Clinical, Dermoscopy and Histopathological features of Nevus Lipomatosus Cutaneous Superficialis: A Multi-Center Case Series of 10 Patients

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Abstract

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous skin condition of unknown etiology. Clinically, NLCS can be classified into two clinical types: 1) Classical Hoffman-Zurhelle or the multiple type and 2) solitary pedunculated type. Histopathologically, hematoxylin-eosin would reveal ectopic mature adipose tissues interspersed with thickened collagen bundles in the dermis separate from the subcutaneous fat which is pathognomonic of NLCS. Although the clinical diagnosis of such condition is straightforward, sometimes it can be mistaken for other skin-colored pedunculated skin lesions. The authors hope that the result of the case series will guide dermatologists in differentiating NLCS from other other skin-colored pedunculated skin lesions. A good clinical eye together with histopathology remains to be the gold standard for the diagnosis of this skin condition. Excision remains to be one of the most effective treatments of choice with minimal recurrence. Other treatment modalities such as carbon dioxide laser excision and cryotherapy may also be offered.

Keywords: amartoma, nevus lipomatosus cutaneous superficialis, excision

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Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous skin condition of unknown etiology characterized by the appearance of ectopic mature adipocytes in the papillary or reticular dermis separate from the subcutaneous fat (1). Clinically, NLCS can be classified into two clinical types. Classical Hoffman-Zurhelle or the multiple type are characterized by multiple soft, skin-colored to yellowish papules or nodules coalescing to form plaques with smooth, wrinkled or peau d'orange appearance of surface. Classical lesions are usually present at birth or in the first two to three decades of life (2), distributed in linear, zonal or segmental fashion over the buttocks, lower back or upper thighs (3). However, lesions can also be found in areas such as the upper trunk, abdomen, axillae, genitalia or face (4). The solitary pedunculated type is characterized by papule, nodule or tumor with either "smooth" or "cerebriform" surface in a study by Baraldi et. Al (5). This type usually appears during the third to sixth decades of life and can appear in different locations of the body but has a predilection for the trunk (6). The exact pathogenesis of this rare skin condition is yet to be determined. In a study by Buch, fat deposition in NLCS may have been secondary to the degenerative changes in connective tissue. In 1937, Robinson and Ellis hypothesized that NLCS may be a true connective tissue nevus which resulted from the focal heterotopic development of adipose tissue (7). In 1975, Light microscopy studies by Jones et. Al. suggest dermal adipocytes of this condition originated from the pericytes of blood vessels during fetal lipogenesis (8). Further cytogenetic study looked into the genetic factor in the development of these lesions which revealed mosaicism for a 2p24 deletion (9).

NLCS is asymptomatic but cosmetically disfiguring. This condition has not been associated with tendency towards malignant changes but are associated with multiple cutaneous disorders such as the following: follicular papules, hypertrophic pilosebaceous units, angiokeratoma of Fordyce, café-au-lait macules, scattered leukoderma, and hemangioma (6).

Case Presentation

A total of 10 patients were included in this case series. All of them are females with Fitzpatrick skin phototype IV, with mean age of 42.4 ± 13.5 years old. All patients reported appearance of solitary pedunculated tumors of varying size and duration (Table 1). No other associated skin abnormalities were present at the time of consultation. All patients voluntary requested removal of lesions for cosmetic reasons. Informed consents on excision biopsy, photography and publications were secured.

In all cases, the authors performed dermoscopy using a manual polarized light device (Dermlite DL2x10; 3Gen, San Juan Capistrano, CA). Dermoscopic findings for cerebriform pattern with sulci and gyri, yellow structureless areas, white structureless areas, irregularly distributed linear loop-like or linear-coiled vessels (8/8; 100%). (Figure 2a). Dermoscopic findings for smooth surfaced revealed yellow structureless areas, white structureless areas, irregularly distributed linear loop-like or linear-coiled vessels. (2/2; 100%). (Figure 2b).

Shave excision prior to electrocautery or carbon dioxide laser were performed on all cases. Histopathologic findings of the cases revealed acanthosis, papillomatosis and absence of spongiosis in the epidermis. (Figure 3a). The dermis contained varying amounts of mature adipose interspersed with fibrous connective tissue septae and blood vessels. Thick and fibrillary brightly eosinophilic collagen in

Figure 1: Solitary brown-colored pedunculated tumor with cerebriform surface on 8 patients (8/10, 80%) and solitary skin-colored nodule with smooth surface measuring on 2 patients (2/10, 20%)

