IN TIME VARIATION OF PATHERGY PHENOMENON IN BEHCET'S DISEASE

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Abstract — The pathergy phenomenon (PP) is one of the hallmarks of Behcet's Disease (BD). It has been reported to be positive from 17.5% to 83% of cases in different studies. PP is used as a diagnosis criterion in the Dilsen criteria, the Japan criteria, the International criteria, and the Iran criteria. It is therefore important to know wether PP is a fix manifestation of the disease or a cyclic manifestation like mucocutaneous disorders. To evaluate the charateristic of PP over a period of time, 77 patients were selected randomly (regaardless of their disease activity or their disease manifestations), to have a second pathergy test. The pathergy test was performed by needle puncture on the skin of the forearm. Three methods were used: 1- needle prick with a 21 - gauge needle. 2- needle prick with a 25 - gauge needle. 3- needle prick with a 25-guage needle and the injection of 1 drop of a sterile normal saline solution. The result was evaluated 24 hours later. The shortest interval between two tests was one month and the longest 13 years. The mean interval was 35.4 months. the standard deviation was 36.6 Thirty-nine patients had a negative PP at their first evaluation. At the second evaluation, 27 remained negative while 12 changed to positive. Thirty-eight patients had a positive PP at their first evaluation. At the second evaluation, 19 remained positive while 19 changed to negative. Although there is a difference between the two groups, it is not statistically significant ($x^2=3.813$, $p=0.05$). Our data demonstrates that the pathergy phenomenon is waxing and waning, like the majority of other manifestations of Behcet's Disease. Therefore, it may be repeated for diagnosis purposes.
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Key words: Behcet's disease, pathergy phenomenon, pathergy test, skin hyper - reactivity.

INTRODUCTION

Pathergy phenomenon (PP) is one of the hallmarks of Behcet's Disease (BD). It is a skin hyper-reactivity to a trauma disrupting the skin barrier. It is mostly seen at the site of a venous puncture or skin scratches. The best way to demonstrate it is the pathergy test which consists of a skin puncture by a 21 or 25 gauge needle. The reaction the needle prick appears usually 24 to 48 hours later. The reaction is a round palpable erythema and edema. Rarely it is surmounted by a pustule. The pustular type is characteristic of severely positive reaction. The definition of the pathergy reaction following the pathergy test differs in the literature. For some authors the pustular reaction is needed to classify the test as positive.

The difference in the definition of the pathergy reaction may be one of the causes of the different range of positivity reported in the literature. pathergy test was found positive in 61.5% of patients in the Iran survey (1) and in 44% of the patients in the Japan survey (2). In case studies the frequency was: 62% in China (3), 83% in Russia (4), 18% in Saudi Arabia (5), 71% in Iraq (6), 62% in Egypt (7), 77% in Morocco (8), and 55% in Germany (9).

Pathergy test is not specific for BD. It can be seen in other inflammatory disorders. It can also be seen in the normal population (10). The sensitivity of the pathergy test was 60.3%, the specificity was 86.7%, and the accuracy was 73.5%. Pathergy test is used as a diagnostic criterion in the Dilsen criteria, the Japan criteria, the International criteria, and the Iran criteria. Usually the pathergy test is done once and the result is used as is. If the pathergy phenomenon was a fixed manifestation of BD then it will suffice to test the patient just once for its presence. However, if the pathergy phenomenon was a cyclic manifestation of BD like the other manifestations of the disease, it would be necessary to repeat the test in case of negativity. This will have important implications for the classification of the patient or the diagnosis of the disease. Pathergy phenomenon has been shown to be associated positively with attacks of oral aphthosis and negatively with attacks of anterior uveitis (11). If the pahtergy phenomenon was a cyclic manifestation of BD, it would be of help to test it perriodically as a predictive test.

Pathergy phenomenon is used in (12) Iran Behcet's Disease Dynamic Activity Measurement (IBDDAM) as one of the parameters of the disease activity index. If the pathergy phenomenon changed periodically, it is important to test it at each visit for the calculation of IBDDAM.

To find out the characteristic of the pathergy phenomenon over time and test the hypothesis of its cyclic evolution a longitudinal study was undertaken in 77 patients.

MATERIALS AND METHODS

Patients

Seventy-seven patients with BD who had a pathergy test at the beginning of their evaluation in the Behcet's Disease Unit were selected to have a second pathergy test. The selection was done randomly regardless of the disease activity or the disease manifestations.

Pathergy Test

Test was performed by a needle puncture on the skin of the forearm. Three methods were used: 1- Needle prick with a 21-gauge needle. 2- Needle prick with a 25-gauge needle. 3- Needle prick with a 25-gauge needle and the injection of 1 drop of a sterile normal saline solution. The needle was inserted diagonally, intradermally, under the skin of the forearm. The result was evaluated 24 hours later.

Statistical Analysis

Results were compared by the Chi square test (x^2).

RESULTS

The shortest interval between two test was one month and the longest interval was 13 years. The mean interval was 35.4 months. The standard deviation was 36.6. Patients were divided into two groups according to the result of their first pathergy test: Positive and negative groups. Thirty-nine patients were classified in the Negative group (50.6%). They had a negative reaction at their first evaluation. At the second evaluation, 27 patients had a negative pathergy test again, while 12 patients changed their reaction and showed a apositive pathergy test. Thirty-eight patients were classified in the positive group (49.4%). They had a positive reaction at their first evaluation. At the second evaluation, 19 patients showed the same positive reaction as their first reaction, while 19 patients changed their reaction to negative. As an overall result, the comparison between the first pathergy test (38 positive, 39 negative) and the second pathergy test (31 positive, 46 negative) did not show any statistically significant difference (x^2=1.287, P=0.26). The comparison of patients who changed their pathergy reaction from positive to negative (19 of 38), with those who changed their reaction from negative to positive (12 of 39), also did not show any statistically significant difference (x^2=3.813,p=0.05).

DISCUSSION

Our data demonstrates that the pathergy phenomenon in Behcet's Disease has the same probability to become positive when it was negative as to become negative when it was positive. This favors the theory that the pathergy phenomenon, like other manifestations of Behcet's Disease, is waxing and waning. This has major implications as pointed out in the introduction part of this report. The positivity of the pathergy test increased from 49.4% (at the first test) to 65% when the number of newly positive patients were added to those classified positive at the first test. In conclusion we suggest that the pathergy test be repeated for diagnosis purposes, as for the calculation of IBDDAM in drug trials or for disease monitoring.

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