



# Nevus Lipomatosus Cutaneous Superficialis with a Histopathological Appearance Resembling Sclerotic Fibroma: A Rare Case Report

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## Abstract

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**BACKGROUND:** Nevus lipomatosus cutaneous superficialis (NLCS) of Hoffmann–Zurhelle is a benign idiopathic hamartoma. There are two types of NLCS; multiple and solitary. They are found in the abdomen, lower back, buttocks, hips, upper posterior thighs, and pelvis. The diagnosis can be evaluated with a typical histopathological of mature fat cells in the dermis, with 10–50% of the dermis.

**CASE REPORT:** We reported a case of NLCS with clinical papules and multiple nodules on the buttocks since the age of 6 years with a history of lipoma removal. The dermoscopic examination was conducted to confirm the diagnosis. The histopathological examination showed a dominant sclerotic fibroma (SF) with two sessions of biopsy and a few mature fats in the dermis after deeper cuts paraffin block. Cryotherapy with an open spray method is the treatment of choice in this patient.

**DISCUSSION:** The appearance of the dermis in NLCS can be normal or an increase in collagen. Interestingly, collagen has sclerosis partially and resembles SF never been reported. NLCS increases the amount of collagen; however, collagen as sclerosis remains obscure. The histopathological features of NLCS with other morphological abnormalities in the dermis have been reported, such as NLCS with perifollicular fibroma (PF) features. The SF features are other morphological abnormalities in NLCS, as reported in the PF.

## Introduction

Nevus lipomatosus cutaneous superficialis (NLCS) is first reported by Hoffmann and Zurhelle in 1921 as a benign idiopathic hamartoma [1]. It is a rare connective tissue nevus in children and young adults [2]. Clinically, there are two types of NLCS: Classic (Hoffman–Zurhelle) and solitary. Classic form presents as multiple, asymptomatic, soft, yellowish lesions that may coalesce to form a plaque, with or without comedones, and follicular holes. Solitary form, on the other hand, presents as isolated papules which are domed on a broader basis than skin tags [3].

Mature fat cells in the dermis are typical findings of NLCS. The number of mature fat cells in NLCS in the dermis varies from 10% to 50% [4], [5]. The dermis can be completely normal or filled with collagen. Besides that, an increase of fibroblasts and vascularity has also been reported [6]. However, no studies have ever been reported regarding the change of fibroblasts into sclerotic tissues seen in sclerotic fibroma (SF).

SF or circumscribed collagenoma or storiform collagenoma is a type of fibrous soft-tissue tumors.

Multiple SF is often associated with Cowden syndrome (CS) [7], with clinical manifestations of papules, round to oval nodules, skin-colored or pearly papules, or well-defined nodules. An increase in the number of atypical collagen and atypical fibroblasts is typical and is pathognomonic for SC [7], [8].

In addition, although NLCS is typically not associated with other malformations, Anzai *et al.* (2015) reported the coexistence of NLCS and perifollicular fibromas, a rare cutaneous hamartoma [9], [10]. In this study, we presented a rare case of NLCS with histopathological features resemble those of SF.

## Case Report

A 25-year-old woman presented to the Dermatology and Venereology outpatient Clinic of Wahidin Sudirohusodo Hospital, Makassar, South Sulawesi, Indonesia, with a chief complaint of multiple skin-colored papules and nodules on the lower back around the buttocks since she was 6 years old. Itching and pain were

denied. A medical history was significant for a presumed diagnosis of lipoma with no histopathological confirmation on the region which was completely excised. However, 3 months following the surgery, the patient noticed the emergence of small papules that eventually increased slowly in number and size.

The patient had sought medical treatment to her local physician and was treated with intralesional steroid injection for a diagnosis of keloid; however, no improvement was shown. No history of malignancies, such as breast, thyroid, and gastric cancer, was reported.

On physical examination, we found coalescing hyperpigmented immobile papules and nodules of various sizes with smooth surface. In addition, keloid with a size of 1.5 cm× 5 cm was also found (Figure 1).



Figure 1: Physical examination day 1 (control I) in the gluteus sinistra region: Papules, nodules, partially skin-colored hyperpigmentation, smooth surface, fixed, scale, and keloid

Dermoscopic examination (Heine, iC1, Germany) was conducted and showed gyri (ridges) and sulci (fissures) with keratinous debris and yellowish structureless areas accompanied by ulceration (Figure 2).

