RESEARCH
www.commprac.com
ISSN 1462 2815

GIANT HOFFMANN-ZURHELLE'S NEVUS LIPOMATOSUS SUPERFICIALIS AT AN ATYPICAL SITE- CASE REPORT OF A RARE HAMARTOMA

Anannya S [1], Abyramy Balasundaram [2]*, Vivekanandh K [3], Vimal Chander R [4], Durgadevi G [5] and Afthab Jameela Wahab [6]

[1,2,3,5,6] Department of Dermatology, Saveetha Medical College,
Saveetha Institute of Medical and Technical Sciences, Saveetha University.
[4] Department of Pathology, Saveetha Medical College,
Saveetha Institute of Medical and Technical Sciences, Saveetha University.
*Corresponding Author Email: b.abyramy@gmail.com

DOI: 10.5281/zenodo.12515140

Abstract

Nevus lipomatosus cutaneous superficialis (NLCS) is an uncommon idiopathic benign cutaneous hamartoma, with the occurrence of ectopic mature adipose tissue in the dermis that does not extend into the subcutaneous adipose tissue. Clinically, there are two forms. The more prevalent classical presentation manifests as numerous yellow papules and nodules that are soft, pedunculated, non-tender, and frequently coalesce to create a plaque. The less common solitary form manifests as a sessile or dome-shaped papule appearing in the 3rd or 6th decade of life. The classical NLCS usually shows up within the first three decades of life or at birth. Although the pelvic or gluteal region is usually affected in this classic type, it can also sporadically affect the chest, face and abdomen with some authors reporting it on the scrotum, vulva and pinna. On very rare occasions the lesions can be giant. In this case report, we describe a giant classical NLCS in the neck region in a 53-year-old male undiagnosed since 40 years.

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS), otherwise known as pedunculated lipofibroma or dermolipoma, was described first in the year 1921 by Hoffman and Zurhelle [1]. There are two clinical forms [2]. The more common classical presentation manifests as numerous yellow papules and nodules that are soft, pedunculated, non-tender, and frequently coalesce to create a plaque. The less common solitary form manifests as a sessile dome-shaped lesion occurring in the 3rd or 6th decade of life. There is no hereditary predisposition or sex predilection of this disease. According to previous studies, the classic type involves the pelvic girdle area with histopathology displaying scattered or clustered lobules of adipose tissue, made up of collagen bundles interspersed with well-differentiated adipocytes, and chronic inflammatory infiltrate in the perivascular region of dermis and subcutis. [3]. There have been no reports of malignant transformation. Excision by surgery is curative, and there have been no documented recurrences. In our case report, we describe a clinicopathological case of a 53 year old male with giant classical NLCS grown unchecked in an unusual location such as the neck and undiagnosed since 40 years.

CASE REPORT

A 53 year old male came with multiple growths over the right side of his neck which was asymptomatic and gradually increasing in size over the past 40 years. He gives history of sudden onset pain, redness, and pus discharge for the past 2 days from one of the growths in the posteromedial region following minor trauma. He was diagnosed to have diabetes mellitus, and dyslipidemia for which he is on regular medications for

2 years. Cutaneous examination revealed a skin-colored to brownish, well-defined, non-tender, soft to firm, sessile, lobulated, cerebriform plaque of size 15x10x5cm over the right side of the neck [Fig. 1a] and few small similar soft papulo-nodules over the right mandible and below the hairline. The posteromedial aspect showed erythema, tenderness, pus discharge, while rest of the lesion showed no surface changes [Fig. 1b]. Multiple cherry angiomas are noted on the trunk and no other cutaneous lesions noted elsewhere on the skin. Systemic examination was unremarkable. On investigation, complete hemogram and blood biochemistry studies were within normal limits. Biopsy was performed from one of the nodules over the right mandibular region.

