Nevus lipomatosus superficialis

Yap F B B

ABSTRACT

Nevus lipomatosus superficialis is a rare benign skin malformation characterised by ectopic adipocytes in the upper dermis. It is classified into two types: a classical Hoffman-Zurhelle type and the solitary type. A case of the classical type with multiple soft, non-tender, pedunculated, cerebriform, skin-coloured papules and nodules over the right lower back is presented in a 21-yearold Malay woman. She had a rare occurrence of ulceration and necrosis of the lesions.

Keywords: benign skin malformation, dermal lesion, ectopic adipose tissue, nevus lipomatosus superficialis, skin papule

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INTRODUCTION

Nevus lipomatosus superficialis is a rare benign developmental disorder characterised by isolated ectopic adipose tissue in the dermis, with predilection to the pelvic girdle. It was first reported by Hoffman and Zurhelle in 1921.⁽¹⁾ A case of nevus lipomatosus superficialis with a rare occurrence of ulceration is reported.

CASE REPORT

A 21-year-old Malay woman was referred for multiple skin-coloured soft nodules on the right lower back. She first noticed the skin lesions when she was five years of age. They progressively enlarged as she grew. Her current presentation was foul-smelling, purulent discharge from the upper part of the lesions. She recalled that she had a similar episode of purulent discharge which had resolved with antibiotics three years ago. She was the youngest of three siblings of non-consanguineous parents. She has no family history of similar skin lesions. She is currently pursuing computer science studies in a local college.

On examination, she was a healthy, thin woman. There were multiple skin-coloured, non-tender, soft, compressible cerebriform nodules coalescing into a plaque, and forming the main lesion in an area measuring 8 cm \times 5 cm on the right lower back. There were also multiple satellite papules and nodules measuring 0.5–3



Fig. 1 Photograph shows multiple soft cerebriform nodules coalescing to form the main lesion, with two small ulcers on the right lower back. Multiple satellite nodules and papules were noted.

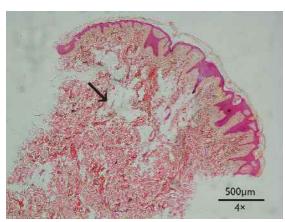


Fig. 2 Photomicrograph shows clumps of adipose tissues in the upper dermis (arrow) (Haematoxylin & eosin, × 4).

cm in diameter from the main lesion. A few nodules on the main lesion had ulcerated with purulent discharge (Fig. 1). There was no café-au-lait spot, hypopigmented macule or neurological abnormality noted.

A skin biopsy taken from one of the nodules showed normal epidermis with groups of mature adipose tissues in the upper dermis, interposed with the collagen bundles (Fig. 2). There was no communication between the ectopic adipose tissue and the subcutaneous fat tissue. Based on the findings, she was diagnosed to have nevus lipomatosus superficialis. Culture of the purulent discharge was negative. Nevertheless, she was given one week of cloxacillin prior to the culture result. The antibiotics did not help the discharging wound, and thus, total excision of the nevus was undertaken. At six

Department of Dermatology, Sarawak General Hospital, Jalan Hospital, Kuching 93586, Malaysia

Yap FBB, MD, MRCP Physician and Clinical Specialist

Correspondence to: Dr Felix BB Yap Tel: (60) 82 276 666 Fax: (60) 82 276 530 Email: woodzlamp @yahoo.com months post-excision, she had no reccurrence of the nevus. It was concluded that she had nevus lipomatosus superficialis with necrosis and ulceration of the lesion.

DISCUSSION

Nevus lipomatosus superficialis is a rare idiopathic benign skin malformation.^(2,3) Clinically, it is classified into the classical Hoffmann-Zurhelle form and the solitary form. In Tunisia, Triki et al noted a predominance of the solitary form.⁽⁴⁾ The solitary form usually occurs after the age of 20 years, with no particular predilection sites. It presents with a single nodular lesion with no predilection site. However, it had been reported to occur on the face, scalp and knee.⁽⁵⁻⁷⁾ The classical form occurs at birth or during the first three decades of life.^(2,7,8) This Hoffmann-Zurhelle form manifests with groups of multiple, soft, non-tender, pedunculated, cerebriform, yellowish or skin-coloured papules or nodules. They often coalesce to form a plaque lesion, usually situated on the pelvic girdle area in a zonal pattern.^(2,3) They are usually unilateral. The patient in this case had a classical form with multiple nodular lesions on the right lower back

This case was interesting in that the patient had ulceration of the lesion with foul-smelling discharge. This occurrence is rarely seen. It is postulated to be due to the compression of the dermal blood vessels by the ectopic fat cells.⁽³⁾ Histologically, there are clusters of ectopic mature adipose tissues among the collagen bundles or subpapillary plexus in the upper dermis.^(2,4) The possible aetiologies for the ectopic adipocytes include adipose metaplasia in a degenerative dermal connective tissue, developmental displacement and perivascular differentiating lipoblasts. This patient had the typical histological changes of nevus lipomatosus superficialis. Nevus lipomatosus superficialis should be differentiated from neurofibroma, fibroepithelial polyp, lymphangioma, haemangioma and focal dermal hypoplasia. Histological studies will usually help the differentiation. Treatment is usually not necessary. This patient had surgical excision of the lesions because of the ulceration, and for cosmetic reasons. Excision is curative as recurrence is rare postexcision.

In conclusion, nevus lipomatosus superficialis is a rare skin malformation. A high index of suspicion is needed to diagnose this benign condition. This lesion rarely ulcerates, so this complication must be taken into account for those presenting with discharge from the lesion.

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