Dermoscopic Analysis of Nevus Sebaceus of Jadassohn: A Study of 13 Cases

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Keywords

Nevus sebaceus of Jadassohn · Analysis · Dermoscopy · Evolutionary stages

Abstract

Background: Nevus sebaceus of Jadassohn (NSJ) is a complex cutaneous hamartoma with various clinical appearances. **Aim:** To describe different dermoscopic patterns of this nevus according to its evolutionary stages. **Methods:** This was an analytical study of NSJ carried out in the Department of Dermatology at the University Hospital Hassan II, Fez, Morocco. **Results:** Thirteen patients with a NSJ were enrolled in the study. Elevated verrucous plaques were noticed in 9 patients, homogeneous tumors were described in 2 NSJ, an alopecic patch was found in 1 case, and we had 1 case of malignant transformation into basal cell carcinoma. The dermoscopic aspects of NSJ described in our study were yellowish or brown globules aggregated in clusters on a yellow background. This pattern was significantly related to the first stage of an alopecic patch ($p = 0.001$). Whitish-yellow lobular aspect and grayish papillary appearance were significantly related to verrucous plaques ($p = 0.003$). Homogeneous yellow-whitish pattern was related to nodules ($p = 0.005$). **Con-**

clusion: The establishment of specific dermoscopic features of NSJ according to its evolutionary stages is important for its diagnosis and especially its monitoring in order to detect malignant transformation.

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Introduction

Nevus sebaceus of Jadassohn (NSJ), or organoid nevus, is a complex cutaneous hamartoma [1], involving not only the pilosebaceous follicle, but also the epidermis and often other adnexal structures [2, 3]. It may present at birth (0.3% of neonates [4]) or develop in early childhood. It occurs most frequently on the scalp (59.3%), less often on the face, the preauricular area, or the neck, and can rarely be identified in other locations [5] such as the trunk or the oral or genital mucosa [6, 7].

NSJ appears clinically as a smooth yellowish hairless patch [3–8]. At puberty, lesions acquire a verrucous appearance; in late adult life, they have a well-documented neoplastic potential [9]. The risk of a malignant transformation increases with age [10, 11].

Table 1. Clinical characteristics of our patients

Patient No.	Age, years	Gender	Clinical aspects	Pathology performed?
1	1	M	alopecic yellow plaque in the parietal area	no
2	12	F	elevated yellow frontoparietal alopecic plaque	yes
3	2	M	verrucous yellow plaque of the forehead	no
4	26	M	elevated brown-yellow plaque in the temporal area	yes
5	39	F	verrucous yellow-grayish occipital plaque	yes
6	30	F	verrucous yellow plaque of the vertex	yes
7	40	F	homogeneous yellow frontal tumor	yes
8	38	M	homogeneous yellow temporal tumor	yes
9	29	F	verrucous yellow parietal plaque	yes
10	32	M	verrucous yellow parietal plaque	yes
11	20	F	verrucous yellow parieto-occipital plaque	yes
12	27	F	verrucous yellow parieto-occipital plaque	yes
13	43	F	blue ulcerated tumor on a yellow verrucous frontal plaque	yes

Fig. 1. Malignant transformation of a papillary yellowish nevus sebaceus of Jadassohn to basal cell carcinoma (aggregated asymmetrical ovoid nets with central ulceration).

Dermoscopy is a noninvasive imaging technique useful for diagnosing a number of skin lesions through the in vivo observation of distinct structures that are considered highly disease specific [12]. Its use in the adnexal affections such as NSJ has not been well described even though it may help in the diagnosis and monitoring of lesions.

The aim of our study was to describe and analyze different dermoscopic patterns of this nevus according to its different evolutionary stages.

Subjects and Methods

Study Design

An observational study was carried out in the Department of Dermatology at the University Hospital Hassan II, Fez, Morocco during a 6-month period. Patients with NSJ were enrolled in the study at different stages of the disease. This NSJ was either the reason for a consultation (8 cases) or it was discovered at the physical examination of patients with other skin diseases (5 cases).

Histopathology was performed in all adult patients. Clinical and dermoscopic images of high quality were taken in order to be analyzed by 2 examiners, and they were then classified for future monitoring. Dermoscopic images were documented with a digital camera (DermLite, Fotofinder) with and without polarized light and with or without immersion. No pressure was used in order to avoid collapse of the vessels.

Statistical Analysis

Descriptive and univariate analysis using the SPSS 20 software were performed. In the univariate analysis, the χ^2 test was used to compare percentages in order to determine the dermoscopic patterns significantly related to each clinical form of NSJ. A p value <0.05 was considered statistically significant.

Fig. 2. Yellowish aggregated globules on a yellow background (alopecic patch in a 1-year-old infant).

