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Unraveling the Rarity: A Case report of Nevus Lipomatosus Cutaneous Superficialis

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Abstract

Nevus Lipomatosus Cutaneous Superficialis, also known as dermolipoma, represents a rare benign hamartomatous skin tumor characterized by the deposition of mature adipose tissue within the dermal collagen matrix. It exists in two forms: solitary and multiple, with the latter being more prevalent, particularly on the lower back. Here, we present a case of a 21-year-old male from South India with non-congenital Nevus Lipomatosus Cutaneous Superficialis on his upper back, offering a comprehensive examination of its clinical, dermoscopic, and histopathological aspects. Remarkably, the patient had no underlying comorbidities and presented with a long-standing, asymptomatic, progressively enlarging skin lesion that had been present since childhood. Diagnostic confirmation was achieved through histopathological examination following a successful surgical excision, devoid of complications. Nevus Lipomatosus Cutaneous Superficialis is an infrequent benign condition that can be overlooked during childhood, emphasizing the significance of early recognition. Timely identification is paramount, as this condition can lead to substantial growth, introducing not only medical concerns but also notable aesthetic and self-esteem challenges for those affected.

Keywords: Nevus Lipomatosus Cutaneous Superficialis, dermolipoma, benign skin tumor, adipose tissue

Introduction

Nevus Lipomatosus Cutaneous Superficialis first documented by Hoffmann and Zurhelle in 1921, an infrequent benign hamartomatous skin tumor characterized by the accumulation of mature adipose tissue within the dermal collagen matrix[1] . This condition presents in two primary forms: solitary and multiple. Solitary lesions are more commonly observed in adults, often distributed more extensively across the skin[2]. In contrast, multiple Nevus Lipomatosus Cutaneous Superficialis lesions tend to manifest from birth to early adulthood, with a predilection for localization on the lower back, buttocks, or abdomen. Both types exhibit similar histopathological features, marked by varying proportions of adipose tissue (ranging from 10% to 50%) interspersed within collagen bundles in the dermis[1]. While the precise pathogenesis remains elusive, it is hypothesized to result from degenerative changes in connective tissues. Notably, Nevus Lipomatosus Cutaneous Superficialis, often referred to as a dermolipoma, does not display a gender predilection or familial inheritance and is not associated with congenital defects[3]. In this case report, we present an uncommon instance of this benign condition in an 21-year-old male, situated in the upper back, which underscores its non-congenital nature.

Case Report

A 21-year-old male, resident of south India with no comorbidities came to our OPD with asymptomatic elevated skin lesion over his upper back that was present since his childhood and had progressed steadily to attain its present size. There is no family history of similar skin problems.No history of itch,pain or discharge. On examination, a 5 * 7 cm well-defined lobulated non tender skin-colored to hyperpigmented plaque present over the left upper back (FIGURE 1).On palpation,it is soft to firm in consistency.There is no evidence of any oozing or discharge. Based on history and clinical examination, diagnosis of Nevus lipomatosus superficialis was identified.It was confirmed with dermoscopy and HPE. Dermoscopy (FIGURE 3) revealed the presence of sulci, keratotic gyri, scales noted inside the sulcus and reticular brown pigmented structure noted. Histopathology (FIGURE 2A) showed flaky hyperkeratosis, irregular acanthosis, and increased pigment seen in the basal layer. Dermis shows dilated blood vessels in papillary dermis . Groups and strands of mature fat cells seen around and within blood vessels (FIGURE 2B). There is no clear cut distinction between dermis and subcutis as it is replaced completely by fat cells.There are frequent foci of fibrosis intermixed with the fat tissue. The lesion was excised surgically and grafting was done.Patient was followed up for 2 months.No recurrence was observed.