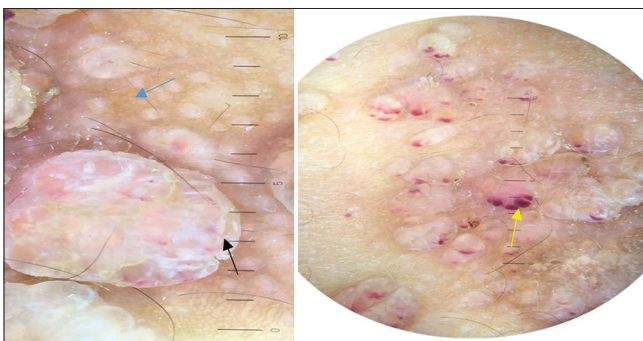


Figure 2: Dermoscopy features (Heine iC1) blue arrow: Yellowish structureless area; black arrows: Gyri and sulci, keratinous debris; yellow arrow: Ulceration keratinous debris; yellow arrow: Ulceration

Biopsy was conducted on a region that was previously excised and histopathological examination demonstrated normal epidermis and thick collagen connective tissue with spindle-shaped, non-cellular, and non-atypical nucleus with inflammatory cells in the upper dermis (Figure 3). These features were in accordance with SF.

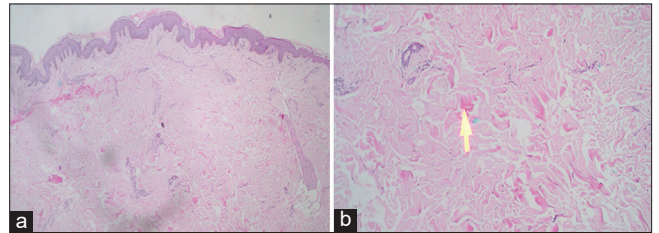


Figure 3: First histopathological examination (HE). (a) Epidermal tissue without any abnormalities ( $\times 4$ ), (b) very thick dermis with abundant subcutaneous collagen tissue ( $\times 40$ )

However, as SF with multiple lesions is known to be highly associated with CS, the absence of features suggestive of CS in this case prompted us to consider other diagnosis. In addition, the clinical presentation of this case was atypical for SF. Therefore, histopathological examination was reperformed in other site and deeper cuts were carried out.

The second histopathological examination showed hyperplastic and hyperkeratotic epidermis (Figure 4). A tumor mass consisting of thick collagen connective tissue with non-atypical spindle core was found in dermis. Perivascular lymphocytes and mild neutrophils were found in the upper dermis. The second histopathological examination was also consistent with SF. Because the clinical and dermoscopic images were pointing toward NLCS, we did deeper cuts in paraffin block. Furthermore, we found several mature fat tissues in the dermis, which were typical for NLCS.

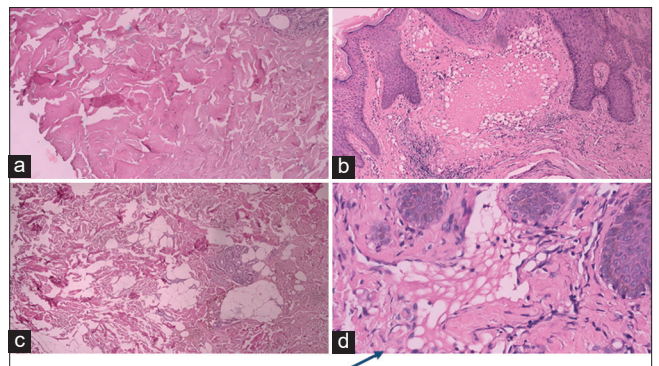


Figure 4: Histopathological examination with deeper cuts in paraffin blocks (HE). (a) Collagen is enlarged and sclerotic; (b) mature fat cells in the dermis, and (c and d) mature fat cells in the dermis surrounded by sclerotic thick collagen

The patient underwent two sessions of cryotherapy with an open spray method. The procedure was done with 1 cm from the skin, lesion margin of

1–2 mm, and maximum time spot of 30 s. One month later, the lesions showed significant improvement where the papules were markedly reduced in number and the nodules were no longer visible. The keloids also turned flat. No recurrence was found within 6 months of follow-up (Figure 5).

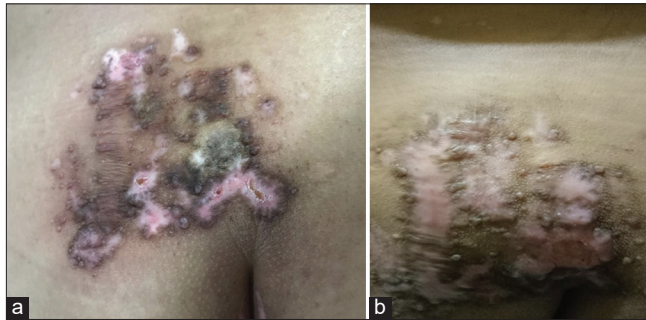


Figure 5: Clinical features (a) erythematous lesion after surgery. (b) a month after the second surgery.

