A Rare Presentation of Multiple Dermatofibromas in a Healthy Patient

[1]**Mahmood KAMAL** *MBBS*, [1,2] **Olusola AYANLOWO** *MBBS, MSc, FWACP*, [1,2]
Ayesha AKINKUGBE *MBBS, FWACP*, [1,2] **Erere OTROFANOWEI** *MBBS, FMCP*
[1]Dermatology Unit, Department of Medicine, Lagos University Teaching Hospital, Idi-Araba, Lagos
[2]Dermatology Unit, Department of Medicine, Faculty of Clinical Sciences, College of Medicine, University of Lagos
Corresponding Author: Dr Mahmood Kamal
Email: kamalmamud@gmail.com; ***Phone number:*** +234 7032448261

ABSTRACT

Dermatofibromas rarely present as its multiple variant, which is the occurrence of more than fifteen lesions in an individual. There are limited reported cases of multiple dermatofibromas in the literature. This is a case report of a 19-year old female with a 10-year history of slowly progressive multiple dermatofibromas involving the upper and lower limbs, back and the face.

Keywords: Dermatofibroma, Multiple eruptive dermatofibromas

Une Présentation Rare de « Dermatofibromes Multiples » Chez un Patient en Bonne Santé

ABSTRAIT:

Les dermatofibromes se présentent rarement dans ses multiples variantes, c'est-à-dire la survenue de plus de quinze lésions chez un individu. Il existe peu de cas rapportés de dermatofibromes multiples dans la littérature. C'est un rapport de cas d'une dame de 19 ans avec des antécédents de 10 ans de dermatofibromes multiples lentement progressifs impliquant les membres supérieurs et inférieurs, le dos et le visage.

Mots clés: Dermatofibrome, Dermatofibromes éruptifs multiples

Introduction

A dermatofibroma is a slow growing cutaneous tumour of mesenchymal origin. Predominantly it involves the dermis, but rarely originates from subcutaneous tissue (benign fibrous histiocytoma).[1] It is a disease of young adults and middle-aged group, and is more common in females.[2] It represents about 3% of skin biopsy specimens received at dermatology laboratories.[3] The aetiology is unclear but thought to represent an abortive immunoreactive process mediated by dermal dendritic cells.[4]

Typical presentation consists of a solitary slow-growing slightly hyperpigmented tumour, usually found on the extremities.[5] Histologic variants include granular cell dermatofibroma, clear cell dermatofibroma, palisading cutaneous fibrous histiocytoma, aneurysmal, and cellular.[5] Various dermoscopic patterns have been reported.[3] Multiple dermatofibromas are defined as occurrence of more than fifteen lesions in an individual.[6,7] Multiple dermatofibromas are rare with very few cases reported in the literature. We report a 19-year-old healthy female with multiple dermatofibromas on the upper and lower limbs, trunk and face. We also reviewed the literature on this rare dermatosis and discussed challenges in management.

Case Report: Our patient is a 19-year-old female who presented with a decade long history of slowly growing dark-coloured lesions on the thighs, which later involved the legs, lower back, arms and the face. The lesions were occasionally itchy but not painful. There was no preceding history of trauma to the affected areas and no systemic symptoms. There was no past history or family history of a similar condition, of atopy or autoimmune disease. A solitary facial lesion was surgically removed in the past before presentation without histology, and there was no recurrence of a facial lesion. She had repeatedly used topical corticosteroids on the lesions in the past with no significant improvement.

Clinical findings were confined to the skin. She had

multiple hyperpigmented dome-shaped and flat-topped firm nodules and papules of varying sizes on both upper and lower limbs, concentrated on the thighs. There were 22 nodules in total, with sizes ranging from 4mm x 4mm to 2cm x 2cm in diameter. Most of them had a smooth surface, however a few lesions on the legs were rough surfaced. They were non-tender and had a positive dimpling sign.

