NEVUS LIPOMATOSUS CUTANEOUS SUPERFICIALIS

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Introduction

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare adipose tissue hamartoma. Two distinct subtypes have been reported. The solitary subtype is more common and presents as a single, pedunculated skin colored nodule.1 The classic subtype of NLCS is rare and characteristically occurs in a segmental or zosteriform distribution most commonly on the posterior surface of the trunk or thigh.2,3 We present a case of classic NLCS.

Clinical and Histopathologic Images

Fig. 1 Fleshy papules coalescing into 22cm x 10cm plaque on posterior thigh and leg.

Fig. 2 H&E demonstrating mature adipocytes in the dermis.

Fig. 3 VVG Elastin Stain

Discussion

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare adipose tissue hamartoma. Two distinct subtypes have been reported including a solitary subtype and a classic form.2 The classic subtype is often in a segmental or zosteriform distribution and it is typically unilateral, as seen in our patient.2,3 It often presents at birth or within the first two decades of life.3,4 Clinically it appears as multiple asymptomatic, soft papules and nodules that coalesce into a plaque that may have a smooth or cerebriform surface.3,4 The classic subtype most commonly forms on the lower back, gluteal area or posterior thighs.3

Histologically, the lesion is characterized by mature adipocytes in the dermis. Clinically, the differential diagnosis includes plexiform neurofibroma, connective tissue nevus and vascular malformations.3 The histologic differential diagnosis includes focal dermal hypoplasia (Goltz syndrome), a rare X-linked dominant disorder, which is invariably present from birth in affected females. While the histologic appearance on H&E can be identical, on special staining, the elastic fibers may be counterintuitively increased or thickened in NLCS. NLCS is benign and typically asymptomatic. However, patients may elect for treatment to optimize cosmesis. Surgical excision is a well recognized treatment approach but is invasive. CO2 laser has been reported to be successful in a localized lesion and may be more appealing to patients due to the less invasive nature of the procedure.5

Case Presentation

A 50-year-old Caucasian female presented with a complaint of a rash on her left leg. The lesion had been present for ten years and had never been previously treated. At the time of presentation, the patient reported it had recently grown in size. The patient was otherwise in good health. Physical examination revealed soft, fleshy papules and nodules coalescing into a 22 cm x 10 cm plaque with cerebriform appearance and various shades of yellow and pink. The lesion was in a segmental distribution on her posterior thigh and leg (Fig 1). Biopsy was obtained and sections demonstrated an admixture of mature adipose tissue lobules within the collagen of the reticular dermis (Fig. 2). A Verhoeff Van Gieson (VVG) stain demonstrated preserved elastic fibers in a normal pattern (Fig. 3). A diagnosis of nevus lipomatosus cutaneous superficialis was made. The pathology was reviewed with the patient. For cosmetic reasons, the patient was interested in treatment. She was referred to plastic surgery where she was offered full thickness skin grafting or excision with tissue expansion and primary closure. However, the patient elected to defer treatment and was lost to follow up.

References