



A GIANT PERINEAL NEVUS LIPOMATOSUS SUPERFICIALIS: CASE REPORT

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ABSTRACT

Nevus lipomatosus superficialis is a rare hamartomatous malformation which is composed of ectopic adipocytes in the dermis. It was first reported in 1921 by Hoffmann and Zurhelle. Two clinical forms of nevus lipomatosus superficialis have been described: classical (multiple) and solitary. Classical form of nevus lipomatosus superficialis is usually found on pelvic girdle, trunk, buttocks and thighs as soft, skin colored papules or nodules. It is usually present at birth or it appears in the first two decades of life. The solitary form of lipomatosus superficialis appears as a solitary papule or nodule on the back, scalp and arms of the patients with late onset.

The lesions are usually asymptomatic, however some patients may complain about pain and itching. Malignant transformation of nevus lipomatosus superficialis has not been reported yet. Therefore, surgical intervention is only necessary for the patients who have cosmetic concerns. Recurrence after surgical removal is very rare.

Perineum is an uncommon localization for nevus lipomatosus superficialis. Hereby, we report a 55-year-old Caucasian female with a 6x5,5x4 cm mass in the perineal region. The patient had cosmetic concerns, therefore she wanted the lesion to be removed surgically. The lesion was surgically removed. The histopathological evaluation of the specimen revealed nevus lipomatosus superficialis.

A solitary type of giant nevus lipomatosus superficialis in the perineal region of a patient over the age of 50 is a very rare condition. Even rarely seen, nevus lipomatosus superficialis should be kept in mind in the differential diagnosis of perineal masses.

UDC CODE & KEYWORDS

■ UDC: 6 ■ Nevus lipomatosus superficialis ■ Perineum ■ Surgical excision

INTRODUCTION

Nevus lipomatosus superficialis is a rare nevoid anomaly of unknown etiology which usually occurs on pelvic girdle, lumbar region, buttocks and thighs. Nevus lipomatosus superficialis is composed of ectopic adipose tissue within the dermis. It usually presents as a group of skin colored or yellowish, asymptomatic papules or nodules with a smooth, wrinkled or peau d'orange appearance on the surface (Ekmeççi, Köşlü & Sakız, 2003). Nevus lipomatosus superficialis may rarely appear as a sessile solitary papule or nodule on the back, scalp and arms of the patients (Uncu et al., 2005; Das, Das, Bandyopadhyay & Kumar, 2015).

Men and women are affected equally, often at birth or in the first two decades of life. There is no known hereditary predisposition (Uncu et al., 2005; Avhad & Jerajani, 2013). Occasionally the lesions may present in the fifth or sixth decade of life. However, nevus lipomatosus superficialis is generally a congenital malformation and it is not associated with any systemic abnormalities.

Despite the fact that nevus lipomatosus superficialis is usually asymptomatic, it may cause cosmetic problems. Surgical removal is the definitive treatment of nevus lipomatosus superficialis. Nevus lipomatosus superficialis has no risk of malignant transformation. Moreover, recurrence is very rare (Ekmeççi, Köşlü & Sakız, 2003). Hereby, we present a 55-year-old Caucasian female with a giant perineal nevus lipomatosus superficialis, which is a very rare entity in daily clinical practice.

