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Solitary Nevus Lipomatosus Cutaneous Superficialis Presenting at Birth

Mary Mathew[1], Bhavna Nayal[2*], Padma Priya J[3], Mohammed Shakir Ali[4]
[1]Professor,[2]Assistant Professor, [3]Associate Professor,[4]Junior Resident, Department of Pathology, KMC, Manipal University, Manipal, India

***Corresponding Author:**
Name: Dr. BhavnaNayal
Email: bhavnayal@yahoo.com

Abstract: Nevus lipomatosus cutaneous superficialis (NLCS) is a rare hamartomatous skin condition characterized by the presence of adipose tissue in the upper dermis. This condition is classified into classical multiple form and the solitary form based on clinical presentation. These lesions are present at birth or appear within the first three decades of life; however, solitary forms tend to occur later in life. Excision of these lesions is curative. Due to rarity of this entity, a high clinical suspicion is required for diagnosis of the same. We report a rare case of solitary NLCS presenting at birth.
Keywords: Nevuslipomatosussuperficialis, Ectopic adipose tissue, Dermal lesion.

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare, idiopathic hamartomatous lesion of the skin. It is characterized by the presence of dermal ectopic adipocytes or mature adipose tissue[1,2]. Clinically, 2 forms have been described: multiple (classical Hoffman-Zurhelle) and solitary [1]. These lesions are present at birth or appear within the first three decades of life; however, solitary forms tend to occur later in life. No familial predisposition or sex preponderance has been described for this condition [3,4]. We present a rare case of solitary NLCS with lesion present since birth.

CASE REPORT

A 10-year-old Indian boy presented with asymptomatic, progressively increasing mass over the back since birth. There was no history of ulceration or discharge at the site of the lesion. He had no significant family history or systemic complaints. Examination revealed a single mass measuring 9x3.5cm on the lower back. The overlying skin surface was thickened and wrinkled. Clinical differentials include lipoma and tuberous sclerosis. The lesion was excised and sent for histopathological examination. Gross examination showed a skin covered cerebriform mass. Cut section of the lesion showed yellow fatty areas and fibrosis. Microscopy revealed a pigmented epidermis with focal acanthosis and papillomatosis. The papillary dermis showed ectopic mature adipocytes with mild perivascular lymphocytic infiltrate (Figures 1 & 2). A diagnosis of solitary nevus lipomatosus cutaneous superficialis was made on the basis of clinical and histopathological features.

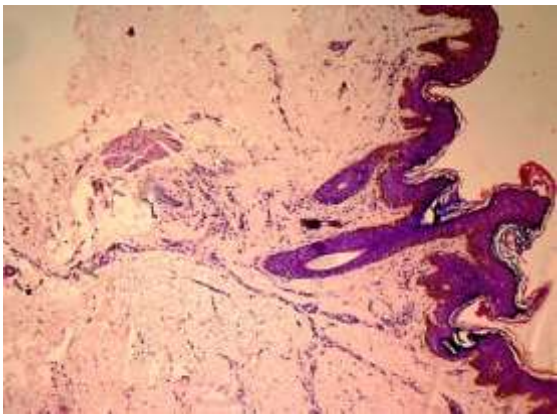

Fig. 1: Pigmented acanthotic epidermis with dermal ectopic adipose tissue (H&E, X10)

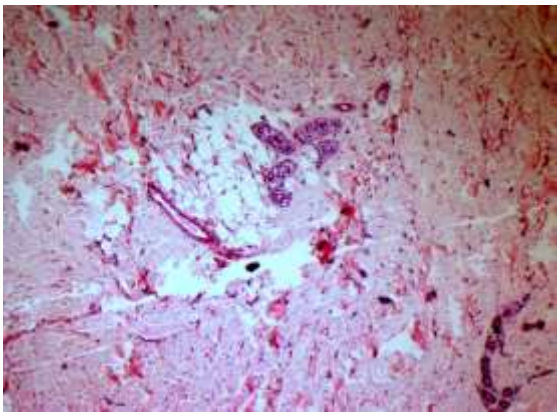

Fig. 2: Periadnexal and perivascular adipocyte tissue (H&E, X20)

DISCUSSION

Nevus lipomatosus cutaneous superficialis (NLCS) is an uncommon benign hamartomatousconnective tissue nevus with mature adipose tissue in the dermis and was first reported by Hoffmann and Zurhelle in 1921 [5]. Since then, few cases have been reported in literature.

Two clinical forms have been described. The classical Hoffman-Zurhelle type is characterized by multiple, asymptomatic flesh coloured or yellow lesions which may coalesce forming plaques with a smooth or cerebriform surface [4]. These lesions usually appear at birth or the first few decades of life and commonly affect the lower trunk and upper posterior thigh. Few cases presenting much later in life and involving scalp and face have also been reported [2,3]. The solitary form on the other hand, usually appear later in life during the third to sixth decade of life. There is no specific site predilection unlike the classical form [3]. Some cases presenting with lesions in childhood have also been reported [6, 7].

Giant forms of NLCS with lesions assuming very large sizes have been documented [8,9]. NLCS may be associated with café-au-lait spots, hypopigmented or leukodermic spots, hypertrichosis and ulceration with necrosis [1,3,10].

Our case had the characteristic clinical presentation of a solitary type of NLCS with absence of hypopigmentation or secondary changes in the lesion. However, unlike most reported cases, the lesion manifested at birth.

Histopathological examination of these lesions reveal dermal collagen bundles with intervening, predominantly perivascular isolated adipocytes or mature adipocytes showing no connection with the underlying subcutaneous fat. The ectopic adipose tissue may comprise 10-50% of the lesion [3,8,10].

It has been hypothesized that the origin of ectopic adipose tissue could possibly be degenerating dermal connective tissue with adipose tissue metaplasia, developmental displacement and perivascular differentiating lipoblasts [1].

The differential diagnosis of NLCS includes nevus sebaceous, lymphangioma, lipoma, neurofibroma, pedunculatedlipofibromas, hemangioma, giant fibroepithelial polyp and focal dermal hypoplasia (Goltz syndrome). Absence of congenital anomalies and characterstic microscopic features helps in the diagnosis of NLCS [2,3,8].

Due to the absence of malignant transformation and rare recurrence of these lesions, treatment is not indicated other than for cosmetic purposes. Excision of these lesions are however curative [1,2].

CONCLUSION

To conclude, NLCS are benign skin malformations with excellent prognosis. Due to rarity of this entity, a high clinical suspicion is required for diagnosis of the same.

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