Fig. 3. Yellowish globules aggregated in clusters on a yellow background with a papillary appearance in some spots (elevated verrucous partially alopecic frontoparietal plaque of nevus sebaceus of Jadassohn).

Results

Our study included 13 patients with a NSJ at different stages of the disease. The average age of the patients was 26 years (range 1–43 years). We had a female predominance (8 females, 5 males) and 1 familial case of NSJ.

The hamartoma was located on the scalp in 11 patients and on the forehead in 2 cases. Elevated verrucous plaques were noticed in 9 patients, homogeneous tumors were described in 2 NSJ, an alopecic patch was found in 1 case, and we had 1 case of malignant transformation into basal cell carcinoma in a 43-year-old adult (Fig. 1). There was no correlation between age and evolutionary stage (Table 1). Histopathology was carried out in 11 patients and was not performed in the 2 infants (Table 1).

The dermoscopic patterns described in this study were yellowish or brown globules aggregated in clusters on a yellow background (Fig. 2–5), grayish papillary appearance (Fig. 6), whitish-yellow lobular aspect (Fig. 7, 8), and homogeneous yellowish appearance (Fig. 9, 10) with or without vascularization (fine linear irregular or arborescent vessels) especially at the periphery (Table 2). The pattern of yellowish globules aggregated in clusters was significantly related to the first stage of NSJ (a patch or a slightly elevated patch) ($p = 0.001$), the yellow or whitish lobular aspect and the yellow-grayish papillary appearance were dermoscopic patterns of elevated verrucous plaques ($p = 0.003$), and the homogeneous yellow-whitish pattern was described in the tumoral stage ($p = 0.005$). Vascularization (linear, irregular, or arborescent) was significantly noticed in nodules and elevated plaques ($p = 0.004$).

The histopathology aspects were also evaluated according to evolutionary stage (Table 3; Fig. 11).

Fig. 4. Yellowish globules aggregated in clusters (verrucous plaque on the forehead of a 2-year-old infant with arborescent vascularization [arrow]).

Color version available online

Fig. 5. Brown globules on a whitish-yellowish background with linear and arborescent vascularization (arrow).

Color version available online

Fig. 6. Yellow-grayish papillary appearance (elevated verrucous partially alopecic occipital plaque of nevus sebaceus of Jadassohn).

Color version available online

Fig. 7. Yellow-whitish lobular appearance with peripheral arborescent vascularization (elevated hillocky plaque of nevus sebaceus of Jadassohn on the forehead).

Fig. 8. Yellowish papillary appearance (elevated verrucous partially alopecic parieto-occipital plaque of nevus sebaceus of Jadassohn).

Discussion

NSJ is a relatively uncommon hamartoma [13] first described by Josef Jadassohn in 1895. It has an epithelial and an adnexal origin [14] and is a predominantly sebaceous hamartoma, with an estimated incidence of less than 1/100,000 [15]. Whereas NSJ is typically congenital and presents at birth, it regresses in childhood and grows during puberty [16], suggesting hormonal control.

Most cases of NSJ appear to be sporadic, and the disease was thought to be caused by sporadic genetic mutations until 1982, when several case reports suggested dominant genetic transmission as a mechanism for this nevus's family inheritance [17, 18]. A paradominant inheritance theory was also suggested [19]. In this pattern of transmission, individuals who are heterozygous for the underlying gene are phenotypically normal and pass on the mutated allele clinically undetected to subsequent generations [15]. In our study, we had 1 family case of NSJ in a girl and her aunt.

NSJ usually affects the face and the scalp [20]. The natural tendency of NSJ is to evolve through 3 stages that

were first described in 1965 by Mehregan and Pinkus [21]. In the infantile stage, the lesion presents as a characteristic bright quiescent 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 [22]. Light microscopy shows masses of hypertrophic sebaceous glands, with possible papillomatosis and hyperkeratosis of the overlying epidermis.

This classic evolution may not be found, as described before. Some publications reported the presence of verrucous plaques in neonates and infants [23], which was also the case of one of our patients.

In the third or the neoplastic stage, many observations confirmed the possibility of the development of secondary tumors, which usually occur in late adulthood. The clinical signs suggesting neoplastic transformation include rapid enlargement or development of a nodularity or an ulceration. Several different tumors of

Fig. 9. Yellowish homogeneous appearance with the presence of peripheral linear and arborescent fine vessels (small tumor of the frontal edge).

Fig. 10. Whitish-yellowish homogeneous appearance with the presence of peripheral linear and arborescent vessels (nodule of the parieto-occipital area).