Case Report

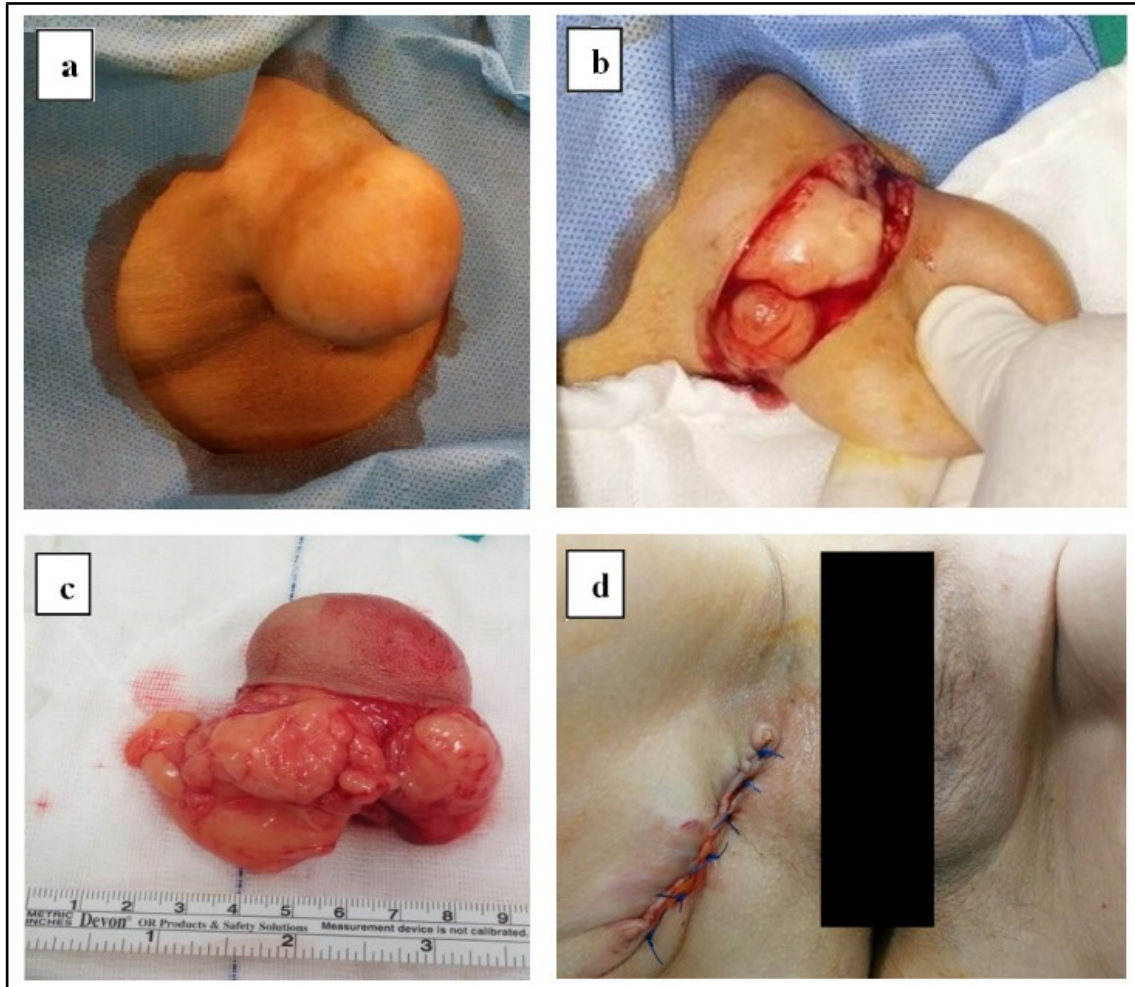
A 55-year-old white Caucasian female who presented with painless mass in the perineal region was admitted to general surgery department for further clinical evaluation. The patient admitted that she had the painless lump for the last 15 years, however it appeared to have enlarged gradually in the last two years. The patient had cosmetic concerns, therefore she wanted the lesion to be removed surgically.

Her past medical history was unremarkable. Laboratory tests did not reveal any abnormalities. The detailed physical examination of the patient revealed a skin colored, 6x5, 5x4 cm, soft, solitary mass with a smooth surface and well defined margins at the right side of the perineal region, lying between vulva and the right inner thigh.

The lesion was surgically removed under local anesthesia without any complications. The histopathological examination of the specimen revealed nevus lipomatosus superficialis. Figure 1 shows the preoperative appearance and the intraoperative surgical excision of the giant nevus lipomatosus superficialis.



