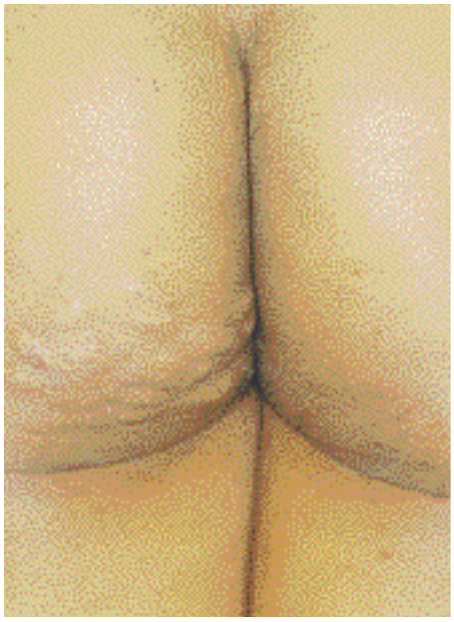
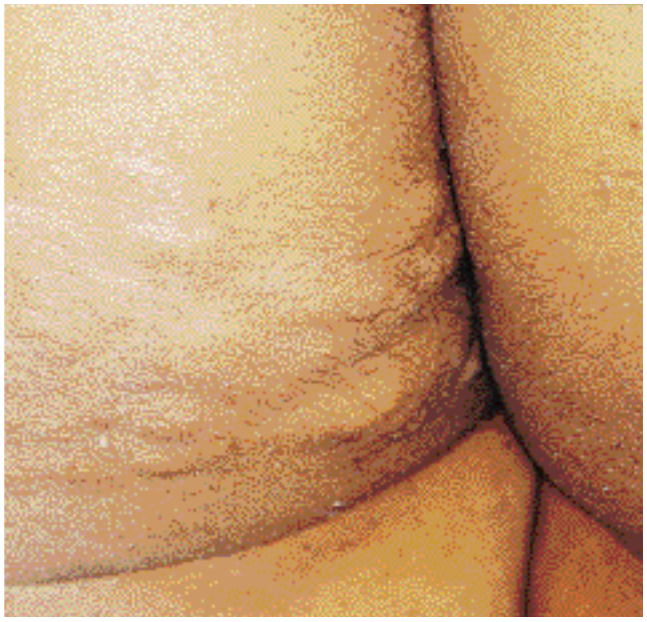
What Is Your Diagnosis?





A 23-year-old black woman presented with an enlarging growth on her left buttock. The lesion had been present for 3 years. She reported only slight discomfort with prolonged sitting but was otherwise completely asymptomatic. The patient was healthy and denied a history of trauma at that site. There was no family history of neurofibromatosis.

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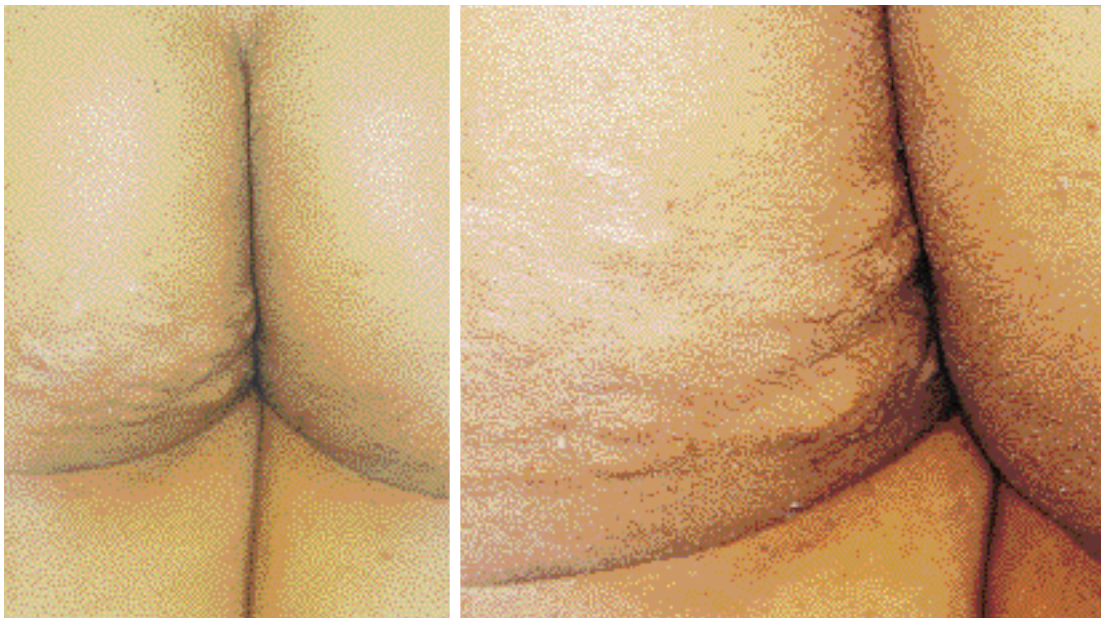
The Diagnosis: Nevus Lipomatosus Cutaneous Superficialis



