irregular and arborescent vessels have been described previously.3,5 We observed brown gobules from crateriform pattern to cerebriform pattern in all eight (100%) patients with verrucous plaque of nevus sebaceus alone from periphery towards centre, and evolved yellow nodules at centre suggestive of maximum epithelial hyperplasia and papillomatosis in HPE at centre and decreasing towards periphery of lesion. This shows maximum evolution of nevus sebaceus at the centre with peripheral dispersion. We did not find any telangiectasia in these nodules.

Table 1: Clinical and dermoscopic features observed in nevus sebaceus (n = 22)

Case number	Age (years)	Sex	Location	Clinical Morphology	Histopathological diagnosis	Dermoscopic features
1	32	M	Face	Plaque	NS	BG in cerebriform pattern at plaques. YG aggregated in clusters over yellow background at slightly elevated plaque
2	13	F	Temporal	Plaque	NS	BG with central crater pattern
3	55	M	Vertex	Patch	NS	YG arranged in clusters on a yellow background
4	22	F	Parietal	Plaque	NS	BG with central crater
5	7	M	Frontal	Plaque	NS	Orange globules and BG in cobblestone pattern (Transition phase), Greyish EPP at places, yellow white scales, linear vessels
6	65	M	Vertex	Verrucous plaque	NS with SCAP	BG at places, EPP, crust, erosion, dotted vessels, yellow white scales
7	24	M	Vertex	Alopecic Plaque	NS	BG and orange globules in cerebriform pattern (Transition phase)
8	24	M	Vertex	Patch	NS	YG aggregated on a pinkish yellow background
9	18	M	Vertex	Verrucous plaque	NS with SCAP	BG at places, EPP, dotted and polymorphous vessels, erosions, white scales
10	24	M	Forehead	Verrucous Plaque	NS	BG in cerebriform pattern, whitish EPP, dotted and linear vessels, white scales
11	25	M	Temporal	Plaque with nodules	NS	BG in cerebriform pattern, yellow homogenous structures
12	52	M	Face	Verrucous plaque	NS	BG in cerebriform pattern, erosion, crust, polymorphic vessels, white scales
13	50	M	Temporal	Plaque	NS	Brown yellow globules with central crater
14	26	M	Vertex	Verrucous plaque	NS with SCAP	BG at places, pinkish yellow EPP, linear vessels, yellow scales, crust
15	16	F	Face	Slightly elevated plaque	NS	BG in crater pattern
16	26	M	Temporal	Nodules to verrucous plaque	NS	Yellow homogenous globules to BG in cerebriform pattern, dotted vessel
17	24	M	Preauricular	Plaque with nodules	NS	Brown-yellow globules in cerebriform pattern, yellow homogenous structures
18	14	M	Face	Verrucous plaque	NS	Grayish EPP, BG in cerebriform pattern
19	26	M	Parietal	Verrucous plaque	NS	BG in cerebriform pattern and crater pattern with peripheral striations, erosion (edge)
20	26	M	Vertex	Verrucous plaque	NS	EPP, linear vessels, yellow scales, BG in cerebriform pattern
21	25	F	Vertex	Plaque	NS	BG and orange globules in cerebriform pattern (Transition phase)
22	3	M	Face (cheek)	Plaque	NS	BG with central crateriform pattern

BG: Brown globules; EPP: Exophytic papillary projections; F: Female; M: Male; NS: Nevus sebaceus; SCAP: Syringocystadenoma papilleferum; YG: Yellow globules

Table 2: Dermoscopic features of nevus sebaceus according to evolutionary stage (n = 22)							
Morphology of NS	Histopathological diagnosis	Number of cases	Dermoscopic finding with frequency (in numbers)				
Patch	NS	2	YG arranged in clusters on a yellow or pinkish yellow background (2)				
Plaque	NS	9	BG with central crater pattern (6) BG and orange globules in cobblestone pattern: Transition phase (3)				
Verrucous plaque	NS	5	BG in cerebriform pattern (5) EPP [at places] (3) Dotted or polymorphous vessels [at places] (3) Scales [white or yellow at places] (3) Erosions or crust [at places] (2)				
	NS with SCAP	3	BG at places (3) Predominant EPP (3) Dotted, linear or polymorphous vessels (3) Erosions or crust (3) Scales (white or yellow) (3)				
Verrucous plaque with nodules	NS	3	Yellow homogenous globules or structures (3) BG in cerebriform pattern (3)				

