CASE REPORT

Congenital multiple clustered dermatofibroma and multiple eruptive dermatofibromas - unusual presentations of a common entity*

Dermatofibroma múltiplo agrupado congênito e dermatofibromas múltiplos eruptivos - apresentações singulares de uma entidade comum

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Abstract: Dermatofibroma is one of the most common entities seen in dermatology clinical practice. Several clinical subtypes have nevertheless been described, all of them of uncommon occurrence. The authors present two rare clinical variants of dermatofibromas: congenital multiple clustered dermatofibroma (the presented case is the 4th congenital case to be reported so far) and multiple eruptive dermatofibromas developing in the setting of a Sjögren's syndrome. Since the uncommon subtypes may not be clinically evident, dermatologists should familiarize themselves with their main features and we advise a high level of clinical suspicion in order to reach the correct diagnosis.
Keywords: Adult children; Congenital abnormalities; Histiocytoma, benign fibrous; Sjogren's Syndrome

Resumo: O dermatofibroma é uma das entidades mais frequentemente observadas na prática clínica dermatológica. No entanto, além do dermatofibroma comum, vários subtipos clínicos de ocorrência incomum têm sido descritos na literatura. Os autores descrevem duas variantes clínicas raras de dermatofibromas: dermatofibroma múltiplo agrupado congênito (o caso apresentado é o quarto caso congênito reportado até hoje) e dermatofibromas eruptivos múltiplos no contexto de uma Síndrome de Sjögren. Estes diagnósticos menos comuns podem não ser clinicamente evidentes portanto os dermatologistas devem estar familiarizados com estas apresentações, sendo de suma importância um elevado índice de suspeita clínica.
Palavras-chave: Anormalidades congênitas; Crianças adultas; Histiocitoma fibroso benigno; Síndrome de Sjögren

INTRODUCTION

Dermatofibroma is a common finding in dermatology clinical practice. Several clinical variants of uncommon occurrence have been described, including giant, atrophic, polypoid, multiple, eruptive and clustered. The authors present two rare clinical variants of dermatofibromas: congenital multiple clustered dermatofibroma and multiple eruptive dermatofibromas.

CASE REPORT

Case Report 1:

A 12-year-old girl presented with a congenital asymptomatic cutaneous lesion on the right thigh. According to the patient, the lesion had been stable for years, but had grown in the previous year and developed new papular areas. She was otherwise healthy and was on no regular medication. Physical examination revealed multiple firm red-to-brown

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papules measuring 5-8 mm in diameter, clustered on the lateral aspect of the right thigh, some coalescing to form one large plaque. The lesions progressed to the posterior aspect of the thigh in a linear arrangement (Figure 1). A cutaneous biopsy was performed and histopathological examination revealed a hyperplastic epidermis with hyperpigmentation of the basal layer and a dermal proliferation of interlacing fascicles of spindle cells that dissociated the collagen (Figures 2 and 3). Immunohistochemistry tests were positive for vimentin and negative for α-actin, desmin and CD34. The diagnosis of congenital multiple clustered dermatofibroma was made, and a conservative attitude was adopted, keeping the patient under surveillance.

Case report 2:

A 42-year-old woman presented with a 3-month history of rapidly-developing generalized brown papules and nodules. She complained that some of these lesions were painful. Her past medical history included a Sjögren Syndrome diagnosed the previous year (xerophthalmia, xerostomia, positive anti-SSA antibodies, Schirmer test, salivary scintigraphy and minor salivary gland biopsy), for which she was on artificial tears and regular oral fluoride treatments. Physical examination revealed multiple (20-30) firm brown papules and nodules of 5-15 mm in diameter, with positive dimple sign, distributed on the trunk, upper and lower limbs (Figures 4 and 5). Histopathological examination showed features similar to the ones found on the previous patient: a spindle cell proliferation in the dermis that dissociated the collagen, superimposed by a hyperplastic epidermis with hyperpigmented basal layer; these features were compatible with a dermatofibroma. Laboratory tests revealed the changes expected to be found in a patient with Sjögren's Syndrome - increased erythrocyte sedimentation rate (38 mm, normal 0-19), anti-SSA antibodies (240,0 U/μL, normal < 10) and antinuclear antibodies (1/640, normal < 1/80). The diagnosis of multiple eruptive dermatofibromas in a patient with Sjögren's Syndrome was made. Surgical excision of the painful lesions was performed and the patient has been kept under surveillance.