Discussion

Nevus Lipomatosis Cutaneous Superficialis (NLCS) typically presents as multiple or solitary papules primarily on the lower trunk, buttocks, or upper thighs[3]. In our patient, the lesions had been present since childhood and have been progressively increasing in size over the years. The clinical presentation varies, but typical nodules in characteristic locations suggest the diagnosis. While biopsy remains the gold standard, MRI can aid diagnosis by revealing uniform skin thickening without irregular septa or abnormal contrast enhancement[3].The exact pathogenesis of NLCS remains unknown, but several proposed hypotheses provide insight. One theory suggests that dermal adipocytes may originate from pericytes[3]. Another hypothesis posits that NLCS may represent a connective tissue nevus. Additionally, the identification of deletion of 2p24 in NCS cases supports the potential role of genetic factors in the development of NLCS.NLCS is generally not associated with systemic findings, but pigment anomalies like café-au-lait spots and hypopigmented macules have been reported[4].Our 21-year-old patient's lesion, which first appeared in childhood, was located on the upper back, lacked ulceration or discharge but exhibited consistent pressure from clothing and progressive growth, warranting surgical excision. Malignant transformation and recurrences are exceptionally rare, though vigilant follow-up is essential[3]. NLCS is a rare connective tissue nevus affecting children and young adults, typically appearing as plaque-like solitary lesions, occasionally multiple, predominantly on the buttocks, lumbar back, and posterior thighs. Histologically, lesions consist of mature adipose tissue and connective tissue components, often associated with thickened collagen bundles, increased elastic fibers, and heightened fibroblast presence in deeper dermal layers[2]. Key differential diagnoses include nevus sebaceous, plexiform neurofibroma, focal dermal hypoplasia, connective tissue nevus, angiolipoma, and lipomatosis. The preferred treatment is surgical excision, generally curative. NLCS lesions are typically nonprogressive, with no systemic or malignant associations, making this case report valuable due to its rarity.

Conclusion

Nevus Lipomatosus Cutaneous Superficialis represents an infrequent benign anomaly often misdiagnosed in childhood. Early recognition is imperative as it may attain substantial size, giving rise to not only medical apprehensions but also posing aesthetic and self-esteem challenges to affected individuals.

Reference:

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Fig. 1. A well-defined lobulated non tender skin-colored to hyperpigmented plaque present over the left upper back