Figure 1: a) Preoperative view of the perineal nevus lipomatosus superficialis b) Intraoperative view: surgical excision c) Specimen: 6x5, 5x4 cm nevus lipomatosus superficialis d) The incision was closed primarily



Source: Authors

Discussion

Perineum is a rare localization for nevus lipomatosus superficialis. Clinically, the perineal lump presented above resembled other lesions such as lipoma, hernia and epidermoid inclusion cyst. However, our initial clinical diagnosis was in favor of a giant perineal lipoma. The histopathological evaluation of the specimen after surgical excision revealed a nevus lipomatosus superficialis.

A nevus lipomatosus superficialis is a benign lesion which shows ectopic adipose tissue in the dermis. It was first reported in 1921 by Hoffmann and Zurhelle. Two clinical forms of nevus lipomatosus superficialis have been described: classical (multiple) and solitary (Ekmeççi, Köşlü, Özeren & Ayabakan, 2005). The classical type presents as grouped, skin colored to yellowish, soft, slow growing, asymptomatic, unilateral, multiple papules, nodules or plaques with a smooth or cerebriform surface. They are usually found on pelvic girdle, trunk, buttocks and thighs at birth or they occur in the first two decades of life. The solitary type, which is also known as pedunculated lipofibroma, appears as a papule or nodule at any part of the body such as clitoris, scalp, eyelid, nose and knee. This form affects older patients in the fifth or sixth decade of life (Goucha et al., 2011; Yap, 2009). The lesions of nevus lipomatosus superficialis vary in size. These lesions are usually stable but they may slowly enlarge and present as giant forms. Trauma and ischemia can lead to ulcerative lesions (Dhamija, Meherda, D'Souza, & Meena, 2012). Nevus lipomatosus superficialis is usually asymptomatic, however pain and itching may occur. In addition, it may cause cosmetic concerns (Ekmeççi et al., 2003). The exact pathogenesis of nevus lipomatosus superficialis is not known. However, it is suggested that degenerative changes in connective tissue, developmental displacement of adipose tissue or mature adipose tissue deposition which is differentiated from perivascular lipoblasts might play a role in the etiology of the disease (Kim, Choi, Kim, Nam & Choi, 2012). Nevus lipomatosus superficialis may be associated with café-au-lait macules, scattered leukoderma, hypertrichosis, capillary hemangioma, trichofolliculoma and comedo-like lesions (Das et al., 2015; Patil, Narchal, Paricharak, & More, 2014). The histopathological examination of nevus lipomatosus superficialis reveals ectopic adipocytes in the dermis, especially around blood vessels and between collagen bundles without a connection to the hypodermis (Dhamija et al., 2012). Nevus sebaceous, epidermal nevi, neurofibroma, lymphangioma, focal dermal hypoplasia, skin tag and lipoma should be included in differential diagnosis of

this rare entity. Histopathological evaluation should be performed to reach a definitive diagnosis (Uncu et al., 2005; Patil et al., 2014). Nevus lipomatosus superficialis can be treated successfully with the surgical removal of the lesion, without postoperative recurrence. It is suggested that better cosmetic results might be obtained by CO₂ laser treatment of large lesions. However, the lesions may reoccur after laser treatment (Kim et al., 2012).

CONCLUSION

Nevus lipomatosus superficialis is rarely seen in the perineum. Our initial clinical diagnosis for the 6x5,5x4 cm perineal mass presented above was in favor of a giant perineal lipoma. The histopathological evaluation of the perineal mass after surgery revealed a nevus lipomatosus superficialis. A solitary type of giant nevus lipomatosus superficialis in the perineal region of a patient over the age of 50 is a very rare condition. Therefore, clinical differential diagnosis of a perineal mass should include nevus lipomatosus superficialis, however, histopathological examination is mandatory to reach a definitive diagnosis.

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