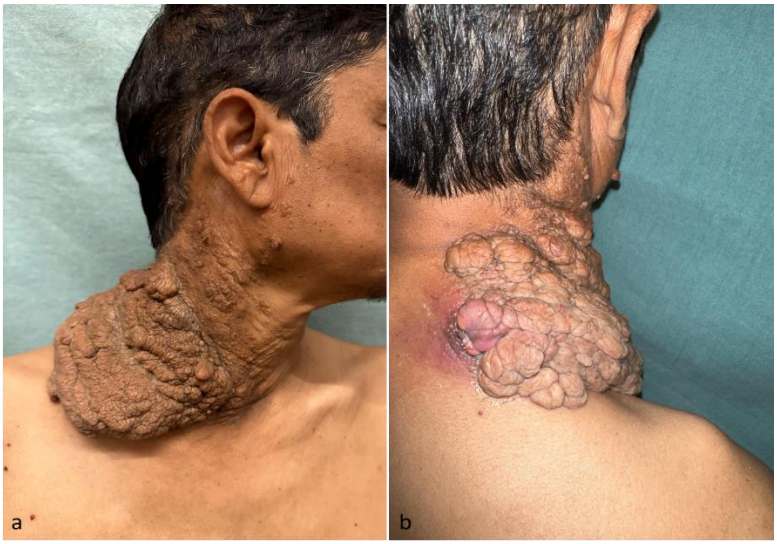

Figure 1: a) Skin-coloured to brownish, well-defined, soft, lobulated, cerebriform plaque of size 15x10x5cm over the right side of the neck; b) Erythema and pus discharge over posteromedial part of the lesion.

On histopathological examination, partial flattening of the rete ridges was seen along with localized thinning of the epidermis. Few scattered mature adipocytes along with minimal perivascular lymphocytic infiltrates were noted in the superficial dermis [Fig. 2a, 2b]. The deep dermis, adnexal structures and subcutaneous mature adipose tissue were unremarkable.

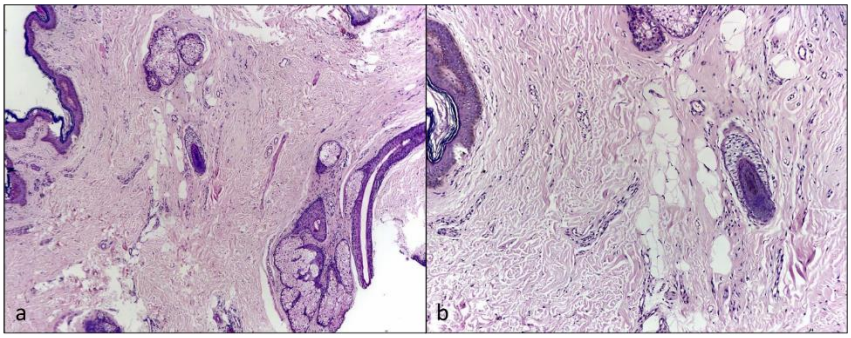

Figure 2: a) Histopathology showing groups of and few scattered mature fat cells embedded in the collagen bundles in the superficial dermis along with minimal perivascular lymphocytic infiltrates [H&E, 40X]; b) Higher power of the same showing aggregates of mature fat cells in the superficial dermis [H&E, 100X].

Based on clinicopathological correlation, nevus lipomatosus cutaneous superficialis was diagnosed. Patient was reassured regarding the benign nature of the lesions and given the choice of surgical removal but he refused. A course of oral Augmentin 625mg twice a day for 7 days was given for the traumatic ulcer with secondary bacterial infection.

DISCUSSION

NLCS is a rare, sporadic, benign skin growth, which is distinguished by the occurrence of ectopic mature fat cells in the dermis.

Clinically, there are two types of presentation [2]. The more common classical presentation manifests in the first 3 decades of life or at birth as numerous yellow papules and nodules that are soft, pedunculated, non-tender, and frequently coalesce into a plaque. The less common solitary form manifests as a sessile or dome shaped papule occurring after the 3rd decade. Rare presentation of NLCS can be linear, bilateral or zosteriform in its distribution [2]. Surface changes like ulceration, secondary infection and hair growth have been described. Typically, NLCS involves pelvic girdle. However, abdomen, chest and face involvement are not uncommon. Only a handful of cases are reported on unusual sites like scrotum, vulva, pinna, and neck. Previous studies have reported cases of NLCS on the neck from India [4,5].

