Peculiarities of the juvenile form of Behçet's disease

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Introduction :

Behçet's disease is a chronic vasculitis that affects vessels of all calibers, involving both veins and arteries. It primarily affects young adults, with a peak frequency between the ages of 20 and 30. Its pathophysiological mechanism is poorly understood and highly heterogeneous, involving both autoinflammatory and autoimmune components. The disease is accompanied by cutaneous, mucosal, articular, digestive, neurological, and ocular manifestations. A distinction must be made between Juvenile Behçet, which involves a diagnosis based on the presence of at least two criteria of the disease in a patient under 15 years of age, and Behçet with juvenile onset, where the first symptom appears before the age of 15 but the diagnosis is only confirmed later. Our study involves 90 patients with Juvenile Behçet out of a total of 1,571 patients.

Objectives:

To assess the different clinical manifestations of juvenile Behçet's disease and compare them with the results collected from adult patients in our series.

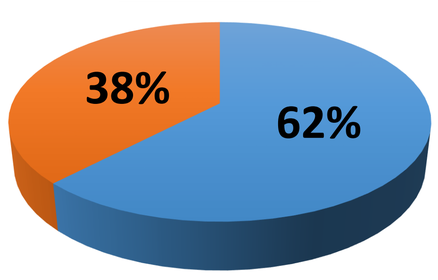
Patients and Methods:

Our work is based on a retrospective, single-center study conducted within the Internal Medicine Department and Geriatrics Unit of Ibn Rochd University Hospital in Casablanca over a period of 42 years, from January 1981 to December 2023. This study allowed us to collect data on 90 patients diagnosed with Juvenile Behçet's Disease, according to international criteria, before the age of 15, out of a total of 1,571 patients treated for Behçet's disease during the same period.

Results:

Our study identified 90 patients meeting the diagnostic criteria for juvenile Behçet's disease, out of a total of 1,571 patients, representing 5.73%, of which 56 were male and 34 were female, giving a sex ratio of 1.6

Figure 1: Distribution of patients by gender



A family history of Behçet's disease was found in 12.5%.

Figure 2: Family History of Behçet Desease:

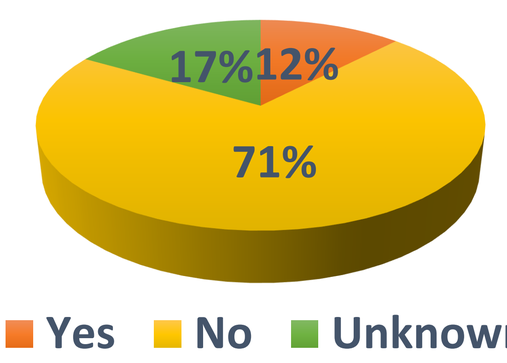
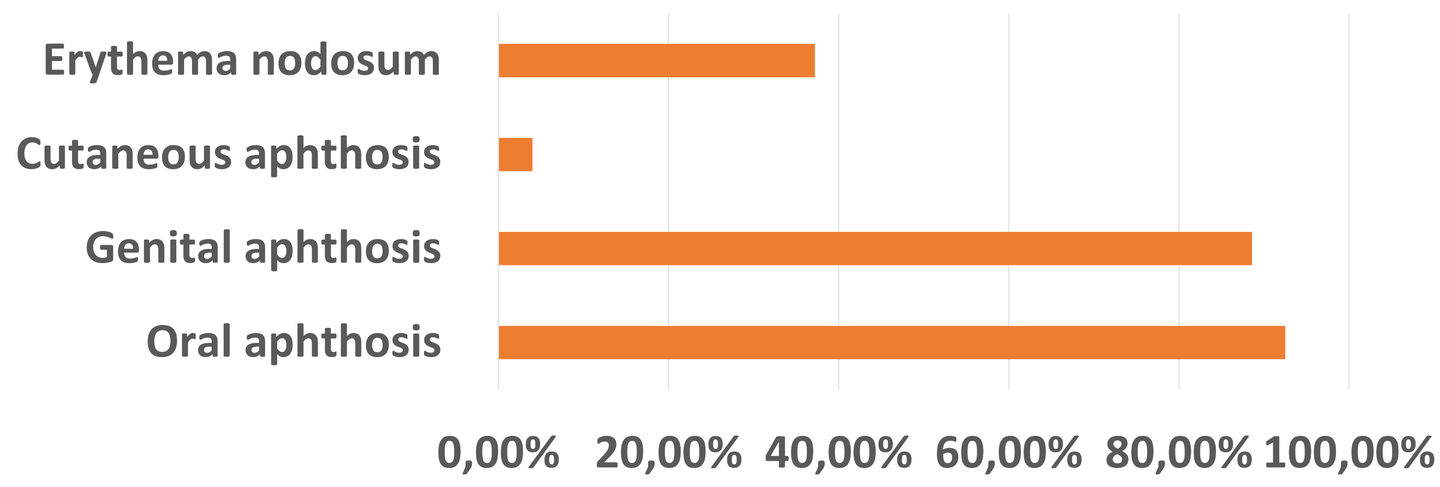