BG: Brown globules; YG: Yellow globules; EPP: Exophytic papillary projections; NS: Nevus sebaceus; SCAP: Syringocystadenoma papilleferum

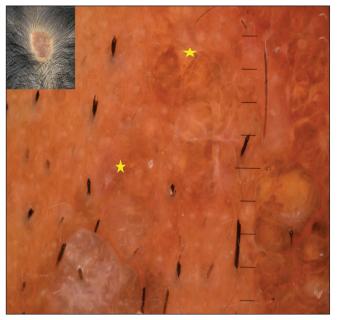


Figure 2a: Yellow globules (yellow stars) arranged in clusters over a yellow background seen in dermoscopy of patch stage of nevus sebaceus (Dermlite DL4, 10X polarised). [Inset: Clinical alopecic patch in childhood stage of NS over vertex of scalp in an adult]

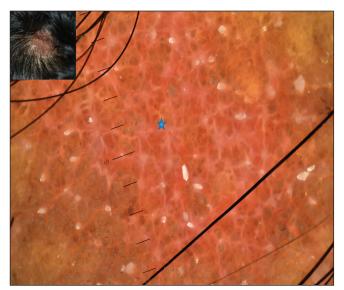


Figure 2c: Dermoscopy showing brown and orange globules in cobblestone pattern (blue star) suggestive of transition phase (Dermlite DL4, 10X polarised). [Inset: Elevated hyperpigmented plaque over vertex of scalp in an adult]

Secondary syringocystadenoma papilleferum associated with nevus sebaceus is seen in 5.2–13% cases.² We encountered three (13.6%) such cases. Dermoscopic features of all 3 cases were predominantly greyish white exophytic papillary projections with dotted, linear or polymorphous vessels, erosions or crust, white or yellow scales with brown globules in cerebriform pattern at places. Hence, replacement of majority of brown or yellow globules to exophytic papillary projections with vessels, erosions, crusts and scales suggested evolution



Figure 2b: Brown globules in crateriform pattern (yellow arrows) seen in dermoscopy of slightly elevated plaque (Dermlite DL4, 10X polarised). [Inset: Slightly elevated plaque located over temporo-parietal region of scalp]

of nevus sebaceus to syringocystadenoma papilleferum which was confirmed histopathologically. In a previous study, syringocystadenoma papilleferum was the second commonest benign neoplasia occurring in nevus sebaceus in 9 out of 23 (15.6%) cases. Dermoscopic features described were symmetric erythematous lesion with "exophytic papillary structures" (78% of cases; 7/9), central depression (66.7%; 6/9), erosions or crusts or ulceration (89%; 8/9) and vascular structures in 100% of cases. 6 Central depression and ulceration in dermoscopy of syringocystadenoma papilleferum may correspond to invaginating cystic spaces opening to skin surface on histopathology. Papillomatosis and papillary projections into cystic space on histopathology represent exophytic papillary projections, while presence of vessels on dermoscopy might be explained by fibrovascular core supporting these projections. Dermoscopy of the entire lesion should be performed to look for development of secondary syringocystadenoma papilleferum within nevus sebaceus.

Conclusion

This is an analysis of the dermoscopic features of nevus sebaceus as per the evolutionary stages in 22 cases. Though, a correlation between dermoscopy and histopathology is always advocated, the above dermoscopic features can guide the clinician towards a diagnosis and its progression to neoplasm without undergoing invasive biopsy or excision especially over the face.

Limitations

The study is limited by small sample size and unavailability of tumours other than syringocystadenoma papilleferum in analysis.

