A Case of Adult Onset Classical Type of Nevus Lipomatosus Cutaneous Superficialis

Geç Başlangıçlı Klasik Tip Nevüs Lipomatozis Kutanöz Süperfisiyalis Olgusu

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ABSTRACT Nevus lipomatosus cutaneous superficialis is a rare malformation characterized by the presence of ectopic adipocytes in the dermis. Two types of it have been described. The classical type, firstly reported by Hoffman and Zurhelle, is presented with multiple, unilateral, soft, non-tender, pedinculated, skin colored or yellowish papules, plaques and nodules with smooth or cerebriform surface. The lesions are usually congenital or appear in the first two decades of life. The main location of the classical type is pelvic girdle. Solitary type of nevus lipomatosus cutaneous superficialis usually occurs as a single nodular lesion after the age of 20 and typical localization has not been described. We report the case of a 46-year-old female with classical type of nevus lipomatosus cutaneous superficialis on the right lumbar region that developed five years ago.

Key Words: Adipocytes; nevus

ÖZET Nevüs lipomatozis kutanöz superfisiyalis, dermisde ektopik adipositlerin varlığı ile karakterize nadir rastlanılan bir malformasyondur. İki tipi tanımlanmıştır. İlk olarak Hoffman ve Zurhelle tarafından tanımlanan klasik tip çok sayıda, unilateral yerleşimli, yumuşak, ağrısız, pedinküllü, deri renginde ya da sarımsı renkte düzgün veya serebriform yüzeyli olabilen papül, plak ya da nodüllerle karakterizedir. Lezyonlar genellikle doğumda veya ilk iki dekad içerisinde ortaya çıkar. Klasik tipin esas lokalizasyonu pelvik alandır. Soliter tip, 20 yaşından sonra nodül şeklinde ortaya çıkar ve tipik bir lokalizasyon tanımlanmamıştır. Sağ lomber bölgede, 5 yıl önce klasik tip nevüs lipomatozis kutanöz superfisiyalis ortaya çıkan 46 yaşında kadın olguyu sunuyoruz.

Anahtar Kelimeler: Adipositler; nevüs

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Nevus lipomatosus cutaneous superficialis (NLCS) is an uncommon, benign, developmental disorder characterized by ectopic adipose tissue in the dermis.[1] Two types of NLCS have been described: classical (multiple) and solitary forms. The classical type occurs from birth to first two decades of life. The onset of solitary type is in the adult life after the age of 20.[2] We report a case of the classical type of NLCS with adult onset.

CASE REPORT

A 46-year-old female presented with a large lesion on the right side of her back since the age of 41. The lesion was asymptomatic but had progresively increased in size during the last six months. She was in good health. Her family history was unremarkable.

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Physical examination showed nontender, soft, skin-colored multiple nodules on the right lumbar area (Figure 1). Their surfaces were smooth. No cafe-au-lait macules, axillary freckling and neurologic abnormality were observed. Routine laboratory examination was normal.

An insicional skin biopsy specimen showed mature adipose tissue interposed with bundles of collagen in the reticular dermis (Figure 2). Histopathological findings were consistent with the diagnosis of NLCS. The patient was refered to the plastic surgery department for complete surgical excision.

DISCUSSION

NLCS is a rare idiopathic skin malformation characterized by dermal deposition of mature adipose tissue. The classisal (multiple) type of NLCS was first described by Hoffman and Zurhelle in 1921.[1] It occurs as a skin-colored or yellowish, nontender, soft, sessile or pedinculated papules, plaques or noduls with smooth or cerebriform surface.[3] The lesions are usually unilateral, linear or zosteriform in distribution.[4] The common predilection is the pelvic girdle area especially buttocks, lower back and upper part of the posterior thighs.[5] The classical type of NLCS on face, nose, pinna and abdominal area have been reported.[4,6] Sexual or familial predisposition have not been described.[5] The classical NLCS occurs at birth or within the first two decades of life. Adult onset of classical NLCS as seen in our case has been reported very rarely.[4,7,8] In contrast the solitary form usually develops later in life and occurs as a single nodul with a predilection of head and neck area.[3,9,10]

