Nevus lipomatosus superficialis on the neck: an unusual location

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Nevus lipomatosus superficialis (NLS) is a hamartomatous skin lesion defined by the presence of mature adipose tissues among the collagen bundles of the dermis. It is classified into two forms: the classical form and the solitary form. The classical NLS most commonly involves the pelvic or gluteal region. In this paper, we report a case of classical NLS over the neck because of its atypical site. In addition, our patient had some uncommon features of NLS such as the presence of comedo-like lesions on the plaque and a foul-smelling discharge.

Keywords: hamartoma, neck, nevus lipomatosus superficialis, skin tumor

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INTRODUCTION

Nevus lipomatosus superficialis (NLS) is an uncommon, benign hamartomatous skin tumor defined by dermal deposition of mature adipose tissue that was first reported by Hoffamn and Zurhelle in 1921 [1]. It is classified into the solitary form and the classical Hoffmann-Zurhelle from. The former type usually appears at a later stage of life, usually after the age of 20 years [2,3], as a single papule or nodule with a wider distribution [4]. The latter type generally presents at birth or within the first two decades of life with multiple, non-tender, soft, cerebriform, and skin colored or yellow papulonodules which are located most commonly on the lower trunk, pelvic girdle, gluteal region, and thigh [5,6]. In this paper, we report a case of classical NLS on the neck because of its unusual location and the presence of some uncommon features of NLS.

CASE REPORT

A 17-year-old boy presented to the Department of Dermatology with a compliant of multiple and skin colored nodules on the left side of the neck. He had noticed these nodules appeared 3 years ago which slowly enlarged and coalesced together, finally forming a plaque. Also, he had a history of intermittent foul-smelling discharge from the lesion. There was no family history of similar skin lesions. There was no systemic symptom or neurological abnormality. Physical examination revealed multiple nodules on the left side of the

neck with a linear distribution. The lesions were soft, skin colored and cerebriform, coalescing into a plaque, with multiple comedo-like lesions over it (Figure 1). There was a little tenderness on pressure. Other examinations were normal and laboratory tests revealed no abnormality. A skin biopsy was performed. Histopathological examination of the biopsy specimen showed an unremarkable epidermis with slight papillomatosis and a cystic hair follicle (comedone). The strands of mature fat cells embedded among the collagen bundles were seen in the reticular and also papillary dermis (Figure 2). Based on the clinical and histopathological features of the skin lesion, the diagnosis of NLS was made.

DISCUSSION

Nevus lipomatosus superficialis (NLS) is an uncommon, hamartomatous skin tumor defined by dermal deposition of adipose tissue [1]. Its precise pathogenesis is unknown. A proposed pathogenesis is that the precursor cells around dermal blood vessels give rise to mature fat cells [7]. The classic form of NLS is characterized by groups of multiple, non-tender, soft, and cerebriform nodules that coalesce to form a unilateral plaque with a zosteriform, segmental, or linear distribution [6], as in our case. They are most commonly located on the lower trunk and the pelvic girdle [5]. Rare involvement of the abdomen, chest, and face or