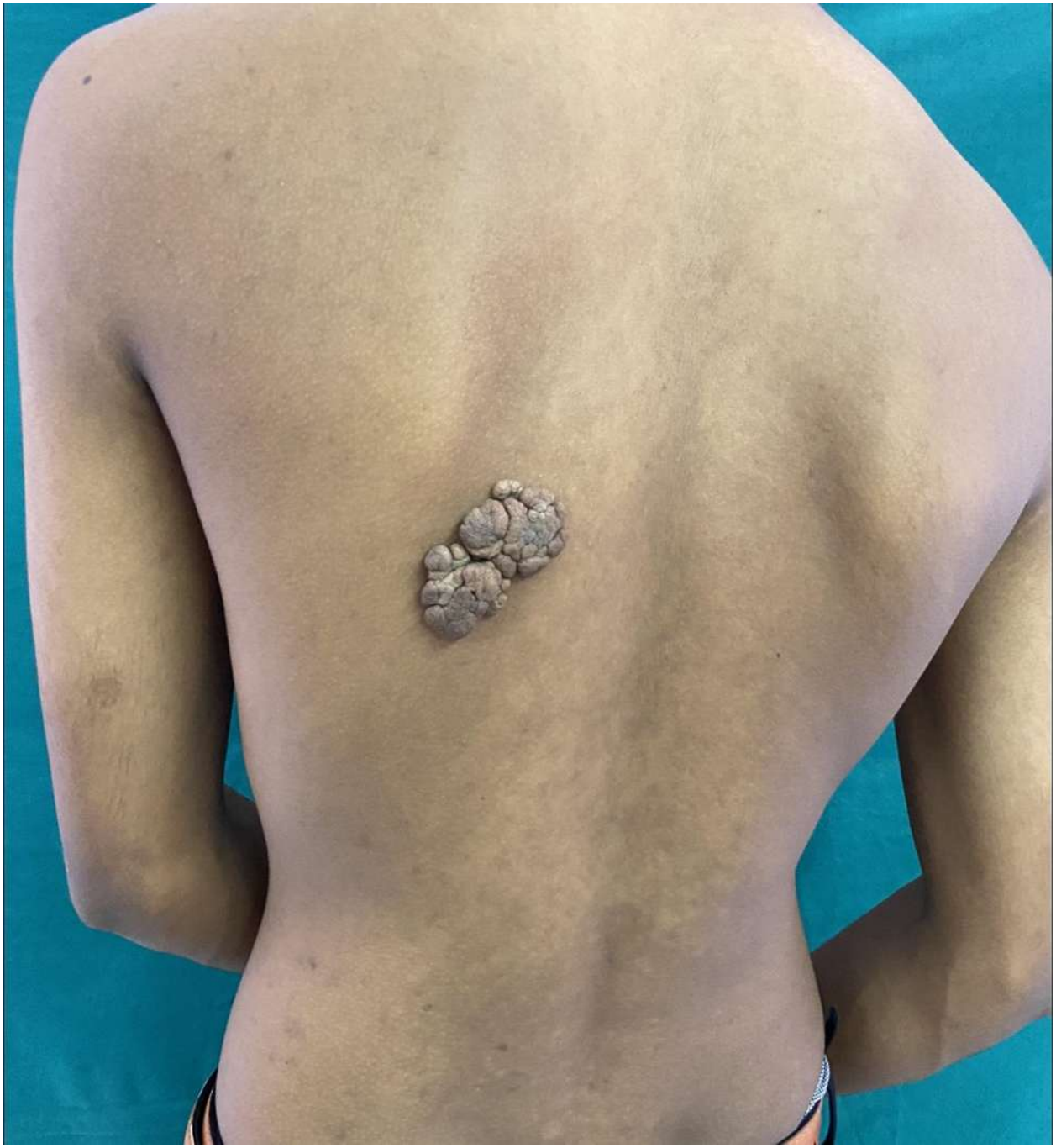

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Fig. 2A. Histopathology specimen on low power microscopy showing flaky hyperkeratosis, irregular acanthosis, and increased pigment seen in the basal layer. Dermis shows dilated blood vessels in papillary dermis.

