Nevus Lipomatosus Superficialis in a Serous Inflammatory Capsule: A Case Report

Nevus lipomatosus superficialis (NLS) is a rare benign hamartomatous lesion characterized by adipocyte infiltration into the dermal layer of the skin. Clinically, there are 2 types of NLS lesions: the classical type and the solitary type. The solitary type of NLS is significantly more common in women, typically presents after age 30 years, and occurs most often in the inner thighs. This lesion is often mistaken for similar cutaneous lesions, including but not limited to acrochordons, fibrolipoma, neurofibromatosis, and lymphangioma. There is limited information in the literature on unique presentations of this rare lesion.

Case Report: We present a case of a 26-year-old woman worried about a pedunculated papule in a fluid-filled capsule on her right inner thigh. She reported that the lesion doubled in size within 48 h prior to presentation. The lesion was biopsied in the clinic and pathology showed lobules of adipose cells in the dermis surrounded by collagen fibers and vascularity consistent with a diagnosis of inflamed NLS. Upon follow-up 2 weeks later, there were no signs of recurrence.

Conclusions: It was hypothesized that the unique presentation of a fluid-filled capsule surrounding a nevus lipomatosus superficialis occurred acutely following torsion of the pedunculated lesion. In addition to the patient’s history of frictional rub between the thighs, histologic signs of lymphocytic infiltration in the dermis and edematous stroma supported the claim of torsional origins. The unique presentation of NLS in a fluid-filled capsule is not often discussed in the literature, and we hope this report will aid providers in identifying such lesions in the clinic.

Keywords: Connective Tissue Nevus • Dermatology • Inflammation • Torsion, Mechanical
Background

Nevus lipomatosus superficialis (NLS) is a rare benign hamartomatous lesion characterized by lobules of adipocytes surrounded by collagen fibers within the dermis. Hoffman and Zurhelle first described this type of lesion in 1921. Clinically, there are 2 types of presentations of NLS: the classical type (also called Hoffman-Zurhelle) and the solitary type. The classical form is characterized by multiple soft cerebriform papules and nodules that coalesce into a plaque. The solitary type presents as a firm round mass often attached to the skin by a peduncle [1].

In general, the solitary type of NLS is significantly more common in women, typically presents after age 30, and occurs most often in the inner thighs [2]. Unusual presentations of NLS have been reported in the literature. This includes unique locations, foul-smelling discharge, and ulcerations [3,4]. However, most previously reported unusual presentations of NLS are in the classical type of NLS and are less frequently reported as part of the solitary form. We present a case of a solitary form of nevus lipomatosus superficialis lesion that developed a fluid-filled capsule over a period of 48 h. Histological analysis showed dermal inflammation and necrosis possibly secondary to torsion. This case had a unique visual presentation that is important to add to the information already known about NLS.

Case Report

A 26-year-old woman presented to the medical clinic for a pedunculated papule on her right inner thigh. She first noticed the thigh lesion 2 years before but attributed it to a small skin tag. She sought evaluation when the lesion nearly doubled in size and developed a serous fluid-filled sac in 48 h. On visual inspection, the inner thigh had an area of red tan wrinkled skin enclosed in a fluid-filled membranous cystic-like tan tissue measuring 1.5×1.5×1.0 cm (Figure 1). She denied any pain or paresthesia in the area. However, she reported discomfort from the frictional rub between her thighs while walking. These combined symptoms motivated her to be evaluated, and she requested excisional removal. A detailed skin physical exam was negative for other similar lesions. She denied any family history of similar lesions. An excisional biopsy was performed in the clinic. Pathology showed adipose cells embedded in the dermis surrounded by bundles of collagen fibers and increased vascularity, consistent with a diagnosis of NLS (Figures 2, 3). Upon follow-up 2 months later, the patient had full clinical resolution.
Discussion

Our case study highlights a unique case of a nevus lipomatosus superficialis in a 26-year-old woman. This case was particularly interesting due to the acute enlargement of the lesion within 48 h and its presentation in a fluid-filled capsule (Figure 1). The differential diagnosis that we considered included fibrolipoma, neurofibromatosis, lymphangioma, and acrochordon. Histopathology confirmed the final diagnosis. Under histologic analysis, the presence of adipose lobules in the dermis and surrounding collagen fibers was indicative of a diagnosis of NLS (Figures 2, 3) [5]. Furthermore, a proliferation of lymphocytes and edematous stroma (Figures 3, 4) revealed that an inflammatory process was likely the cause of the acute enlargement of the lesion. An inflammatory response was likely responsible for the rapid enlargement and serous fluid collection around the lesion, as seen by separating the epidermis from the dermis layer (Figure 2). Given that the patient endorsed friction rubbing at the lesion site with walking, the inflammatory process was likely due to torsion of the lesion. One previously reported case of inflamed and necrosed NLS in a neonate reported similar histological findings and had also attributed the presentation to torsion of the lesion [6].

The etiopathogenesis of NLS is not well understood. Hoffman and Zurhelle’s proposed pathogenesis of NLS included adipocytic metaplasia in the course of degenerative changes within the dermal tissue [1]. Given that NLS lesions are more likely to have increased vascularity on histopathologic analysis, some have postulated the development of mature adipocytes from primitive lipoblasts associated with mesenchymal perivascular cells [7]. Increased vascularity of the lesion may have also contributed to our patient’s acute and quick inflammatory response to torsion of NLS.

Conclusions

In summary, nevus lipomatosus superficialis is a rare benign skin lesion. In the setting of mechanical trauma, the lesion can grow exponentially and activate an inflammatory response. NLS lesions may be more susceptible to torsion than other benign lesions such as acrochordons because they are generally larger in size and present in locations prone to mechanical trauma such as the inner thighs and trunk. Inflammation of the lesion can present as a solid mass within a fluid-filled capsule, as demonstrated in our patient. This case report may be helpful for practitioners in recognizing a rare benign lesion with a unique clinical presentation.

Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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Figure 4. Histology shows lymphocytes dispersed across the stroma and surrounding vessels within the dermis (200×).