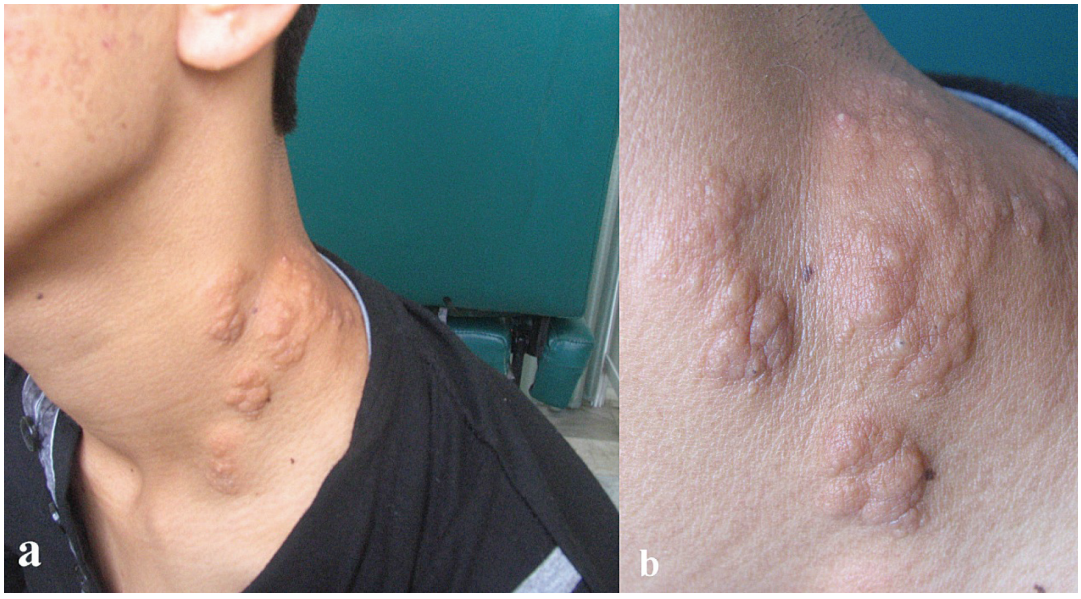

Figure 1. (a) Photograph shows the left side of the neck with multiple, soft and cerebriform nodules coalescing into a main plaque. **(b)** There are multiple comedo-like lesions over the plaque.

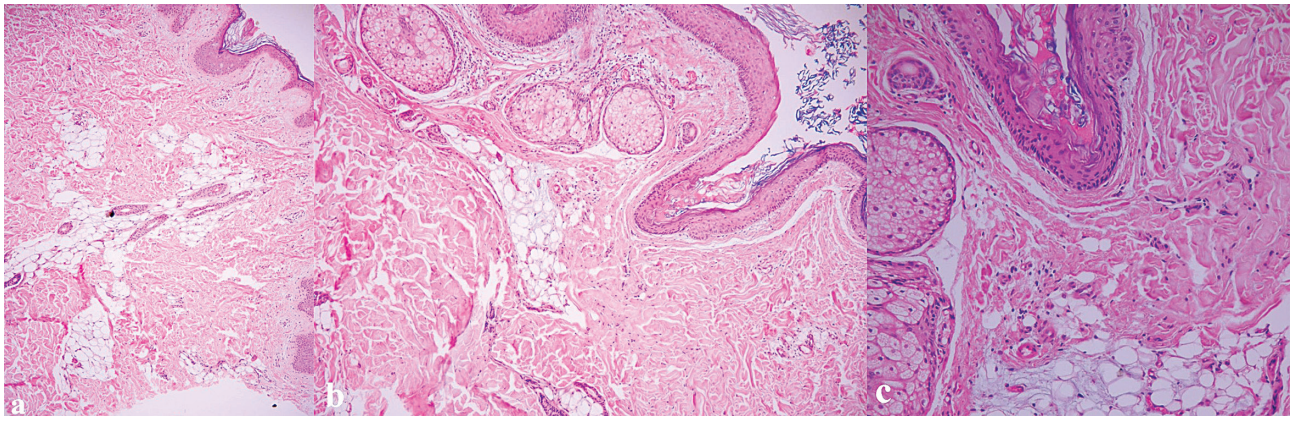

Figure 2. Aggregation of adipose tissues in the upper dermis intermingled with collagen (H&E (a) ×10, (b) ×20 and (c) ×40 respectively)

scalp [6,8,9] has also been reported. Moreover, Dos [10] and Khandpur [2] reported NLS over the neck as a rare and atypical site of involvement, as in our case. Ulceration may occasionally occur [11]. A foul-smelling discharge has been rarely reported [3,6], as seen in our patient. Other coexistent cutaneous disorders such as angiokeratoma of Fordyce [2], café-au-lait macules, scattered leukoderma, overlying hypertrichosis, and comedo-like lesions [6,12] have also been reported rarely. The latter was seen in our patient.

