A 36-year-old man presented with a group of partially erythematous, yellowish papules and plaques ranging from 5 to 20 mm in diameter on the right side of the upper back of 20 years’ duration. They were surgically excised 8 years prior but recurred and spread. The lesions occasionally were painful and tender with redness and discharge.

What’s the diagnosis?

a. leiomyoma  
b. lymphangioma  
c. nevus lipomatosus cutaneous superficialis  
d. plexiform neurofibroma  
e. sebaceous nevus
The Diagnosis: Nevus Lipomatosus Cutaneous Superficialis

A punch biopsy was obtained from a skin lesion, which showed orthokeratosis, irregular acanthosis, papillomatosis, intense edema in the upper dermis, and mature fat lobules that dissected collagen fibers in the reticular dermis (Figure). Classical-type nevus lipomatosus cutaneous superficialis (NLCS) was diagnosed based on these clinical and histopathological findings. The patient was referred to the plastic surgery clinic for total excision of all lesions.

Nevus lipomatosus cutaneous superficialis is a rare hamartoma characterized by ectopic deposition of mature adipose tissue in the dermis. It was first described by Hoffmann and Zurhelle in 1921. Clinically, NLCS is classified into 2 subtypes: classical (multiple) and solitary. Classical-type NLCS is characterized by multiple pedunculated or sessile, soft, cerebriform, yellowish papules and nodules, especially in the pelvic area. Solitary-type NLCS presents as a sessile papule or nodule with no predilection for localization. Although the classical form of NLCS generally occurs in the first 2 decades of life, the solitary form usually appears in adulthood.

Nevus lipomatosus cutaneous superficialis has no gender predilection and there is no genetic or congenital defect association.

The pathogenesis of NLCS is still unknown, but some theories have been proposed, such as the development of adipose metaplasia secondary to degeneration of connective tissue, the formation of a true nevus resulting from heterotopic development of adipose tissue, and the development of mature adipocytes from pericytes in dermal vessels.

Histopathology of NLCS shows clusters of ectopic mature adipose tissue in varying rates (10%–50%) between collagen bundles in the dermis. Characteristically, there is no connection between the ectopic mature adipose tissue and the subcutaneous adipose tissue. The differential diagnosis of NLCS includes neurofibroma, lymphangioma, sebaceous nevus, fibroepithelial polyps, leiomyoma, and lipomas.

Treatment of NLCS generally involves basic surgical excision; however, patients treated with CO2 laser also have been reported in the literature. Because of the growth tendency and the large size of the classical form of NLCS, recurrence may occur, as in our case. In such cases, gradual surgical excision is recommended. We present this case to indicate that undesirable surgical results or relapse may occur in untreated patients because of lesion growth and delayed diagnosis.

REFERENCES