Figure 3 Cutaneous manifestations:



Ocular involvement was observed in 65.7% of the juvenile patients

Ocular Manifestations

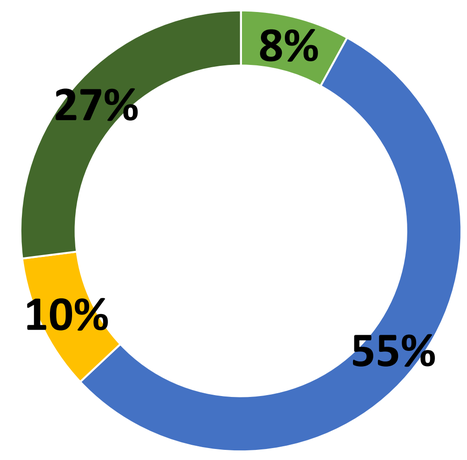
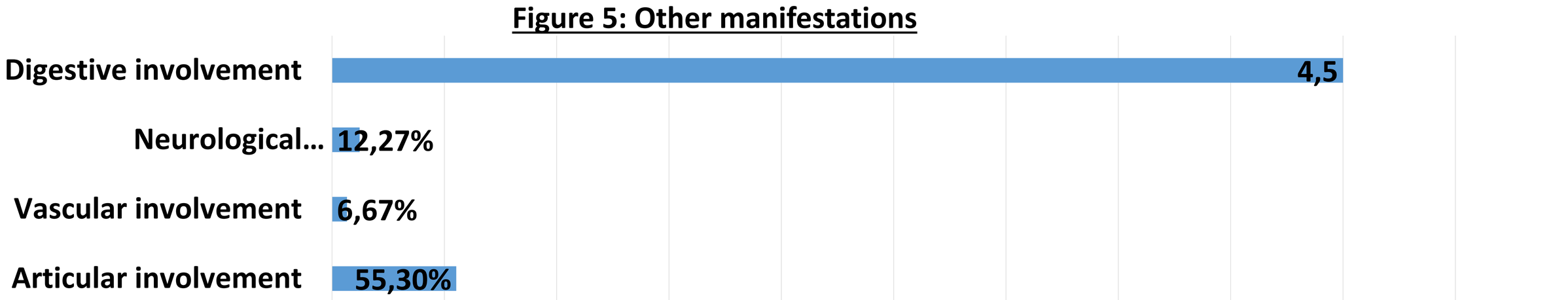


Figure 5: Other manifestations



Discussion:

The average age of diagnosis in the juvenile group was 7 years, while in the adult group it was 12 years. Family history of Behçet's disease was found in 12.5% of juvenileserie compared to only 5.1% in the adult serie. Family history of aphthosis was found in 50% of the juvenile serie compared to 13.3% in the adult serie. Skin involvement manifested as oral aphthosis in 92.5% of juvenile patients compared to 99.6% in adult patients, genital aphthosis was found in 88.6% of the juvenile group versus 87.9% in the adult group, dominated by scrotal localization in both series. Cutaneous aphthosis was seen in 4% of the juvenile serie compared to 7.8% of the adults and erythema nodosum was present in 37.2% of juveniles versus 15.43% in the adults. Ocular involvement was observed in 65.7% of the juveniles, with 55% presenting with posterior uveitis, 8% with anterior uveitis, 10% with panuveitis, and 27% being at the stage of blindness. In the adult serie, ocular involvement affected 60.8% of the patients. Joint involvement was found in 55.3% of juveniles, and vascular involvement was present in 6.67%, with 33.33% having superior vein thrombosis, 33.33% deep vein thrombosis of the limbs, 16.66% internal jugular vein thrombosis, and 16.66% aortic aneurysm. In the adult serie, vascular involvement was 23.85%. Neurological involvement was observed in 12.22% of the juveniles, with 63.6% presenting parenchymal involvement, 27.2% cranial nerve involvement, and 9.2% non-parenchymal involvement, compared to 18.36% neurological involvement in the adult serie. Digestive involvement was seen in 7.8% of juvenile patients compared to 9.18% in the adult group.

Conclusion:

Behçet's disease is a vasculitis that affects vessels of various calibers and presents with cutaneous, mucosal, neurological, digestive, articular, and ocular manifestations. Its pathophysiology is poorly understood. Although the disease classically affects young adults, with a peak incidence between the ages of 20 and 30, it can occur at extreme ages. Juvenile Behçet has some specific characteristics compared to adult Behçet, as demonstrated by our study, including a stronger familial predisposition for the disease or aphthosis, earlier and more severe ocular involvement, but paradoxically with better treatment response and fewer relapses in juvenile Behçet compared to adult Behçet. Articular involvement is equally frequent in both juvenile and adult cases, while vascular, neurological, and digestive involvement are more common and more severe in adults, which is consistent with the literature.