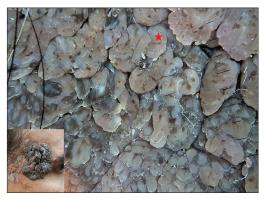


Figure 3a: Dermoscopy of an evolving verrucous plaque alone showing brown globules in cerebriform pattern (red star) (Dermlite DL4, 10X polarised). [Inset: Verrucous plaque in adulthood stage of NS]

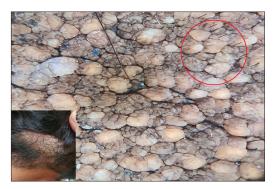


Figure 3c: Dermoscopy of verrucous plaque with nodules showing predominant yellow homogenous globules (red circle) along with brown globules (blue star) in cerebriform pattern (Dermlite DL4, 10X polarised). [Inset: Plaque with verrucous surface and yellow nodules over temporal region of scalp]

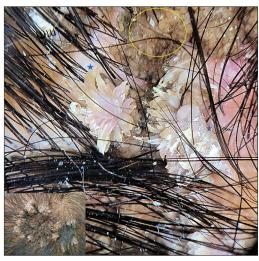


Figure 4a: Dermoscopy of nevus sebaceus with syringocystadenoma papilleferum depicting predominantly greyish white exophytic papillary projections (blue stars) with linear and polymorphous vessels and white scales with brown globules (yellow circle) in cerebriform pattern at places (Dermlite DL4, 10X polarised). [Inset: Plaque over vertex with verrucous surface and papillary projections]



Figure 3b: Dermoscopy of an evolving verrucous plaque showing brown globules in cerebriform pattern (yellow star) along with at places, greyish to white exophytic papillary projections with linear vessels and white scales (yellow circles) (Dermlite DL4, 10X polarised). [Inset: Hyperpigmented verrucous plaque over forehead]

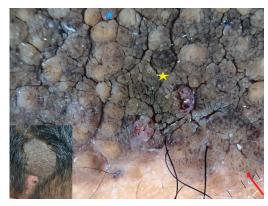


Figure 3d: Dermoscopy of an evolved verrucous plaque showing crateriform pattern (red arrow) at periphery to cerebriform pattern (yellow star) to yellow homogenous globules (blue star) towards centre of NS (Dermlite DL4, 10X polarised). [Inset: Verrucous plaque over temporal region of scalp]



Figure 4b: Dermoscopy of nevus sebaceus with syringocystadenoma papilleferum showing greyish exophytic papillary projections with polymorphous vessels and white scales, predominant erosions, crust and yellow white scales (yellow star) (Dermlite DL4, 10X polarised). [Inset: Verrucous plaque of nevus sebaceus over vertex]

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The patients or guardians of minors enrolled in this study have given written informed consent for the publication of their case details.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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Clinical characteristics of 81 patients with maculopapular cutaneous mastocytosis: A 10-year experience

Dear Editor,

Cutaneous mastocytosis (CM) is characterised by abnormal dermal accumulation of mast cells. Maculopapular CM (MPCM) is the commonest clinical type and is subdivided into pigmented variant (urticaria pigmentosa, UP) and telangiectatic variant (telangiectasia macularis eruptiva perstans, TMEP). This study reviewed all patients with histologically confirmed diagnosis of MPCM between 2009 and 2022 and characterised its detailed clinical manifestations. UP diagnosis was based on typical clinical lesions and monomorphic mast cell infiltrate. TMEP was characterised by characteristic telangiectatic macules and increased mast cells at the periphery of dilated blood vessels in the upper dermis.

In total, 81 patients were analysed, with 54 males and 27 females (2:1). The demographics and clinical characteristics of MPCM have been summarised in Table 1. There were 70 patients diagnosed with UP and 11 patients of TMEP. The mean age (\pm SD) of disease onset in years was 18.04 ± 15.93 in UP and 38.41 ± 14.96 in TMEP. Twelve patients with UP had congenital lesions. Most TMEP lesions developed between 31 and 50 years (8/11, 72.7%). None of our patients reported a positive family history or had any extracutaneous disease.

TMEP presented with multiple discrete red to brown telangiectatic macules [Figure 1a]. On the contrary, UP displayed a heterogeneous appearance in the morphology and arrangement of lesions as multiple yellow-orange to

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