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Acute Febrile Polyarthritis Revealing very Late-Onset Behçet's Disease

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Abstract

***Introduction:** Late-onset forms of Behçet's disease (BD), by definition after the age of 40, remain rare and highly controversial. The revelation of these forms by articular manifestations is not uncommon and poses a real diagnostic challenge; particularly in elderly. We report an original observation of acute febrile polyarthritis revealing a very late-onset BD.*

***Case Report:** A 65-year-old patient with simple and well-balanced primary hypertension under combination therapy with a beta blocker and an angiotensin-converting enzyme inhibitor, was hospitalized for etiological assessment of acute and febrile polyarthritis affecting hands, wrists, knees and ankles, evolving for two weeks with fever at 38°C.*

The examination noted multiple oral aphtosis, genital ulcers and scares, and pseudo-folliculitis at the inner sides of the thighs and back.

The infectious, radiological and immunological assessments were negative, eliminating particularly infections, endocarditis, lupus, ANCA-associatedangiitis, spondylarthropathies, and rheumatoid arthritis. The Pathergy-test and HLA B51 typing were positive.

The diagnosis of BD was retained in accordance with the new international criteria for BD and the patient was placed on Colchicine®, Aspégic® and low dose oral corticosteroids with a favorable evolution.

***Conclusion:** The maximum incidence of BD is observed between the second and fourth decade of life. Late-onset forms of BD are rare and very late-onset forms are exceptional and uncommon. As rare as it is, BD deserves to be mentioned in front of any arthritis that is not proven, occurring in a geriatric environment.*

Keywords: *Acute polyarthritis, late-onset, Behçet's disease, arthritis, vasculitis, elderly.*

Introduction

Behçet's disease (BD) is a chronic inflammatory disease of unknown etiology. It appears to be multifactorial involving infectious, toxic, immunological, and predisposing genetic factors. It was individualized in 1937 by Hulusi Behçet [1], and was recognized to be a systemic vasculitis since the 2012 revision of the Chapel Hill consensus for the classification and nomenclature of systemic angiitis[2]. It is a cosmopolitan affection with a very variable prevalence depending on the country, ranging from 0.30/100,000 in Western countries to 420/100,000 in Central Asia and Middle East (80/100,000 in Iran and 420/100,000 in Turkey) [3,4]. It is particularly common in countries located on the former "Silk Road" [4].

The maximum incidence of this disease is observed between the second and fourth decade of life [4-6]. Late-onset forms of BD, defined by the onset of symptoms after the age of 40, are rare [7,8], representing less than 3% of cases [9], and are very little known and studied [5]. The occurrence of the disease after the age of 60 remains exceptional and unusual [5,6].

We report an original observation of BD with very late-onset, and revealed by acute polyarthritis.

Case Report

A 65-year-old Tunisian patient with simple and well-balanced primary hypertension under combination therapy with a beta blocker and an angiotensin-converting enzyme inhibitor, was hospitalized for etiological assessment of acute and febrile polyarthritis affecting hands, wrists, knees and ankles, evolving for two weeks.

The examination noteda febrile patient at 38° C,peripheral joints (wrists, elbows, knees, ankles, proximal and distal metacarpophalangeals, and metacarpo-carpals) swollen, painful, and with significant synovitis, multiple oral aphtosis(inner side of the lower lip (Fig. 1), tongue tip (Fig. 2), and inner faces of the cheeks), as well as multiple genital ulcerations and hypochromic scars (Fig. 3), and multiple pseudo-folliculitis at the inner sides of the thighs and back.The joint involvement was bilateral and symmetrical.