NLS should be differentiated from nevus sebaceous, neurofibroma, connective tissue nevus, angiolipoma, lymphangioma, and focal dermal hypoplasia [6,11]. Histopathological evaluation is required for differentiation. The histopathology of NLS usually shows aggregation of mature fat cells embedded among the collagen bundles in the dermis, often with extension to the papillary dermis. The proportion of the fatty tissue is variable from < 10% of the dermis to > 50%. Apart from the presence of fat tissue, the dermis is otherwise normal. However, in some cases, the density of the collagen bundles and vascularity is more than normal [13].

Our patient was disinclined to receive any treatment. Since no malignant alteration has been reported, treatment is usually not necessary other than for cosmetic reasons. The treatment of choice is surgical excision and recurrence is rare [3,14]. If surgery is not an option, alternative therapies include cryotherapy [2] or CO2 laser [15]. In conclusion, we reported a case of classical NLS on the neck because of its unusual location and the presence of rare features like a foul-smelling discharge and comedo-like plugs on its surface.

REFERENCES

1. Hoffmann E, Zurhelle E. Über einen naevus lipomatodes cutaneus superficialis der linken glutaalgegend. Arch Dermatol Syphilol 1921;130:327-33.
2. Khandpur S, Nagpal SA, Chandra S, et al. Giant nevus lipomatosus cutaneous superficialis. Indian J Dermatol Venereol Leprol 2009;75:407-8.
3. Yap FB. Nevus lipomatosus superficialis. Singapore Med J 2009;50:161-2.
4. Bancalari E, Martínez-Sánchez D, Tardío JC. Nevus lipomatosus superficialis with a folliculosebaceous component: report of 2 cases. Patholog Res Int 2011; 2011:105973.
5. Goucha S, Khaled A, Zéglaoui F, et al. Nevus lipomatosus cutaneous superficialis: report of eight cases. Dermatol Ther (Heidelb) 2011;1:25-30.
6. Ghosh SK, Bandyopadhyay D, Jamadar NS. Nevus lipomatosus cutaneous superficialis: an unusual presentation. Dermatol Online J 2010;16:12.
7. Kaddu S, Kohler S. Muscle, adipose and cartilage neoplasms. In: Bolognia JL, Jorrizzo JL, Schaffer JV, editors. Dermatology. 3rd ed. Mosby: Elesivier; 2012: 1988.
8. Chanoki M, Sugamoto I, Suzuki S, et al. Nevus lipomatosus cutaneus superficialis of the scalp. Cutis 1989; 43:143-4.
9. Park HJ, Park CJ, Yi JY, et al. Nevus lipomatosis superficialis on the face. Int J Dermatol 1997;36:435-7.
10. Das JK, Sengupta S, Gangopadhyay AK. Nevus lipomatosus superficialis over neck, an atypical site. Indian J Dermatol Venereol Leprol 2006;72:66-7.
11. Ranjkesh MR, Herizchi QH, Yousefi N. Nevus lipomatosus cutaneous superficialis: a case report with histologic findings. J Turk Acad Dermatol 2009; 3: 93103c.
12. Dhar S, Kumar B, Kaur I. Naevus lipomatosus superficialis of Hoffman and Zurhelle. Indian J Dermatol Venereol Leprol 1994;60:39-40.
13. Ragsdale BD. Tumors with fatty, muscular, osseous, and/or cartilaginous differentiation. In: Elder DE, Elenitsas R, Jhonson BL, Murphy GF, Xu X, editors. Lever's histopathology of the skin. 10th ed. Wolters Kluwer-Lippincott Williams & Wilkins, New Delhi; 2009: 1057-106.
14. Pujani M, Choudhury M, Garg T, et al. Nevus lipomatosus superficialis: A rare cutaneous hamartoma. Indian Dermatol Online J 2014; 5: 109-10.
15. Fatah S, Ellis R, Seukeran DC, et al. Successful CO2 laser treatment of naevus lipomatosus cutaneous superficialis. Clin Exper Dermatol 2010; 35: 559-60.