She was non-reactive to HIV-1 and 2, Hepatitis B and C screening

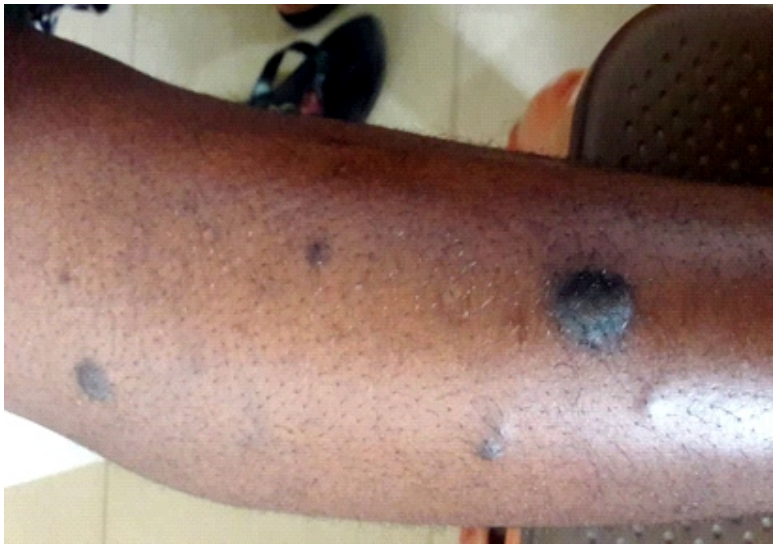

Figure 1: Multiple DF on the arms

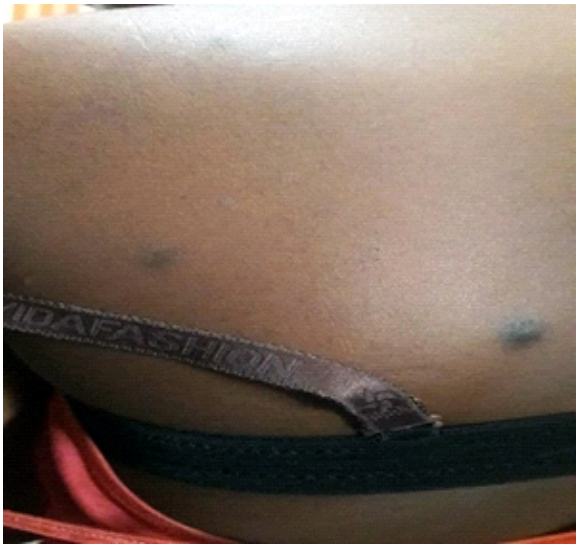

Figure 2: lesions on the back

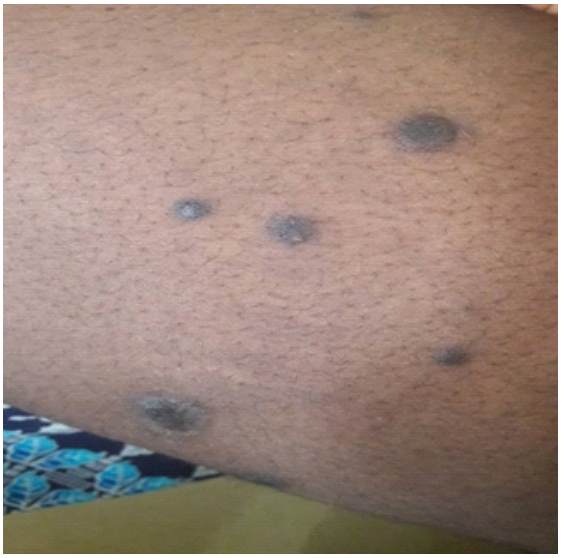

Figure 3: Multiple hyperpigmented nodules on the thigh

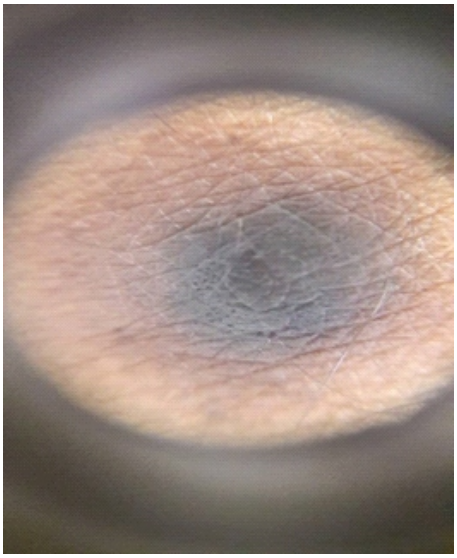

Figure 4: Dermoscopy of the nodule

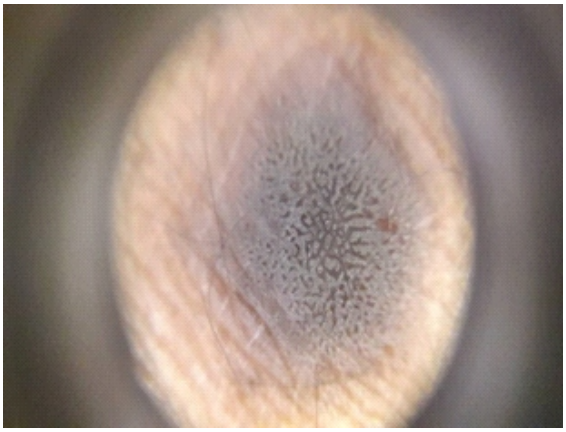

Figure 5

Dermoscopic findings: Central homogeneous hyperpigmentation and hyperkeratosis with a peripheral pigment network is the most common dermoscopic feature found in our patient (Figure 4). Other lesions showed central homogeneous hyperpigmentation and whitish streaks with peripheral pigment networks, atrophic thinly-scarred center, and pseudo hair follicles (Figure 5).

Based on the clinical findings and dermoscopy features, a diagnosis of multiple dermatofibromas was made and an excisional biopsy of a lesion on the right lower limb was performed to confirm this.

Histology Findings: A dermal lesion comprising of benign proliferation of spindle shaped cells with bland nuclei. No atypical cells were seen. The cells had elongated vesicular nuclei and there was abundance of extracellular collagenous matrix. The deep margin was free of the lesion. In view of the above, diagnosis of multiple dermatofibromas was confirmed.