DISCUSSION

Common dermatofibroma is a frequent cutaneous condition that can affect people of any age, but it usually develops in the 20-30 age range.[1] The lesions are isolated, single or in reduced number, with predilection for the lower limbs. Diagnosis is clinical and usually does not pose diagnostic difficulties. No treatment is recommended unless it is accompanied by discomfort, which is uncommon.[1] There is no association with systemic diseases. On the other hand,

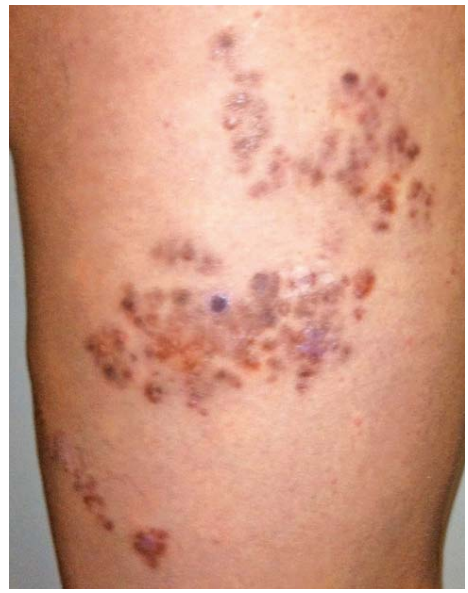

Figure 1: Clinical features of the cutaneous lesion on the lateral aspect of the right thigh

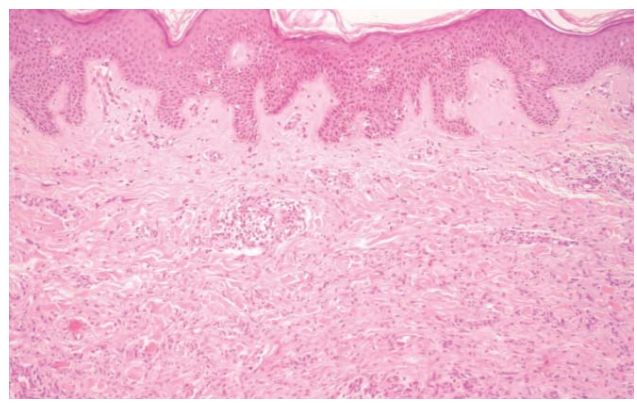

Figure 2: Histopathologic examination showing a hyperplastic epidermis with hyperpigmentation of the basal layer and a dermal proliferation of interlacing fascicles of spindle cells that dissociated the collagen (hematoxylin and eosin, original magnification 100x)

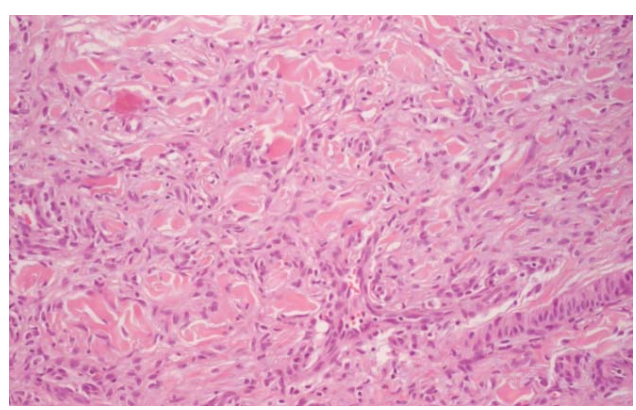

Figure 3: Amplification of the interlacing fascicles of spindle cells dissociating the collagen (hematoxylin and eosin, original magnification 200x)