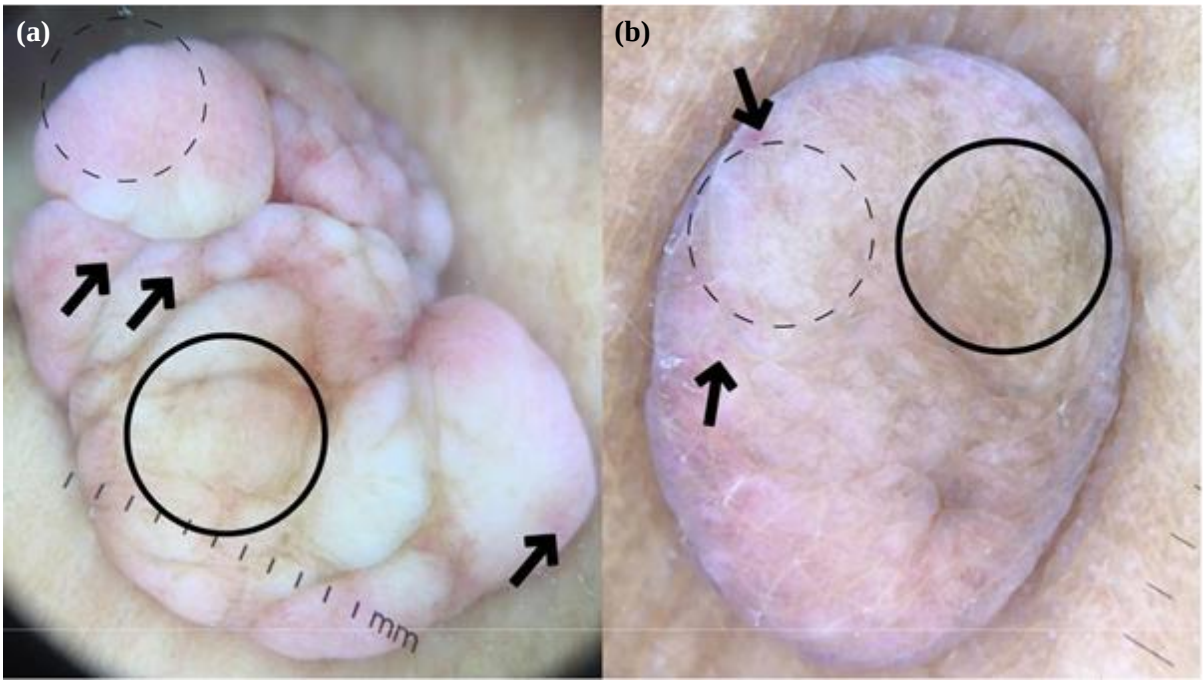

Figure 2: Representative photos of the dermoscopy of nevus lipomatosus cutaneous superficialis. Yellow structureless areas (black circle), white structureless areas (dotted circle) and irregularly distributed linear loop-like or linear-coiled vessels (black arrow). (a. Dermlite DL2x10 ; b. Dermlite DL2x10)

haphazard array with mild diffuse inflammatory infiltrate of lymphocytes were also noted. (Figure 3b). In the cases of nevus cutaneous lipomatosus cutaneous superficialis with a smooth surface, acanthosis and papillomatosis were absent as compared with the variability in the cerebriform type. There was also noted absence of mature adipose tissue in the superficial dermis fat in the case of NLCS with smooth surface as compared with the presence in all superficial, mid-dermis and deep fat in the cerebriform type. The group also noticed that the ectopic fatty tissue present in the superficial, mid and deep dermis is not connected with the fat of

