Elastic Tissue in Nevus Lipomatosus Cutaneous Superficialis

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Abstract: Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous condition characterized by the presence of mature ectopic adipocytes in the dermis. We studied twelve cases of solitary type of NLCS to demonstrate the status of elastic fibers. All were stained with elastic-van Gieson stain for elastic tissue. Most of the cases showed moderate to marked reduction and thinning of elastic fibers (EF) as compared to adjacent normal dermis. We conclude that thinned and decreased elastic tissue is the cause of NLCS.

Keywords: Nevus lipomatosus superficialis, elastic fibers.

INTRODUCTION

NLCS is a rare benign developmental disorder characterized by isolated ectopic adipose tissue in the dermis. It was first reported by Hoffman and Zurhelle in 1921. Clinically it is classified into two forms. The classical form is characterized by groups of multiple, soft, pedunculated papules, nodules, or plaques. The other form manifests as a solitary dome-shaped or sessile nodule/papule. The precise etiopathogenesis of NLCS is not yet understood.

MATERIAL AND METHODS

Twelve specimens, processed in histopathology lab at king Khalid university hospital, whose histopathologic diagnosis was solitary type NLCS were identified. Age, sex and locations of each case were assessed. These specimens were then re-examined after staining for elastic tissue with elastic-van Gieson stain. The number and thickness of elastic fibers were evaluated as normal, reduced or thinned. Normal adjacent skin was used as normal control.

RESULTS

The mean age of patients was 39 years (range 6–67 years). There were 6 females and 6 males. The lesions were located on thigh (n: 8), lower abdomen (n: 2), and shoulder (n: 2). The mean size of the lesions was 1.5 cm (range 1-3 cm) (Table **1**). By Hematoxylin and eosin (H/E) stain, all the cases showed polypoid lesion with adipose tissue located high in dermis which is the diagnostic feature for NLCS (Figure **1**). Elastic-van Gieson stain showed moderate to marked reduction and thinning of elastic fibers (EF) in most of the cases as compared to normal control skin (Figure **2**). Two cases revealed mild reduction and thinning of EF.

DISCUSSION

Nevus lipomatosus superficialis also known as nevus lipomatosus cutaneus superficialis of Hoffmann snd Zurhelle is a rare type of connective tissue lesion characterized by the presence of mature adipose tissue in the dermis. It is found as plaques or solitary lesions, or in an extremely rare generalized form. It was first reported by Hoffman and Zurhelle in 1921 [1].

Clinically, it is classified into the classical Hoffmann-Zurhelle form and the solitary form. The solitary form usually occurs after the age of 20 years, with no particular predilection sites. The classical form occurs at birth or during the first three decades of life. This form manifests with groups of multiple, soft, non-tender, pedunculated, cerebriform, yellowish or skin-coloured papules or nodules. They often coalesce to form a plaque lesion, usually situated on the pelvic girdle area in a zonal pattern [2, 3].

Histopathologically, NLCS usually shows a papillated epidermal surface associated with a dermal proliferation of mature adipocytes in the reticular dermis that may extend to the papillary dermis. The proportion of the dermal fat is variable, ranging from less than 10% of the dermis to over 50% .The adipocytes may show connections to the underlying subcutaneous fat or be separated from the subcutis by collagen [4]. The precise etiopathogenesis of NLCS is not yet understood. The proposed pathogenesis of NLCS includes adipose tissue metaplasia in dermal connective tissue, developmental displacement of adipose tissue; alternatively, the lesions could be explained by the possible origin of adipocytes from the pericytes of dermal vessels [5].

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Table 1:

CASE NUMBER	AGE	SEX	LOCATION	SIZE(CM)	ELASTIC FIBERS NUMBER	ELASTIC FIBERS THICKNESS
1	34	male	thigh	1.7	REDUCED ++	Thin++
2	40	male	thigh	1.5	REDUCED ++	Thin++
3	32	male	thigh	1.5	REDUCED ++	Thin++
4	26	male	thigh	2	REDUCED +++	Thin+++
5	63	male	neck	1.6	REDUCED ++	Thin++
6	6	female	thigh	2	REDUCED +++	Thin+++
7	67	female	Lower abdomen	1.5	REDUCED ++	Thin++
8	35	female	Shoulder	1.3	REDUCED +	Thin+
9	62	female	Shoulder	1	REDUCED +	Thin+
10	31	male	thigh	1.5	REDUCED ++	Thin++
11	30	female	thigh	3	REDUCED +++	Thin+++
12	44	male	Lower abdomen	2.5	REDUCED +++	Thin+++

Mild +, moderate++, severe+++.