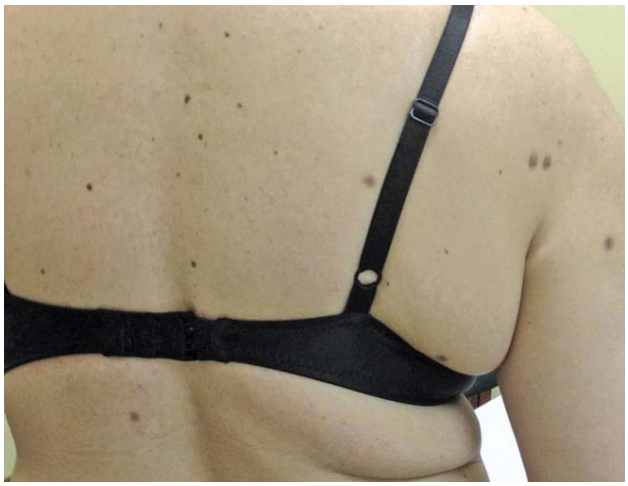

Figure 4: Lesions distributed on the back

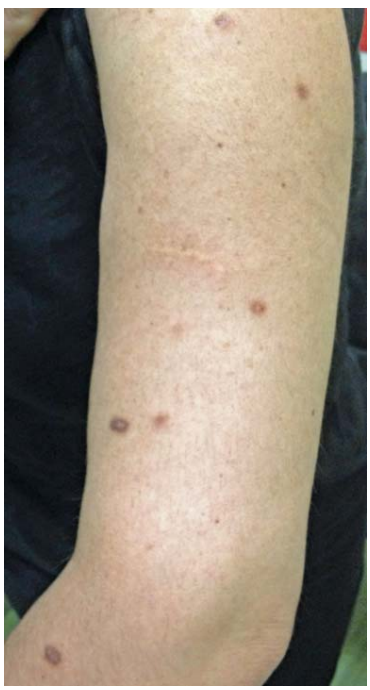

Figure 5: Multiple dermatofibromas on the right arm

multiple clustered dermatofibroma (MCD) is of exceptional occurrence.[2] Lesions normally appear during puberty and continue to develop over the years.[2] Congenital MCD is exceedingly rare and, to our knowledge, our patient is only the 4th congenital case reported so far in the international literature.[3-5] Clinically, it is characterized by the development of multiple asymptomatic dermatofibromas clustered in one anatomical area, mainly on the lower limbs, that usually merge into one plaque or are arranged in a linear pattern.[2-5] A cutaneous biopsy is essential since the diagnosis depends upon histopathological examination, which shows the same features found in common dermatofibroma.[2-6] Treatment consisting of complete surgical excision is directed by related discomfort or for aesthetic reasons. No association with systemic disorders has been reported to date.[2-5] Multiple eruptive dermatofibromas (MED) refer to a rare entity that by definition is present when there are > 15 dermatofibromas (multiple) and when 5-8 dermatofibromas develop over a 4-month period (eruptive).[7] Both these requirements were fulfilled in our patient. Any age can be affected, but individuals aged 20-30 are mostly affected, and distribution over the body is generalized.[7-9] The lesions are usually painful. Diagnosis is made on clinical grounds and treatment is symptomatic, frequently involving surgical excision of the painful lesions.[7-9] MED have been associated with auto-immune diseases, immunosuppressant drugs, HIV, hematologic malignancies and pregnancy, among others. Therefore the development of MED should prompt investigation of an underlying disease.[7-9] It is thought that an impaired immune function may play an important role in its development, although the underlying mechanisms have not been completely elucidated.[7-9] In the presented case the most probable association is with Sjögren Syndrome, creating the microenvironment of immune dysfunction needed for such lesions to develop. The unusual nature of these cases resides in the fact that such a common entity as dermatofibroma can present with so many different and uncommon clinical features, thereby sometimes making diagnosis difficult and the cause not always immediately evident. Since the uncommon subtypes may not be clinically evident, dermatologists should familiarize themselves with the main features and maintain a high level of clinical suspicion. A cutaneous biopsy is vitally important and the clinico-pathologic correlation is crucial for correct diagnosis. ❑

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