Table 2. The dermoscopic features of nevus sebaceus of Jadassohn according to evolutionary stages

Dermoscopic features	Evolutionary stages of the disease	Number of cases	<i>p</i> value
Yellowish globules aggregated in clusters on a yellow background	slightly elevated patches and plaques	5	0.001
Brown globules	elevated plaques	1	0.06
Whitish-yellow lobular aspect	elevated verrucous plaques	1	0.003
Yellow-grayish papillary appearance	elevated verrucous plaques	5	0.003
Homogeneous yellowish	nodules or small tumors	3	0.005
Vascularization (linear irregular or arborescent vessels)	nodules (+++) and elevated plaques	6	0.004

Fig. 11. Nevus sebaceus of Jadassohn. **a, b** Verrucous plaque showing immature inflammatory infiltrate in the area between the papillary and the reticular dermis. **c, d** Homogeneous tumor showing proliferation of mature sebocytes in the papillary dermis.

Table 3. The histopathology aspects of nevus sebaceus of Jadassohn according to evolutionary stages

Clinical stages	Histopathology aspects
Slightly elevated and elevated plaques	Immature hair follicles Immature sebocytes especially at the periphery Deep localization in the dermis
Verrucous plaque or tumor	Immature and mature sebocytes Inflammatory infiltrate Localization between the reticular and papillary dermis
Homogeneous tumors	Mature sebocytes Superficial localization in the dermis

epidermal, adnexal, and mesenchymal origin are known to arise (one or more simultaneously [24–26]). Benign tumors [12, 27–32] are the most common neoplasms, including syringocystadenoma papilliferum, apocrine cystadenoma, trichoblastoma, trichilemmal cysts, and keratoacanthoma. Although less common, malignant degeneration [33–47] occurs, with a lifetime risk of 5–20%. The majority of these tumors are basal cell car-

cinomas, but squamous cell, sebaceous, and apocrine carcinomas have also been recognized [2, 13]. In our study, we had 1 case of malignant transformation into basal cell carcinoma.

For this reason, NSJ should be managed with surgery before the period of puberty, as was proposed in many reports [48, 49]. CO₂ lasers [50] enhance cosmesis, but can only reach the papillary dermis, therefore they are

not recommended because of the possibility of malignant degeneration of the remaining cells in the lower dermis [51].

Another possibility of managing NSJ is its clinical and dermoscopic monitoring, especially for those lesions of the face whose excision may leave permanent scars, but first the dermoscopic features of this nevus must be well described. This was the objective of our study, where we defined 5 specific patterns according to evolutionary stage.

For the first stage, we noticed yellowish globules aggregated in clusters on a yellow background, while in the second stage of elevated verrucous plaques, 4 dermoscopic features were described – whitish-yellow lobular or papillary structures, yellow-grayish papillary appearance, brown globules on a yellow background, and yellowish globules aggregated in clusters, which was the most frequent dermoscopic aspect in the first and the second stages of NSJ. In the final stage or the tumoral stage, we noticed a homogeneous yellowish aspect. Although NSJ was not among the vascularized tumors described by Argenziano et al. [52], we found linear irregular and arborescent vessels especially in the tumoral stage of this hamartoma.

To our knowledge, the identification of dermoscopic features according to the different evolutionary stages of NSJ has not been well described before. There are only a few case reports that pointed out the benefit of dermoscopy in NSJ. Those reports found similar aspects, such as the round-shaped structures of a yellowish-white color which may correspond to clusters of mature superficial sebaceous glands [53].

Dermoscopy is also useful in differentiating NSJ in the tumoral stage from other sebaceous affections such as sebaceous adenoma and sebaceous hyperplasia (crown peripheral vascularization on a yellow background and a central crater), as was discussed in the publication of Kim

et al. [22]. In these 2 differential diagnoses, there is no risk of malignant transformation, and the decision of excision is taken by the patient for aesthetic reasons, without the need for a close follow-up as in the case of NSJ.

Dermoscopy is also useful in detecting the type of tumor arising on an NSJ in case of malignant transformation. This was reported by Zaballos et al. [54], who found that the pattern composed of asymmetrical large blue-gray ovoid nests – as in our patient – was more common in basal cell carcinoma than in trichoblastoma.

Although the correlation between histopathology and dermoscopy is highly emphasized, these results help the clinician to be more confident in order to make the diagnosis clinically and dermoscopically, without invasive diagnostic tools such as pathology.

Conclusion

In our original observational study, we identified specific dermoscopic patterns of NSJ according to its evolutionary stages. This is important for its diagnosis and especially its monitoring in order to detect malignant transformation and to avoid unnecessary excisions, especially on the face, in addition to their use in differentiating this nevus from sebaceous hyperplasia or adenoma.

Statement of Ethics

All subjects were informed about the conditions related to the study and gave their consent for publication.

Disclosure Statement

The authors have no conflicts of interest to declare. There were no funding sources.

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