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A Bizarre Presentation of Vulvar Nevus Lipomatosis Superficialis

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A 24-year-old girl presented to the Outpatient Department with multiple painless, insidious-onset swellings on the pubic area. Initially, they started as pin-head nodules when she was just eight years old and gradually increased in number and size. Apart from occasional itching in the area, she never experienced any noticeable symptoms over the years. However, as she grew older, concerns about cosmetic issues at marriage age compelled her to seek medical advice. Her past and family histories were not contributory. She had no addictions, medical co-morbidities, or local trauma.

On local examination, numerous, non tender, firm-to-hard, skin-coloured, mulberry-like nodules ranging from 0.5-1 cm in size were observed, densely covering both the mons pubis and the labia majora, but sparing the labia minora [Table/Fig-1]. There were no café-au-lait macules, hypertrichosis, or comedo-like lesions on them. She did not have any regional lymphadenopathy. The rest of the physical examination was unremarkable. Routine haemogram, urine, and stool examinations were within normal limits.

[Table/Fig-1]: Nevus Cutaneous Lipomatosis Superficialis (NCLS) of the vulva. Note that, multiple sessile densely clustered dermal nodulopapular lesions of varied sizes drape the entire mons pubis yielding a typical deceptive 'cobblestone' appearance to the female genitalia.

With a provisional diagnosis of NCLS, she underwent an incisional biopsy of the vulvar nodules, which revealed islands of mature adipocytes arranged in nests and cords in the dermis around blood vessels and eccrine glands. These ectopic fat cells interposed with collagen bundles without extending into the subcutaneous tissue. These histopathological features confirmed the diagnosis of NCLS.

Subsequently, the patient was prepared for complete surgical excision with skin grafting; however, she was lost to follow-up thereafter for reasons unknown to us.

The NCLS is a rare benign hamartoma of dermal adipose tissue that presents with certain striking clinical manifestations when it involves the female external genitalia. It was first reported by Hoffman and Zurhelle back in 1921 [1-4]. As the name implies, it is a nevoid anomaly characterised by aggregates of mature adipocytes embedded among dermal collagen [5-7]. A very high index of suspicion is needed to avoid misdiagnoses and subsequent anxiety among these patients [2,4,6,8].

Clinically, NCLS is classified into two types-classical (multiple) and solitary [1-4,9]. The former, also known as the Hoffman-Zurhelle nevus, may appear at birth, in infancy, or even during the first two decades of life [2,3,7]. Generally unilateral, it has a marked predilection towards the pelvic girdle, trunk, buttocks, and thighs [1-3]. It presents as a cluster of soft fleshy nodules that coalesce into large plaques, giving a typical pleated cerebriform appearance to the affected dermatome [4-6]. Sometimes, they may progress to form comedo-like plugs, foul-smelling discharge, and ulcerations of ischemic or traumatic origin [3,4]. Left untreated, it can grow as large as 106×30 cm-the largest reported NCLS to date-and further escalates therapeutic challenges [1,2]. However, the solitary variant presents in adulthood as a sessile dome-shaped discrete papule with a propensity for the trunk [2,3,5,9]. Follicular papules, hypertrophic pilosebaceous lesions, angiokeratoma, café-au-lait spots, and leukoderma can be rare associations [5,9].

Though the exact pathogenesis of NCLS is unclear, various theories have been proposed, such as adipose metaplasia, developmental fat displacement, dermal collagen degeneration leading to fat deposition, and capillary pericytes maturing into abundant adipocytes [1-4]. Recently, deletion of the 2p24 locus has emerged as the genetic basis [5,9].

Interestingly, during the counselling session, patient's family revealed a strong superstitious belief regarding the cause of the condition. In ancient times, rural areas of developing countries like India have been deeply influenced by myths surrounding various deities from Hindu mythology. This has led to the creation of fervent myths; one such belief is that this illness is the result of an unavoidable curse from a prominent deity-Lord Shiva, famously known as the 'destroyer' of the universe [10]. Consequently, it has been logically conceptualised that the genital manifestation of this disease requires the removal of offspring. Despite psychiatric counselling, it took us four days to dispel this delusion and an additional two days to persuade them to undergo further evaluation. Authors believed this may have been the reason for her becoming non compliant with treatment at later stages.

The common differentials of NCLS include skin tags, lipofibromas, and focal dermal hypoplasia [2,3,5,9]. Their close clinical resemblances necessitate histopathological examination to arrive at an accurate diagnosis [2,9]. In contrast to NCLS, skin tags have all their appendages intact with no adipocytes in the dermis [2,9]. In lipofibromas and focal dermal hypoplasia, adipocytes can be observed, but the dermal appendages are conspicuous by their absence [2,9].

Martinez-Ortega JI et al., recently reported a successfully treated case of a huge 20×10 cm left vulvar mass in a 38-year-old lady

presenting 20 years post-carbon dioxide laser excision [1]. Additionally, Singh N et al., documented a 10-year-old girl with a classical NCLS presenting as a 1.5×1 cm polyp over the left vulva [7]. Authors believe present NCLS case is unique in certain ways- first, it has a vulvar origin, an unusual site for its occurrence; second, the lesions were draping both the vulva; and lastly, it attained an atypical cobblestone-like appearance that posed a unique diagnostic challenge. To the best of authors knowledge, such a case has not been reported before.

Malignant transformation in NCLS is extremely rare [1-3]. Surgical excision with appropriate plastic reconstruction is curative and indicated only for aesthetic purposes and functional disturbances [2,4,9]. Staged resection is undertaken for giant masses spreading over several dermatomes [2,7]. Some non operative treatment options like topical corticosteroids, carbon dioxide ablative laser, cryotherapy, and intralesional injections of phosphatidylcholine and sodium deoxycholate render satisfactory results [2,7]. Thus, early recognition and timely interventions are crucial for better prognosis, psychological well-being, and quality of life of these individuals [2,7].

In conclusion, NCLS, though rare, should always be considered as one of the key differentials when evaluating patients with multiple papulomatosis of the vulva. Understanding its typical gross and microscopic findings helps in clinching the diagnosis convincingly and largely prevents patient apprehension. However, further large cohort studies are required for a better understanding of its aetiopathogenesis and tailoring treatment strategies for optimised outcomes.

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