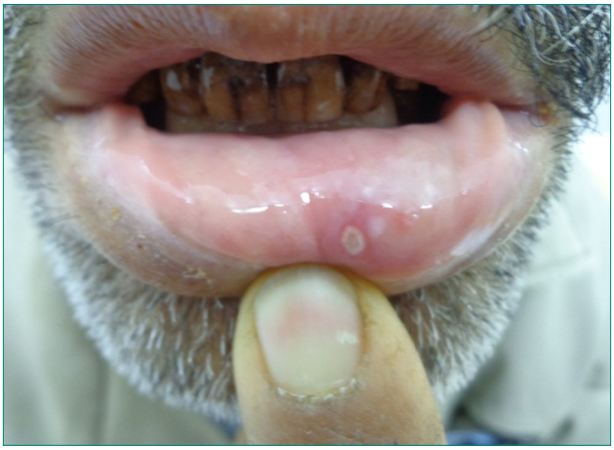

Fig 1. *Aphtosis of the inner side of the lower lip.*

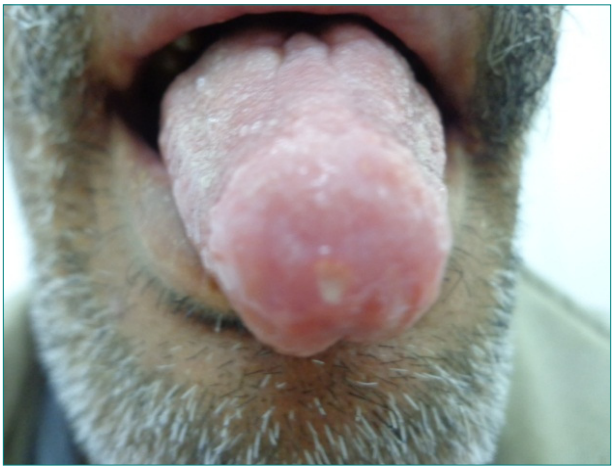

Fig 2. *Aphtosis of the tip of the tongue.*

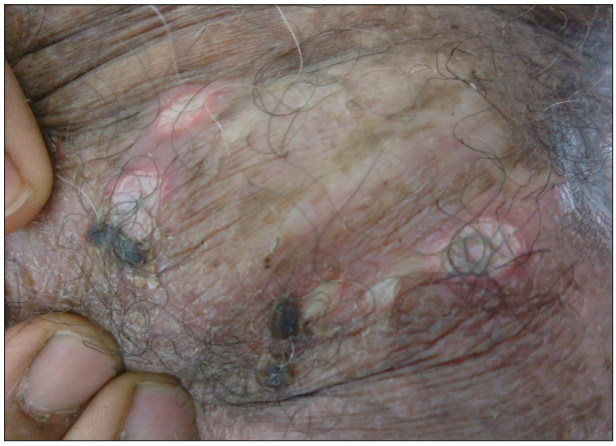

Fig 3. *Multiple hypochromic scars and multiple active scrotal ulcerations.*

The cardiopulmonary, neurological, and abdominal examination was without abnormalities. The biology showed a marked inflammatory syndrome with an erythrocyte sedimentation rate at 98mmH1 and a C reactive protein at 36mg/l, and standard radiographs of affected joints were without abnormalities.

The infectious, radiological and immunological assessments were negative, eliminating particularly infections, endocarditis, lupus, ANCA-associatedangiitis, spondylarthopathies, and rheumatoid arthritis: leucocytes, platelet, hemoglobin, creatinine, uric acid, ionogram, glycemic parameters, ASAT, ALAT, muscle enzymes, electrophoresis of serum proteins, urine analysis, bacteriological, viral and tuberculous tests, rheumatoid factor, ANA, anti-DNA, p-ANCA, c-ANCA, anti-CCPantibodies, chest X-ray, thoraco-abdominopelvic CT, abdominal ultrasound, transthoracic echocardiography, blood and urine cultures, X-ray and CT scan of sacroiliac joints, and HLA B27typing.

The Pathergy-test was positive. The specialized ophthalmological examination revealed no abnormalities, ant HLA B51 typing was positive

The diagnosis of BD was retained in accordance with the new international criteria for Behçet's disease (score at 6 points: oral aphtosis, genital ulcerations, pseudofolliculitis, and positive Pathergy test). Systemic visceral impairment was negative. The patient was placed on Colchicine®, Aspégic®, and oral corticosteroid therapy at a dose of 30mg/d for four weeks followed by gradual tapering. The evolution was rapidly favorablewith apyrexia on the third day,

gradual disappearance of joint complaints, and rapid cicatrizations of oral and genital ulcers (Fig.4).

