Clinical and Prognostic Profile of Behçet's Disease (A Series of 111 Cases)

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Introduction

Behçet's disease (BD) is a complex systemic vasculitis of unknown etiology. It is characterized by various clinical manifestations, including cutaneous-mucosal, ocular, articular, vascular, and neurological manifestations. This study describes the epidemiological, clinical, and prognostic characteristics of a series of 111 patients with Behçet's disease.

Patients and Methods

This was a retrospective study involving 111 cases of Behçet's disease meeting the revised 2014 International Criteria for Behçet's Disease. Patients were recruited from the internal medicine department of Oued Eddahab Military Hospital (Agadir) and Hassan II Regional Hospital (Agadir).

Results

A total of 111 cases were included in this study. These patients consisted of 67 men and 44 women, with a male-to-female ratio of 1.5. The mean age at the time of Behçet's disease diagnosis was 35.2 years (range: 14–73 years), and the mean time to diagnosis of BD was 5 years. Clinically, cutaneous-mucosal manifestations were found in 100% of patients. The distribution of other manifestations was as follows:

- Vascular involvement, affecting 33 patients (29.7%):
- Deep vein thrombosis of the lower limbs in 18 patients;
- Venous thrombosis in unusual locations in 10 cases;
- Arterial involvement in 5 patients.
- Ocular involvement in 31 patients (27.9%), including nearly bilateral blindness in 8 patients and monophthalmia in one patient;
- Articular involvement in 29 patients (26.1%);
- Neuro-Behçet in 6 patients (5.4%) and 28 cases of neuropsychiatric manifestations observed mainly in male subjects;
- Four cases of bilateral optic neuropathy (3.6%);
- Digestive involvement in one patient (0.9%).

All patients were on colchicine (100%). 69 patients (62.1%) received systemic corticosteroid therapy. Azathioprine was prescribed in 18 cases (16.2%), cyclophosphamide in 16 cases (14.4%), anti-TNF alpha agents for 8 patients (7.2%), and anticoagulants for 19 patients (17.1%).

In our series, there were 2 cases of relapse, both of which involved vascular manifestations.

Only one death was recorded, caused by a Budd Chiari.

Discussion

The distribution of clinical manifestations varies across studies, which may be attributed to population heterogeneity and differing clinical criteria for each manifestation. In our study, buccal aphtosis was observed in all patients, aligning with the findings reported in the literature [1].

Angio-BD showed a negative correlation with ocular involvement, consistent with literature observations [2].

Our study, along with others, noted a high incidence of venous thrombosis and a scarcity of arterial manifestations during BD progression [3].

Additionally, our study confirms a predilection for thrombosis in the lower limbs [3].

Conclusion

Our study confirms most of the epidemiological observations made in other countries of the Mediterranean region regarding Behçet's disease. The results confirm the male predominance of the disease, which are known to be more severe and to have a very poor prognosis. We also observe a high prevalence of cutaneous-mucosal lesions, and that half of the patients presents with severe manifestations, especially ocular and vascular. However, parenchymal neurological and digestive involvement were less frequent in our series [4].

Références

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