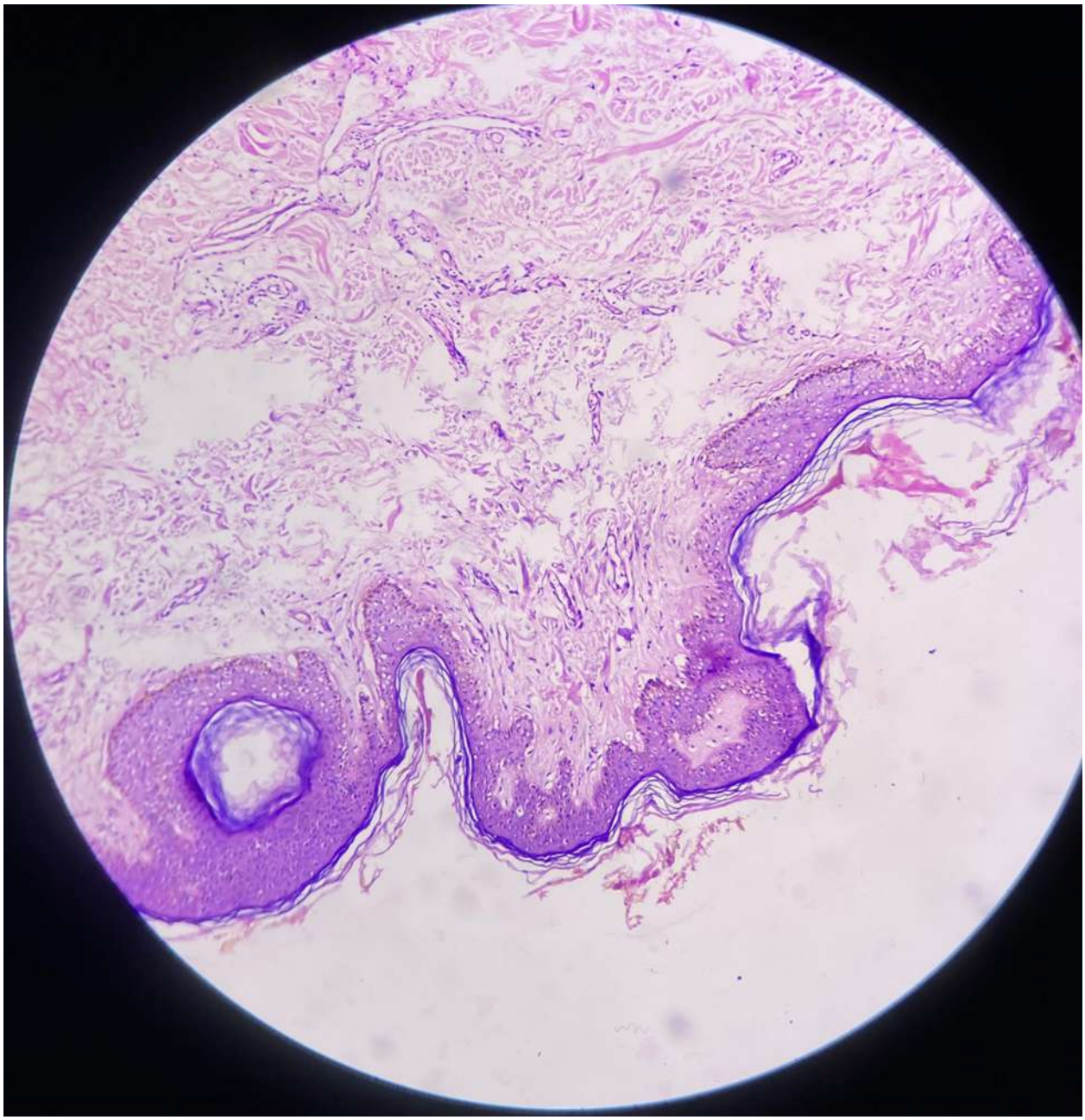

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Fig. 2B. Histopathology specimen on high power microscopy showing groups and strands of mature fat cells seen around and within blood vessels along with frequent foci of fibrosis intermixed with the fat tissue.