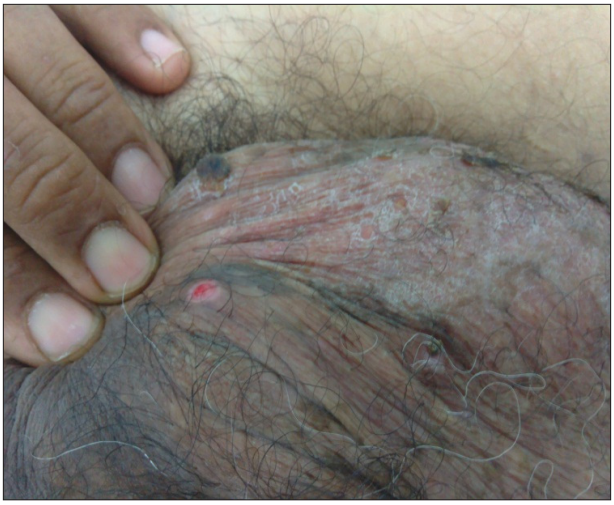

Fig 4. *Progressive cicatrization of the scrotal ulcerations.*

Discussion

BD is classically a disease of the young adult, and it is estimated that in 80% of cases the first symptom of the disease occurs between 16 and 35 years, and that in 97% of cases the disease begins before the age of 40 [9]. The disease can, however, begin at any age, from the first months of life (neonatal BD) [10], childhood (Pediatric and juvenile Bd) [11], adult (adult BD) [12], and until old age (even after 70 years) [6,7,13].

The onset of the disease after the age of 50 is very rare [4, 14]: only nine out of 439 patients with BD in the Saricaoglu H et al series, started their illness after the age of 50 (1.56%) [14]. The very late-onset forms of this disease, beginning after the age of 60, remain exceptional and unusual with only a few sporadic cases reported in the world literature [5,6]. These presentations represent a real diagnostic challenge for clinicians [5, 6], especially with other chronic inflammatory rheumatisms particularly common in the elderly such as rheumatoid arthritis, psoriatic arthritis, RS3PE, giant cell arteritis, and polymyalgia rheumatica [15].

Joint manifestations are common during BD; they can be seen in more than half of cases [16], may be the first sign of the disease [17], and may have several clinical presentations: inflammatory arthralgia, peripheral arthritis, inflammatory lower back pain, pseudo-gout, spondyloarthropathies, sacroiliitis, and fibromyalgia [16-19]. Arthritis is considered a minor clinical manifestation of the disease [4], and can be in the form of mono-, oligo-, or polyarthritis affecting all joints (small and large) [16-19].

Among all rheumatologic manifestations of BD, polyarthritis remains classically rare; in fact, its frequency was only 5% in the series of Ait Badi MA et al of 176 patients with BD of whom 79 had osteoarticular manifestations [17]. This frequency of 5% was also confirmed by the large prospective study of Fatemi A et al of 2321 patients with BD and followed over a period of 5 years with a total of 289 cases of recorded arthritis [18]. Monoarthritis is the most common and knees are the most affected joints [18].

This polyarthritis can be acute, recurrent, or chronic, and is typically non-erosive, non-destructive, and non-deforming, with a good clinical response to colchicine [20,21].

Sometimes misleading forms of pseudo-gout, pseudo-rheumatoid arthritis with erosion and destruction [17,20,22], as well as rapidly progressive, refractory and pseudo-septicemic presentations can be seen [21], presenting a real diagnostic challenge.

These particular forms of late-onset and very late-onset BD deserve to be well known by health care workers because they do not seem to be as benign as we thought [4,5]. In fact, serious systemic damage and severe ocular complications are often associated with it, threatening the prognosis [5,14].

Conclusion

Late-onset BD is rare and often unknown and overlooked in current medical practice. Very late-onset forms (after the age of 60) remain exceptional, unusual, and represent a real diagnostic challenge for clinicians, especially with the frequency of rheumatological diseases at this age.

The BD deserves to be mentioned in front of any arthritis that is not proven, occurring in a geriatric environment.

Early diagnosis and adequate management are the only guarantors of a better prognosis, both articular and systemic, of this disease.

Abreviations

ASAT: Aspartate aminotransferase, ALAT: Alanine aminotransferase, ANA: antinuclear antibodies, p-ANCA: perinuclear antineutrophil cytoplasmic antibodies, c-ANCA: cytoplasmic antineutrophil cytoplasmic antibodies, Anti-CCP: anticy cliccitrullinated peptide antibodies, anti-DNA: anti-double stranded deoxyribonucleic acid antibodies, RS3PE: remitting seronegative symmetrical synovitis with pitting edema.

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