The plan: she was counselled and prepared for intralesional triamcinolone injection monthly. She was however lost to follow up after the first session of intralesional injection.

Discussion

Typically, dermatofibroma is a common benign skin tumour but some of its clinical variants are rare. Clinical variants include giant, atrophic, polypoid, multiple, eruptive and clustered dermatofibroma.[8]

A dermatofibroma is a superficial (dermal) form of benign fibrous histiocytoma, and it is composed of a mixture of fibroblastic and histiocytic cells.[9] It can be described as "solitary", "several" (<5 lesions), or "multiple" dermatofibromas (>15 lesions).[10] Definition of multiple eruptive dermatofibromas (MEDFs) was proposed, arbitrarily, as "the presence of 15 DFs appearing within a period of 4 months", and usually with background immunodeficiency or autoimmune disease.[4,10] Few cases of multiple eruptive dermatofibromas in association with HIV infection have been reported in the literature.[11] However, about 30% of MEDFs have no systemic diseases.[10] Our patient has a longer duration with no clinical or laboratory evidence of immune suppression, connective tissue disease or other systemic diseases. Dermatofibromas can result from trivial trauma like insect bite.[9] No trigger factor was found in our patient.

Dermatofibromas typically occur on the lower extremities of young adult females as seen in this case being reported. Dermoscopic features as seen in our patient includes a central homogeneous hyperpigmentation with peripheral pigmentary network and this differs from most common dermoscopic features reported in literature, which is a central white patch with peripheral pigment network.[3] Other dermoscopic features reported in literature include white scar-like patch, brown dots and globules, pigment network, irregular crypts and pseudofollicular openings, homogeneous pigmentation, and vascular structures.[12,13] Camara et al. reported a homogenous pigmentation as the third most common dermoscopic finding in their study,[13] and this was the most prominent feature seen in our patient. Also, pseudofollicular openings seen in our patient is one of the specific dermoscopic features reported in literature. However, the atrophic, thinly scarred centre seen in some of the lesions could be as a result of prior topical corticosteroid used by the patient.

Benign fibrohistiocytoma (BFH) originating from subcutaneous tissue which can mimic dermatofibrosarcoma usually requires immunohistochemical staining (CD34, factor XIII) to confirm the diagnosis.[1] However, diagnosis of dermatofibroma can be made from typical clinical features, dermoscopy and histological examination.

The main reason for the treatment of dermatofibromas is usually cosmetic except in rare cases when it becomes symptomatic. Many treatment options for multiple dermatofibromas have been tried with varying outcomes.

Surgical excision is the definitive treatment especially when the diagnosis of benign dermatofibroma is not certain, or when an aggressive subtype is suspected.[14] however, this can result in more conspicuous scar formation. Kim et al. successfully treated 151 dermatofibroma patients with 3mm margin complete surgical excision with no recurrence during the follow up period.[14] Shave biopsy is also a form of surgical treatment but not completely successful.[15]

Cryotherapy is an effective well tolerated option

with a more cosmetically acceptable outcome but higher recurrence rate.[16] It requires a short preparation time, has low risk of infection and minimal wound care. Some of the possible complications of cryotherapy include bleeding, blister formation, loss of hair and hypopigmentation.[17] Lamgan et al. reported 27 dermatofibroma cases treated with cryotherapy with and without topical corticosteroid, with more than 90% good-excellent outcome.[16]

Laser therapy is one of the more recent treatment options for the management of dermatofibromas with good cosmetic outcome and patient satisfaction. Cases of non-ablative pulsed dye laser (PDL) and ablative carbon dioxide laser treated dermatofibroma have been reported.[18,19] Alonso et al treated 15 dermatofibroma cases with non-ablative PDL, with global clinical improvement in more than 50%.[19] Common side effect associated PDL is post-inflammatory hyperpigmentation, which usually resolves within six months.[20] Combination of fractionated carbon dioxide laser with topical corticosteroid has been shown to be very effective in treating symptomatic dermatofibroma.[18] Ultrapulse (UP) mode of carbon dioxide laser is more effective than continuous wave (CW) and super-pulse (SP) modes with minimal skin damage.[21] Carbon dioxide laser, being an ablative procedure, can be associated with pain, bleeding, pigmentary changes and scaring.[20]

Intralesional and/or systemic steroid administration, particularly where there is autoimmune association, are other treatment options.[9,22] This can result in atrophy, hypopigmentation and incomplete resolution. To avoid excessive scar formation in our patient, since she had multiple lesions and coupled with the increased risk of keloid formation in blacks, intralesional triamcinolone was favoured over surgical removal in the management of the dermatosis.

Conclusion: We have reported the case of a 19-yearold girl female with multiple dermatofibromas involving the limbs, trunk, and face. Dermoscopic features showed a homogeneous hyperpigmentation, hyperkeratosis, whitish streaks with peripheral pigment network, atrophic thinly scarred centre and pseudo hair follicles. She had the first course of intralesional triamcinolone administered as treatment but was subsequently lost to follow up.

The authors declare no conflicts of interest

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