The etiology of NLCS is still unclear. Various explanations have been proposed, such as the concept of metaplasia wherein alterations in connective tissue, causes fat deposition in the dermis [1], ectopic adipocytes originating from pericytes of dermal vessels or mononuclear cells in perivascular zone [3] and adipocytes representative of a true nevus secondary to heterotopic adipose tissue found focally [6]. Groups and strands of adipocytes buried in the reticular dermis, which may reach the papillary dermis without extending into subcutaneous fat tissue, are the histological hallmark of this disease [3]. Dermoscopic features include cerebriform surface, yellow structureless areas representing dermal adipocytes, regular pigment network due to preservation of normal rete ridge anatomy. Baraldi et al. described that dermoscopic diagnosis may be aided by the presence of linear loop-like or linear-coiled structures [7].

Histopathological features show that the mature adipocytes can be solitary or in small groups around eccrine glands or blood arteries, or they can form between collagen bundles. Fatty tissue can make up as much as 50% of the dermis, though the percentage varies widely. Although a biopsy is the gold standard of diagnosis, magnetic resonance imaging and ultrasonography could aid in the diagnosis.

The classical form is not typically associated with other malformations, but café au lait macules, folliculosebaceous cystic hamartoma, nevus sebaceous of Jadassohn, basal cell carcinoma has been reported to occur alongside. There is no evidence suggesting malignant transformation so far. NLCS needs to be distinguished from hairy nevus, skin tags, nevus sebaceous, plexiform neurofibroma, Goltz syndrome, lipoblastomatosis, lymphangioma, hemangioma, and mucinous nevus [8]. Dermal aggregates of adipocytes have also been seen in some melanocytic nevi, Goltz syndrome and lipofibromas.

The primary indication for treatment is cosmetic. Surgical excision is curative and recurrences are uncommon. Carbon dioxide laser and cryotherapy have been found to be useful. Intralesional phosphatidylcholine injection has also been attempted [9].

CONCLUSION

As NLCS is a rare, benign condition requiring high clinical suspicion and expertise to identify it in its early stages for the reason that it can grow to reach enormous sizes, leading to medical, aesthetic and psychological concerns. The distinctive histopathological findings help in clinching the diagnosis and excision is the ultimate cure. Reconstruction of the defect can be more conservative if the lesion is removed promptly, early, and with less invasiveness.

Acknowledgement

No external funding was received for this study.

Declaratıon

The authors declared no conflict of interest.

Informed Consent

Written informed consent was obtained from the patient for publication of this case report.

References

1) Hoffmann E, Zurhelle E. Über einen Naevus lipomatodes cutaneaous superficialis der linken Glutaalgegend. Arch Dermatol Syphilol. 1921;130:327-33.

2) Yap FB. Nevus lipomatosus superficialis. Singapore Med J. 2009;50(5):e161-e162.

3) Jones EW, Marks R, Pongsehirun D. Naevus superficialis lipomatosus. A clinicopathological report of twenty cases. Br J Dermatol 1975;93:121-33.

4) Das JK, Sengupta S, Gangopadhyay AK. Nevus lipomatosus superficialis over neck, an atypical site. Indian J Dermatol Venereol Leprol. 2006;72(1):66-67. doi:10.4103/0378- 6323.19729

5) Kulkarni S, Chhabra N, Prabha N, Hussain N, Bugalia A. Nevus Lipomatosus Cutaneous Superficialis on the Nape -A Rare Presentation. Skinmed. 2018;16(3):189-190

6) Kim YJ, Choi JH, Kim H, Nam SH, Choi YW. Recurrence of Nevus Lipomatosus Cutaneous Superficialis after CO2 Laser Treatment. Arch Plast Surg. 2012; 39:671–673.

7) Baraldi C, Barisani A, Fanti PA, Patrizi A. Clinical, dermoscopic and histopathological features of solitary nevus lipomatosus cutaneous superficialis. Indian J Dermatol Venereol Leprol 2021;87:399-401.doi:10.25259/IJDVL_399_19

8) Kim E JJS, Jo S J, Cho K H. A case of mucinous nevus clinically mimicking nevus lipomatosus superficialis. Ann Dermatol. 2014;26(04):549–550.

9) De Paula Mesquita T, de Almeida H L, Jr, de Paula Mesquita M C. Histologic resolution of naevus lipomatosus superficialis with intralesional phosphatidylcholine. J Eur Acad Dermatol Venereol. 2009;23(06):714–715.