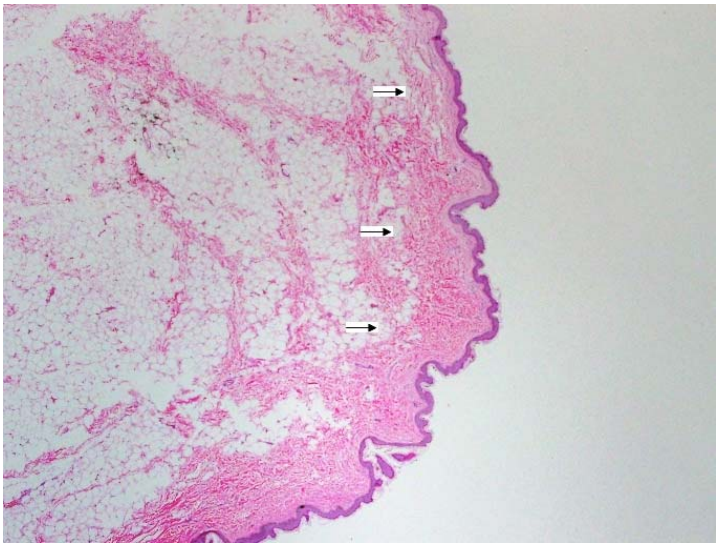
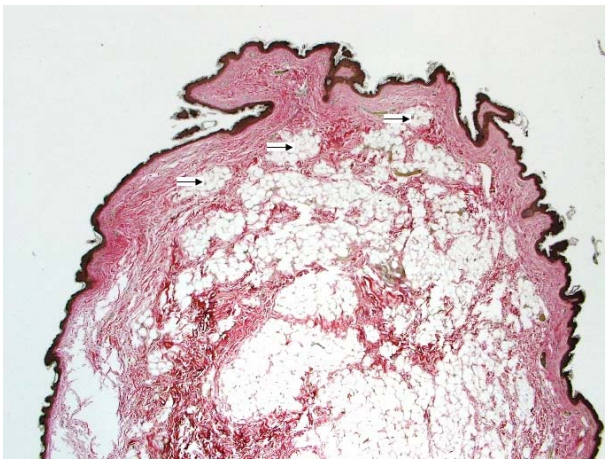
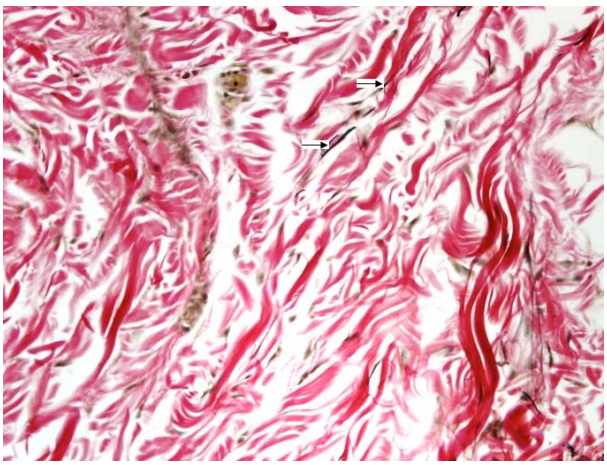


Figure 1: Low power of one case shows polypoid skin lesion with adipose tissue (arrows) located in the dermis. (H/E stain, x20).

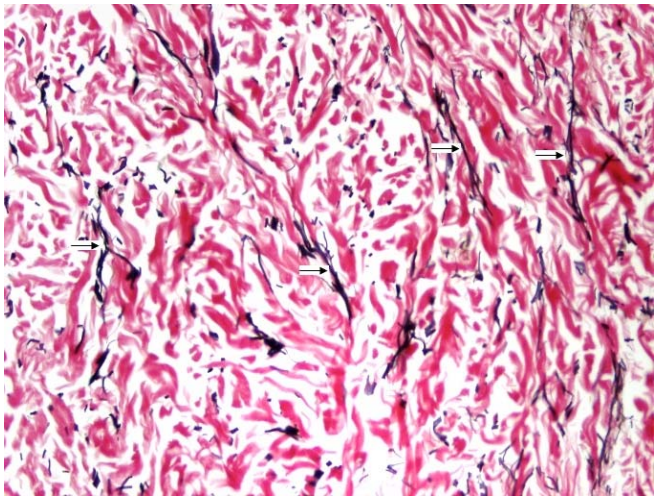


A



B

(Figure 2). Continued.



C

Figure 2: Low power of elastic Van Gieson stain shows marked reduction of elastic fibers and show adipose tissue in the dermis (arrows) (**A**) (EVG stain x20). High power shows few and thin elastic fibers (arrows) (**B**) (EVG stain x200) as compared to normal control dermis (**C**) (EVG stain x200).

Elastic fibres are essential extracellular matrix macromolecules comprising an elastin core surrounded by a mantle of fibrillin-rich microfibrils. They endow connective tissues such as blood vessels, lungs and skin with the critical properties of elasticity and resilience [6]. From our results, we found considerable abnormalities (reduction and thinning) of dermal elastic fibers in all the cases of NLCS examined. We also noticed that the larger the lesion, the more reduction and thinning of elastic fibers and vice versa.

We conclude that reduction and thinning of dermal elastic fibers is the cause of solitary type of NLCS that will lead to herniation of subcutaneous fat in the dermis and subsequent polypoid skin nodule.

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