Nevus lipomatosus cutaneous superficialis (NLCS) is a rare condition characterized by the presence of mature adipose tissue within the dermis. Three types of this condition have been described in the literature. The classical nevus, first described by Hoffmann and Zurhelle[1] in 1921, presents at birth or within the first 2 decades of life as a plaque of flesh-colored to yellow, soft, nontender papules and nodules in a linear distribution, usually occurring unilaterally. These lesions occur most commonly in the pelvic girdle region, particularly the buttocks. However, NLCS also has been reported to occur on the scalp,[2] knee,[3] axilla,[4] and arm.[4]

In 1968, Weitzner[5] described a second subtype of NLCS as a solitary lesion of the scalp. This solitary NLCS lesion presents as a single soft, flesh-colored to yellowish papule or nodule. It can appear anywhere on the body but usually occurs on the trunk. Unlike the plaque type, the lesion appears later in life, usually in the fourth or fifth decade. Although this subtype of NLCS is histologically similar to the plaque type, Mehregan et al[6] proposed the term *pedunculated lipofibroma* based on its distinctive clinical and pathologic features. A review of 32 cases by Nogita et al[7] further supported the designation of these lesions as *solitary pedunculated lipofibromas*. Solitary lesions of NLCS in the gluteal region have been referred to as a form of Hoffmann-Zurhelle type NLCS.

There have been reports of a third, more generalized, type of NLCS. Characterized by marked folds over the skin surface, this type is referred to as the *Michelin tire baby*. Ross[8] described the first case of a child in South Africa with extensive folding of the skin with underlying nevus lipomatosus. Another such case was later described in 1979 by Gardner et al.[9]

Histopathologic examination of NLCS is characterized by the presence of mature adipose tissue in the dermis. The amount of ectopic fat present varies greatly, ranging from less than 10% to 70% of the lesion. The fat tends to localize around subpapillary blood vessels in lesions with little adipose tissue. In lesions with large amounts of adipose tissue, the fat lobules are unevenly arranged throughout the

dermis and blur the distinction between the dermis and the hypodermis. The rest of the dermis may show irregularly arranged or thickened collagen bundles and an increase in the number of fibroblasts in the papillary dermis. There also may be an increase in mononuclear cells, including mast cells and blood vessels. Furthermore, pilosebaceous follicles and elastic fibers may be reduced.[4] The epidermis and appendages essentially are unaltered.

Various theories on the pathogenesis of NLCS exist. Early theories by Hoffman and Zurhelle[1] suggested that the deposits of fat are caused by primary degenerative change in connective tissue. In 1937, Robinson and Elis[10] proposed that the ectopic fat cells result from focal heterotopic development of adipose tissue.

Holtz suggested a vascular origin of the ectopic adipose tissue present in NLCS. He observed the growth of immature adipose tissue from blood vessels, such as that seen in embryonic tissue, which destroys the surrounding connective tissue.[11] Holtz theorized that degenerative changes in the connective tissue were therefore secondary to the growth of adipose tissue from blood vessels. Lynch and Goltz[12] further concluded that the process is similar to embryonic development of normal adipose tissue.

Wilson Jones et al[4] reported that the lipogenesis in NLCS recapitulates the way fat develops in the primitive mesenchyme of the fetus. They further noted that mononuclear cells in immature lesions differentiate into lipoblasts around the proliferating capillaries. An electron microscopic study of 4 cases of classic NLCS by Reymond et al[13] also demonstrated an angiocentric origin of the adipose cells and concluded that the mature adipocytes appeared to stem from a precursor of young adipocytes, further supporting the comparison to fetal adipogenesis.

Another electron microscopic study by Dotz and Prioleau[14] examined an unusual case of NLCS that developed in a 43-year-old patient. They observed small mature lipocytes but were unable to demonstrate lipoblasts or the transition of mesenchymal cells to lipocytes. However, numerous mast cells were observed throughout the dermis, adipose tissue, and surrounding blood vessels. In a study of solitary NLCS that developed after the third decade, Nogita et al[7] also reported an increase in mast cells throughout the adipose tissue and a lack of lipoblasts on electron microscopic evaluation. These similar findings may represent either a feature of late onset lesions or lesions of long duration. Further research is needed to determine if there is a relevant histologic difference between the 2 subtypes.

The lesions of NLCS usually are asymptomatic, and we were unable to find any reported case of malignant transformation in the literature. Treatment consists of surgical excision or, given the benign course of the disorder, the lesion may simply be observed.

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