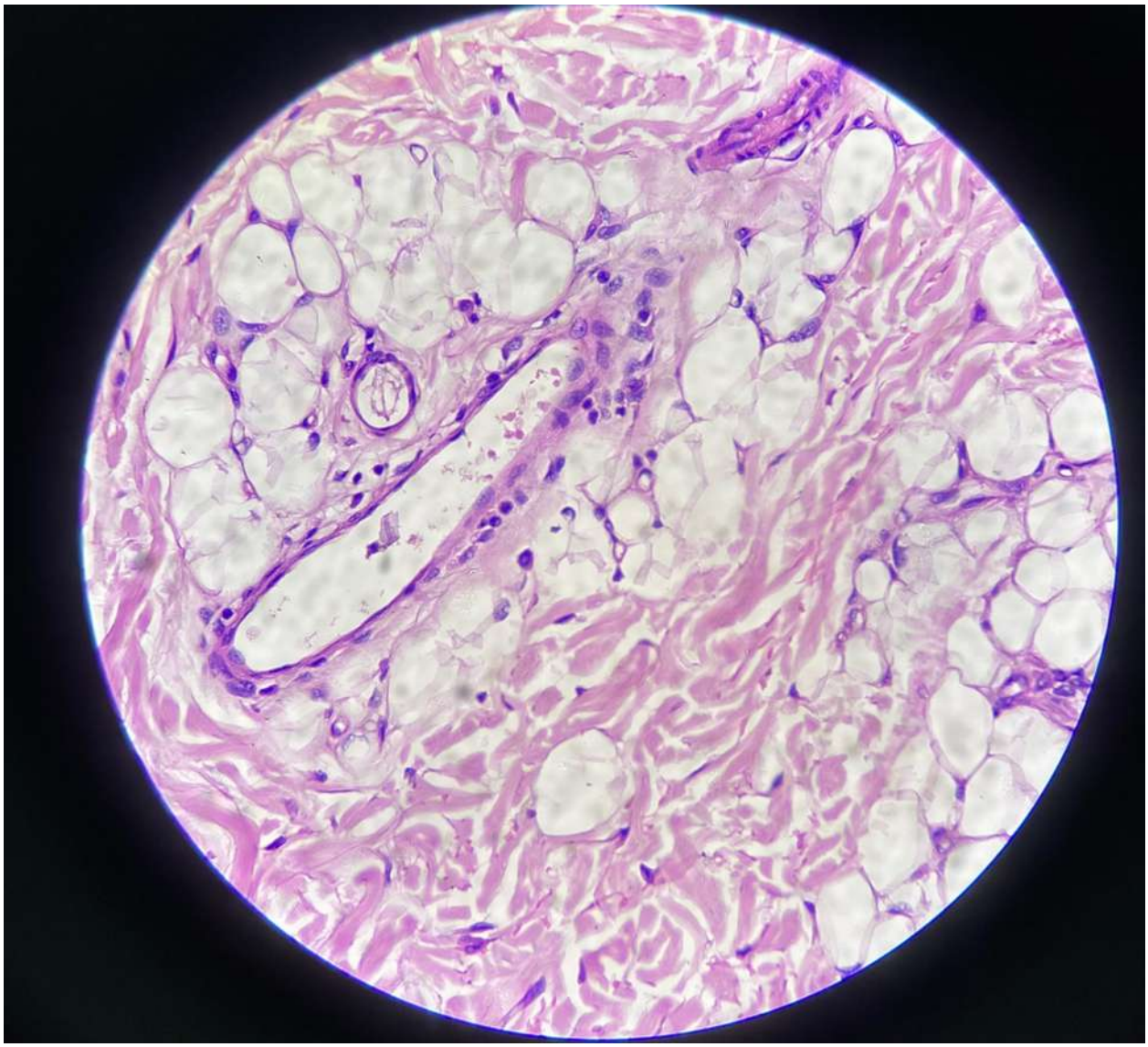

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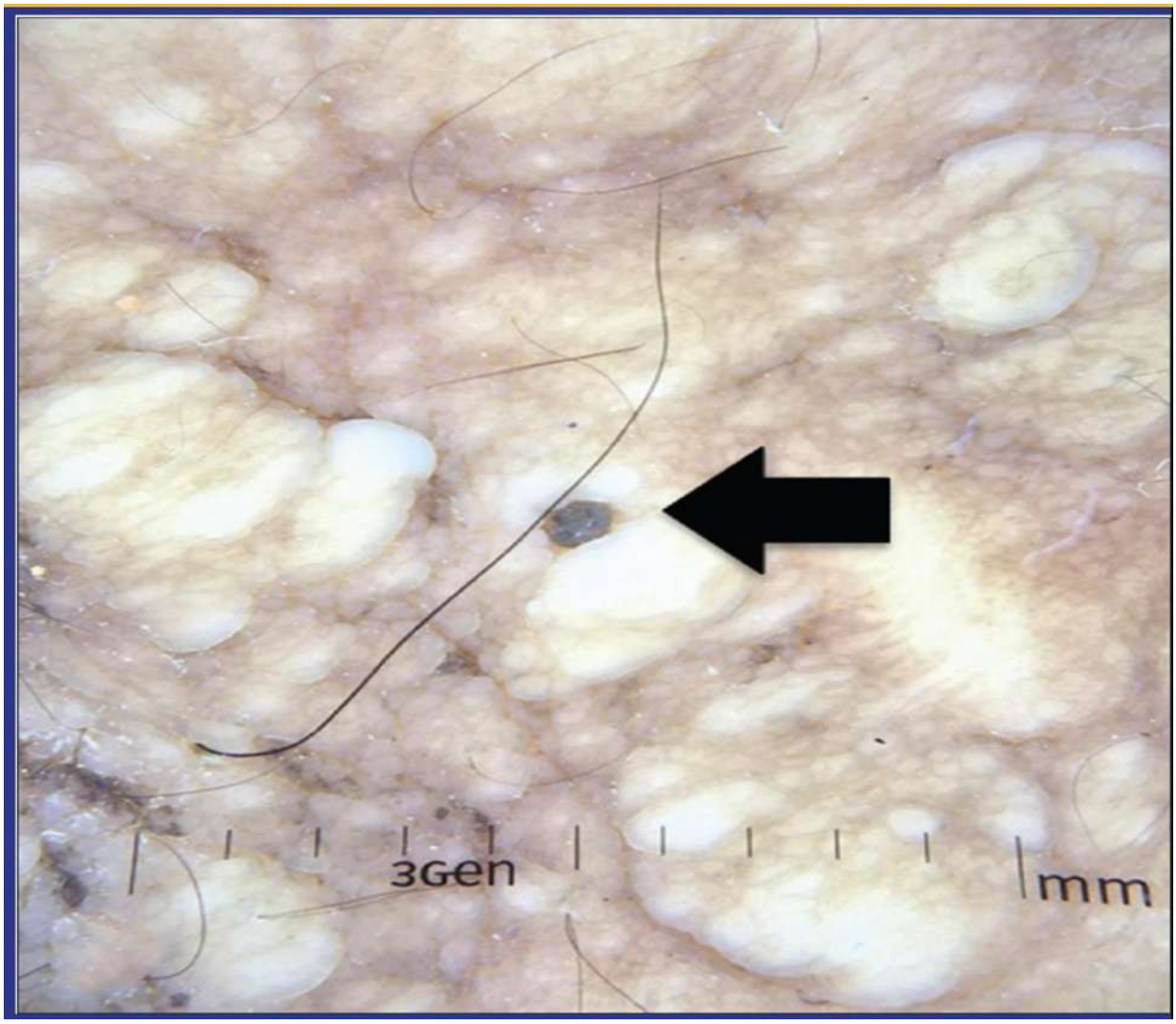
Fig. 3.Dermoscopy revealed the presence of sulci, keratotic gyri, scales noted inside the sulcus and reticular brown pigmented structure



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