Behcet disease: a case study

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INTRODUCTION: Behçet disease (BD) is more common in Mediterranean countries and the Middle East, and in Japan the prevalence is 10 per 100,000 of the population (Chamberlaine, 1977, 1978). The etiology of the disease is unknown, but an immunological basis seems possible. The histology is of vasculitis, particularly involving venules and capillaries, with mononuclear cell infiltration. Circulating autoantibodies to oral mucosa were first described in 1963 (Oshima et al., 1963). The main clinical features of Behget's disease are relapsing iridocyclitis with recurrent oral and genital ulceration.

METHODS: We report a cross-sectional and multicentric study (in 3 centers in Algeria) in a series of 70 cases followed in rheumatology since 1986 to date. The diagnosis of BD was made on international criteria (ICBD).

Table 1: Clinical data collected in the Behcets’s cohort.

RESULTS:
70 cases reported: 51 male for 19 female, index ratio = 3, the average age of diagnosis is 37 years (18-72), the juvenile onset seen in 5 cases. Mucocutaneous disorders are reported as buccal aphtosis in 38% (bipolar aphtosis 62%), pseudofolliculitis in 55% , erythema nodosum in 16%, vasculitis in 9%, positif pathergy test in 10%, joint disorders reported in 95%, thrombophlebitis in 11% and ocular involvement in 39%, neurologic damage in 18% (Table 1) , inflammatory syndrome seen in 49%, HLA B5 in 23%.

Gastrointestinal involvement was more frequent than in patients without vascular involvement (p<0.001); ocular involvement was less frequent (p<0.05). Only 3 patients (12%) required surgery. Patients received prednisone and immunosuppressants, including etanercept, for vascular and/or concurrent gastrointestinal involvement. Nine patients received warfarin, without bleeding complications.Two patient died during the observation period.

Feature	Number (%)
Buccal aphtosis	27 (37.8)
Bipolar aphtosis	6 (9.1)
Pseudofolliculitis	38 (54.8)
Erythema nodosum	11(15.7)
Joint disorders	66 (94.7)
Neurologic damage	12 (18)
Ocular involvement	27 (39)
Vasculitis	6 (8.7)

International comparison between the Erasmus MC and four other cohorts for diagnostic criteria for Behçet’s disease					
	The Netherlands	Germany	UK	Turkey	Morocco
N	110	590	419	2147	1034
♂:♀ ratio	1	1.4	0.5	1.03	2
Oral ulcers	100%	98%	100%	100%	100%
Genital ulcers	79.1%	64%	89%	88%	86%
Erythema* nodosum	31.8%	42%	.	.	16%
Pustules*	80.9%	62%	.	.	64%
Uveitis	61.8%	53%	68%	29%	44%
Positive pathergy test	57.1%**	34%	32%	56%	53%

CONCLUSION Behcet disease needs attention for it’s multi-organ involvement. The takeover depends in severity of disease, often needing to have recourse to use: corticoids, immunosuppressive drugs, less than interferon alpha, thalidomide or anti TNF alpha.