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Brief communication

The dermatoscopy in the skin pathergy testing: case series in patients with suspected Behçet's Disease



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ABSTRACT

Behçet's disease is a multisystemic disease consisting of a varying combination of ocular, mucocutaneous, neurologic, cardiovascular, gastrointestinal and other manifestations. Its diagnosis is based on clinical criteria, in which a positive pathergy test scores 1. A case series with 26 suspected patients is presented, and the skin pathergy test was performed in 23. The results were read in 48 hours, and they were considered negative when without papule, and positive with a papule or pustule. Positive results were divided by papule size, and dermatoscopy was done to measure and observe its clinical aspects. After the readings, a biopsy was performed, with annotation of histopathological aspects. The test was negative in 2 (8.7%) and positive in 21 (91.3%) patients. The results and the literature review are presented.

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Dermatoscopia no teste cutâneo da patergia: série de casos de pacientes com suspeita de Doença de Behçet

RESUMO

A doença de Behçet é uma doença multissistêmica que consiste de diferentes combinações de manifestações oculares, mucocutâneas, neurológicas, cardiovasculares, gastrintestinais e outras. Seu diagnóstico se fundamenta em critérios clínicos, em que o teste da patergia positivo recebe um ponto. Apresenta-se uma série de casos com 26 pacientes suspeitos, tendo o teste da patergia da pele sido realizado em 23 deles. Os resultados foram avaliados em 48 horas, tendo sido considerados como negativos diante da ausência de pápula e positivos na presença de pápula ou pústula. Os resultados positivos foram divididos pelo tamanho da pápula, efetuou-se uma dermatoscopia para medir e observar seus aspectos clínicos. Após as leituras, foi realizada uma biópsia, com anotação dos

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aspectos histopatológicos. O teste foi negativo em 2 (8,7%) e positivo em 21 (91,3%) pacientes. Apresentam-se os resultados e a revisão da literatura.

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Introduction

Behçet's disease (BD) is classified among vasculitides.¹ It was first defined as a triad of recurrent aphtous stomatitis, genital aphtae and relapsing uveitis in 1937 by Hulusi Behçet. It is considered a multisystemic disease consisting of varying combination of ocular, mucocutaneous, neurologic, cardiovascular, gastrointestinal and other manifestations.²⁻⁷

Its diagnosis is based on clinical criteria. The New International Criteria for Behçet's Disease are one of the most recently revised diagnostic criteria. A diagnosis of BD consists of a sum of three or more points according to a score system. Positive pathergy test scores 1 point (Table 1).⁸

Pathergy is a hyper reactivity of the skin after a needle trauma. It was first described in 1937 and it is considered pathognomonic, although it can be seen in pyoderma gangrenosum, erythema elevatum diutinum and other neutrophilic dermatoses, including Sweet syndrome and the blind loop syndrome.⁹ It is reported that about 8% of inflammatory bowel disease patients show this phenomenon.¹⁰

In spite of its high specificity, the skin pathergy response has variable sensitivity and inconstant reproducibility, which limit its use. Regardless of this, it is used in many sets of classification/diagnosis criteria.^{1,9,11,12}

Methods

A case series study was done with twenty-six suspected BD patients (23 women and three men) referred to a private clinic to be tested for pathergy phenomenon.

Out of 26 patients, the pathergy test was performed in 23 patients, as three of them (two men and one woman), which already fulfilled the diagnosis of BD did not agree to be tested. Eight patients did not present disease activity (presence of

symptoms or signs) during the test, contrasting to 15 who did. Five patients were in treatment (taking less than 20 mg of prednisone) when it was performed.

Six needle pricks using 21 gauge disposable needles were done intradermally at the same point on the skin of the forearm, after cleaning the site with 70% ethanol swabs. Results were read by the same observer 48 hours later.

They were considered negative if without papule and only needle mark or erythema and positive if with papule or pustule surrounded by an erythema. Dermatoscopy was done to measure the reaction and observe its clinical aspects.

After the readings a biopsy was performed and stained with hematoxylin and eosin (HE). In some cases more than 1 point of multiple punctures were performed (two prick tests) to choose the biggest papule to biopsy.

Results

Among the patients, 23 were women and 3 men. The age ranged from 11 to 72 years and the average age was 33.11, with standard deviation 14.73.

The oral lesions were present in all patients (100%), followed by genital lesions, observed in 12 (46.1%), ocular lesions in 10 (38.5%), skin lesions in 10 (38.5%), and joint and neurological involvement in 13 (50%) and two (7.7%), respectively (Table 2).

The pathergy test was negative in two BD patients (8.7%) and positive in 21 (91.3%). Among the positive tests, four (17.4%) were less than 1 mm, 16 (69.5%) were between 1-2 mm and one (4.3%) more than 2 mm. These results were not correlated with the disease activity or treatment. Dermatoscopy showed needle marks and mild erythema in the negative tests (2 patients) and erythematous papule/pustule or exulcerocroscous lesions surrounded by erythematous and/or edematous area in the positives (21 patients) (Table 3). It was a good tool especially to measure and examine the inflammatory aspects of the small lesions (less than 2 mm). Figure 1 shows the dermatoscopic aspect of a pathergy test.

The main histopathologic findings of the 23 biopsed patients were: perivascular inflammatory infiltration in 19

Table 1 – Revised international criteria for Behçet's disease.

Criterion	Score (point)
Oral aphtosis	1
Skin manifestations (erythema nodosum-like lesions, papulopustular lesions or pseudofolliculitis, acneiform nodules)	1
Vascular lesions (arterial and venous thrombosis, aneurysm)	1
Pathergy phenomenon (test)	1
Genital aphtosis	2
Ocular lesions	2
Behçet's Disease, three or more points.	

Table 2 – Clinical manifestations in 26 patients with suspected BD.

Clinical manifestation	Absolute number (%)
oral	26 (100)
genital	12 (46)
ocular	10 (38)
cutaneous	12 (46)
arthritis	13 (50)
neurological	2 (7,6)

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