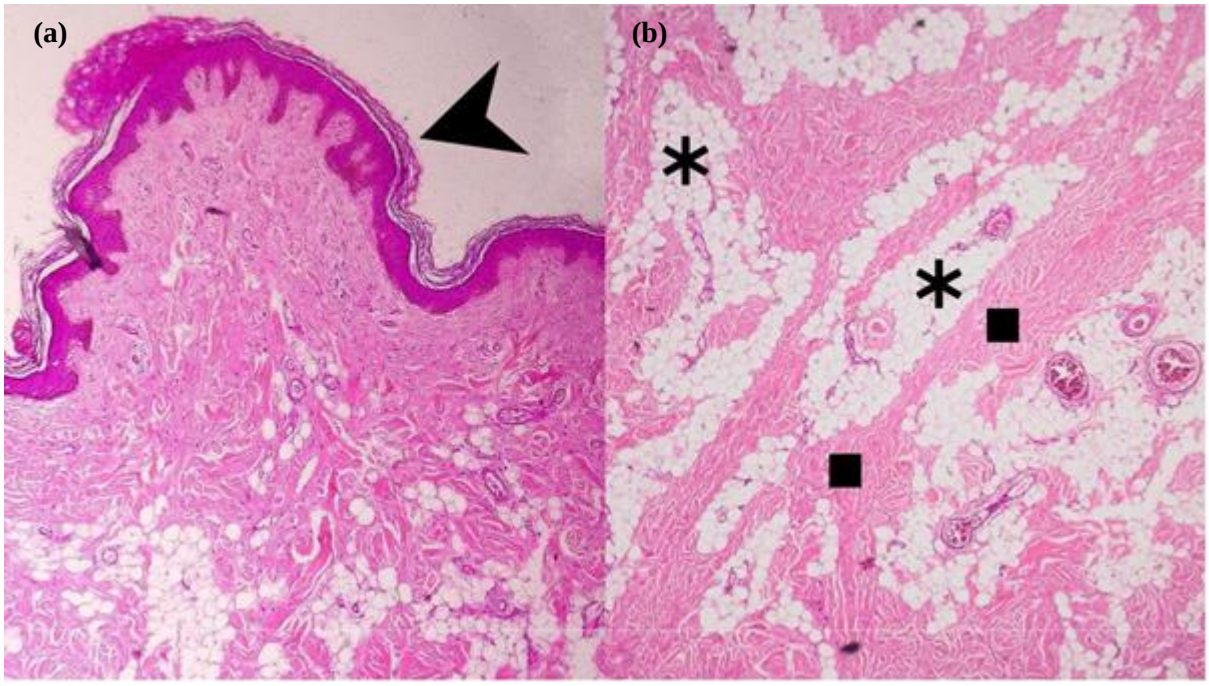

Figure 3: H&E shows acanthosis and papillomatosis (arrowhead), varying amounts of mature adipose interspersed with fibrous connective tissue septae and thick and fibrillary brightly eosinophilic collagen in haphazard array (black square) with mild diffuse inflammatory infiltrate of lymphocytes (a&b. H&E, 100x)

the underlying subcutaneous tissue. Summary of all the histopathological findings observed in our patients can be seen in Table 2.

Management And Outcome

In approaching patients with nevus lipomatosus cutaneous superficialis, reassurance that the condition is benign is very essential. In our patients, electrocautery or carbon dioxide laser excision was done in all cases and no recurrence were noted.

Discussion

NLCS appears clinically as a multiple or solitary skin-colored to yellowish papule, nodule or tumor with smooth or cerebriform surface. Dermatologists and other practitioners should be guided with the possibility of an NLCS diagnosis when evaluating a patient with an isolated, pedunculated skin-colored papule, nodule or tumor. In this case series, the group investigated the dermoscopic and histopathological correlation of NLCS in order to differentiate it from other skin-colored pedunculated lesions.

For the dermoscopic findings of the solitary type with cerebriform surface, our group found the appearance of sulci and gyri, yellowish structureless areas, white structureless areas, irregularly distributed linear loop-like or linear-coiled vessels on all eight cases. This is in contrast with the dermoscopic findings of the solitary type with smooth surface which revealed yellow structureless areas, white structureless areas, irregularly distributed linear loop-like or linear-coiled vessels on the two cases. The findings of the yellow and white structureless areas in our study were similar to the findings of Kinnera et.al. The yellowish structures correspond to the dermal adipocyte while the white structures correspond to the thickened collagen in the dermis. (10). Our findings were consistent with the dermoscopic features previously described by Vinay et al who

Table 1: Summary of Patients

Case No.	Sex	Age	Duration	Location	Clinical features	Treatment
1	M	40	5 years	Right gluteal area	solitary brown-colored pedunculated tumor with cerebriform surface measuring 2.5cm x 1.0cm	Excision
2	M	40	3 years	Right gluteal area	solitary flesh-colored pedunculated tumor with cerebriform surface measuring 0.5cm x 0.5cm	Excision
3	F	38	10 years	Left chest	solitary skin-colored pedunculated tumor with cerebriform surface measuring 1.0cm x 1.0cm	Excision
4	M	34	4 years	Right pelvic area	solitary skin-colored to brownish pedunculated tumor with cerebriform surface measuring 2.0cm x 2.0cm	Excision
5	M	28	5 years	Right gluteal area	solitary flesh-colored pedunculated tumor with cerebriform surface measuring 1.0cm x 1.0cm	Excision
6	M	56	35 years	Right shoulder	solitary skin-colored pedunculated tumor with cerebriform surface measuring 3.0xm x2.0cm	Excision
7	F	54	10 years	Right medial thigh	solitary skin-colored pedunculated tumor with cerebriform surface measuring 1.0cm x 1.0cm	Excision
8	M	64	2 years	Trunk	solitary skin-colored pedunculated tumor with cerebriform surface measuring 0.5cm x 0.5cm	Excision
9	F	20	4 years	Back	solitary skin-colored nodule with smooth surface measuring 0.5cm x 0.5cm	Excision
10	M	50	4 years	Left posterior thigh	solitary skin-colored nodule with smooth surface measuring 0.5cm x 0.5cm	Excision