## Discussion

NLCS is a rare benign hamartoma. There is no difference in prevalence in terms of sex, genes, or family [11]. The classic type, as in this case, typically presents as multiple skin-colored or yellowish papules with a tendency to coalesce and form a plaque. The surface of the cerebriform is segmentally distributed [2], [12] and is not accompanied by pain [1] NLCS has been reported to occur in people aged 3 months–35 years and is commonly reported on the buttocks and thighs with size varying from 1 cm to 10 cm × 7cm [11].

Comedo-like opening, cerebriform surface, honeycomb-like pigmentary network, yellowish holes, and rim of white veils are reported dermoscopic features of NLCS. Although no typical dermoscopic features of NLCS have been previously reported, the presence of a yellow structureless area representing dermal adipocytes is considered a specific marker of NLCS [12]. These features were also reported by Pardo *et al.* in his case report [13].

In this case report, the patient was initially diagnosed with SF based on histological features, where an increased amount of collagen with sclerotic changes was observed. SF is a benign fibroblastic tumor which is characterized by histiocytic differentiation, eosinophilic collagen, and inconspicuous nucleus [14]. Clinically, SF may present as solitary lesion in patients without CS or as multiple lesions in patients associated with CS [7]. The latter is an autosomal dominant disease characterized by mucocutaneous hamartoma and various extracutaneous benign and malignant tumors. In addition, CS is associated with an increases risk of neoplasms of the thyroid, mammary, endometrial, colorectal, and renal [1].

Both SF and NLCS are diagnoses that depend on histological features. However, although the first

histopathological descriptions were suggestive of SF, the clinical and dermoscopic features were very typical for NLCS. Thus, a second biopsy followed by histopathological examination was reperformed and also consistent with SF. After deeper cuts of the paraffin block were carried out, histopathology revealed the presence of dermal mature fat cells.

The characteristic histopathological finding in NLCS is mature fat cells in the dermis which may range from 10% to more than 50% [4]. In addition, an increase in collagen with abundant fibroblasts and increased vascularity may also be found [6]. An interesting finding in this study was that besides the typical finding of NLCS, an increased number of sclerotic collagen fibers with spindle-shaped, non-cellular, and non-atypical nucleus, which more closely resembled SF, was also observed, thus perplexing the diagnosis.

Although the presence of sclerotic collagen has been reported in NLCS [13], the coexistence of histopathological findings of SF and NLCS has never been reported.

Increased blood vessels in the papillary dermis [6] could explain the dermoscopic finding of ulceration in our patient. Epidermal changes as seen in this case have been described in literatures. However, epidermal changes, such as epidermal acanthosis, hyperkeratosis, and focal flattened or elongation of rete pegs, have also been reported [3], [4], [5], [11].

Although the presence of mature fat cells in the dermis is the typical and often the only histopathological abnormalities in NLCS, we managed to demonstrate a histopathological overlap between SF and NLCS in this case. To the best of our knowledge, only two studies have described dermal abnormalities other than the presence of dermal adipocytes in NLCS [9], [10]. However, different from our case, both studies reported the presence of folliculosebaceous components. Thus, to our knowledge, we were the first to report histopathological features of SF in a patient with clinical presentation suggestive of NLCS.

Therapy for NLCS is generally not necessary except for cosmetic reasons. Surgical therapy is an option with very low rate of recurrence. Patients who refuse surgery can choose cryotherapy with satisfactory results [11]. Cryotherapy is commonly used to treat many benign, premalignant, and malignant skin lesions by utilizing extreme cold to destroy abnormal cells and tissues [15], [16]. This technique causes cell death in four ways: Ice crystals formation inside cells that induced cellular damage, intracellular clot formation during freezing that leads to osmotic differences which, in turn, cause cell disruption, cold injury to small blood vessels causing ischemic damage, and immunological stimulation produced by the release of antigenic components resulting in cell damage [17].

The cryotherapy procedure in this case was conducted by open spray technique where the tissue

was frozen for 30 s and repeated twice with a margin of 2–3 mm. This procedure was performed twice every month. The treatment of NLCS by an open spray technique with a 2-week interval between therapies has previously been reported [18]. These findings indicated that cryotherapy is a therapy option with satisfactory results in patients with NLCS.

## Conclusion

We presented a rare case of NLCS resembling histopathological features of SF. This finding raises the possibility that NLCS may be related with other hamartomas. Furthermore, cryotherapy was shown to be effective in treating NLCS.

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