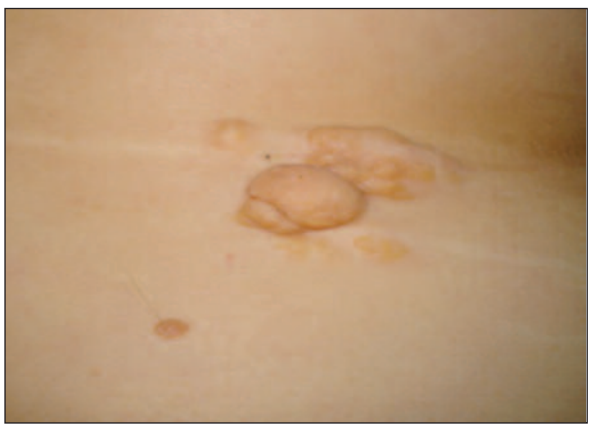

FIGURE 1: Soft, skin-colored multiple nodules on the right lumbar area.

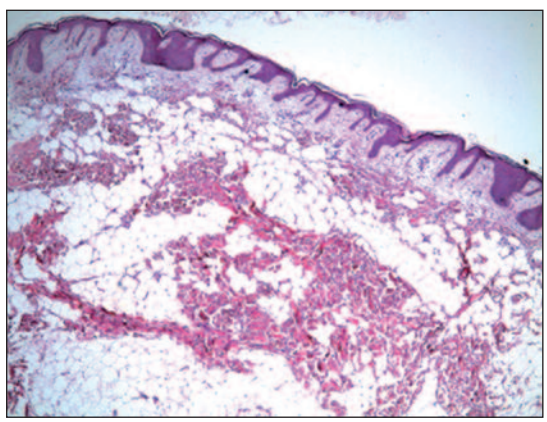

FIGURE 2: Mature adipose tissue interposed with bundles of collagen in the reticular dermis (H&EX40).

NLCS is mostly asymptomatic, however ulceration with foul smelling and discharge of the lesion have been rarely reported.[3,4] The reason of the ulceration is thought to be due to external trauma or ischemia of dermal blood vessels depending on ectopic adipose tissue compression.[7] Abnormalities associated with NLCS have not been reported; but sporadic cases with coexistent pigmentary disorders such as cafe-au-lait macules, leukodermic spots, hypertricosis and commedo-like lesions on the surface of NLCS have been described.[4,7,11-13]

Histopathologically, the presence of ectopic mature adipocytes among the collagen bundles and subpapillary plexus are observed in the dermis.[3] In some cases, abnormal folliculosebaceous structures such as sebaceous trichofolliculoma, folliculosebaceous cystic hamartoma, dermoid cysts and epidermal changes like acanthosis, increased basal pigmentation, basket weave hyperkeratosis and focal elongation have been reported.[14-16] Etiopathogenesis of NLCS has not been understood yet. Hoffman and Zurhelle suggested that it might be caused by adipose metaplasia due to degenerative differantiation in dermal collagen and elastic tissue. Other theories include focal heterotopic development of adipose tissue and preadipose tissue derived from dermal blood cells.[12,17] Only one cy-

togenetic study that showed 2p24 deletion was reported.[18] Further studies are needed to determine the role of genetic in the development of NLCS.

Treatment of NLCS is only needed for cosmetic problems. Surgical excision is the most preferred therapy with a low recurrence rate. Surgical excision of multiple type of NLCS usually requires skin graft that cause unsatisfying cosmetic results. CO_2 laser treatment has been reported as a successful alternative therapy of multiple NLCS.[19] In 2012, a case of multiple NLCS treated with CO_2 laser therapy was reported with a recurrence after a few months of treatment. The authors suggested that staged excision was an effective therapy with better cosmetic results and no recurrence.[20] In conclusion, we present a case of classical but adult onset form of NLCS on the flank which has been reported rarely up to date.

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