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Table 2: Histopathological findings of nevus lipomatosus cutaneous superficialis

	Acanthosis	Papillomatosis	Spongiosis	Superficial Dermis fat	Mid-dermis fat	Deep dermis fat	Blood vessels	Infiltrate
Case 1	mild	present	absent	Present	present	present	present	mild
Case 2	moderate	present	absent	present	present	present	present	sparse
Case 3	absent	present	absent	present	present	present	few	sparse
Case 4	absent	present	absent	present	present	present	few	sparse
Case 5	mild	present	absent	present	present	present	few	sparse
Case 6	absent	present	absent	present	present	present	few	sparse
Case 7	absent	present	absent	present	present	present	few	sparse
Case 8	absent	present	absent	present	present	present	few	sparse
Case 9	absent	absent	absent	absent	present	present	present	sparse
Case 10	absent	absent	absent	absent	present	present	few	sparse

was able to describe five features of NLCS: cerebriform appearance, web-like regular pigment network, rim showing a white veil, yellowish structureless areas, and comedo-like openings. In addition to the yellowish and whitish structureless area seen in the dermoscopy, irregularly distributed linear loop-like or linear-coiled vessels were also observed in both the “cerebriform” and “smooth” type of solitary pedunculated NLCS. The presence of irregularly distributed linear loop-like or linear-coiled vessels correspond to the vascularity histopathologically. Our study was similar with the findings of Buch et al showing increased vascularity in the subpapillary and papillary dermis with perivascular with mononu clear cell (11).

In a study by Triki et al, they found out that the epidermis may show mild to moderate acanthosis, basket weave hyperkeratosis and focal elongation of rete ridges (12). The group were able to observe similar findings of acanthosis, papillomatosis and absence of spongiosis in the epidermis of our specimens. In all cases, the groups observed aggregates of mature adipose tissue embedded among the collagen bundles of the dermis were separated from the subcutanetous fat. This is similar in with the study of Ionnidou et al who stated that the most characteristic feature of NLCS is that there is usually no connection with the subcutaneous fat tissue (13). Our findings were also in line with the findings of Kinnera et al, that the adipose tissues typically form small aggregates around blood vessels or eccrine sweat glands and separate collagen bundles. (10) The adipocytes may extend to the papillary dermis (14). Avhad and Jerajani observed that the proportion of adipose tissues in the papillary and reticular dermis varies greatly and ranges from from 10% to 50% of the lesion. (15). In a study by Baraldi et al on the clinical, dermoscopic and histopathological features of solitary NLCS, they concluded that the histopathological features of

the solitary type nevus lipomatosus cutaneous superficialis are similar. They were able to conclude that the adipocytes are present both in reticular and papillary dermis in the cerebriform type and adipocytes are present only in the reticular dermis in the smooth-surfaced type. (5). This is quite similar with our findings, as the group noted absence of mature adipose tissue in the superficial dermis fat in the case of NLCS with smooth surface as compared with the presence in all superficial, mid-dermis and deep fat in the cerebriform type.

Due to the similarities between acrochordon, neurofibroma and nevus sebaceous clinically, histopathology still remains to be the gold standard of diagnosis. Acrochordon are usually less than 1 cm in size and with variable adipose tissue in the dermis of its larger variants. Neurofibroma would reveal proliferation of spindle shaped cells with wavy nuclei embedded in a myxoid matrix. Nevus sebaceous would reveal presence of adnexal structures (immature sebaceous gland, immature hair structures) and/or ectopic apocrine gland.

Treatment options for NLCS are mainly limited to excision with either electrosurgery, carbon dioxide laser or cryotherapy. Excision is curative as report of recurrence is rare post-excision (16). As any hamartomas, NLCS can gradually increase in size, causing apprehension among patients. The authors hope that the result of the case series will guide dermatologists and surgeons in differentiating NLCS from other skin-colored pedunculated skin lesions. Previously, cases of NLCS were misdiagnosed as acrochordons. In contrast with acrochordons however, NLCS is not associated with insulin resistance or metabolic disorders. Patient reassurance that this condition is benign and not a known marker for any other underlying conditions is essential. (17). A good clinical eye together with histopathology remains to be the gold standard for the diagnosis of this skin condition. Excision remains to be one of the most effective treatments of choice with minimal recurrence.

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