Clinical course of Pediatric Onset Behçet's Disease in young adulthood

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Introduction

- Behçet's disease (BD) is a chronic inflammatory disease affecting the eye, central nervous system (CNS), vascular and gastrointestinal systems (GIS), as well as skin, mucosa and joint involvement.
- Although BD onset is commonly in the second or third decades, the initial symptoms occur under the age of 16 years in 4%–26% of the patients [1].

Aim

- To assess the clinical course of pediatric onset BD in adulthood period.

Methods

- The files of 1114 BD patients were reviewed retrospectively.
- 51 (4.6%) (F/M:21/30) pediatric-onset BD patients were included in the analysis.
- Demographic and clinical characteristics, follow-up and treatment data of the patients were recorded from files.
- Categorical variables are presented as frequency (n) and percentage (%). According to data distribution, numerical data were displayed as mean and standard (SD) deviation or median and interquartile range (Q1-Q3).

Results

- The median age at diagnosis was 16 (14-17) years.
- The median follow-up duration was 51 (26-96) months.
- Erythema nodosum was more common in females (p=0.008) and vascular involvement was more common in males (p=0.025). The distribution of disease findings was shown in Table 1.
- At the end of follow-up, 32 (62.8%) patients had major organ involvement,
  - Sixteen (50%) patients had a major organ involvement at the time of diagnosis.
  - Sixteen (50%) patients developed a major organ involvement a median of 54 (12-105) months after diagnosis.
- Male patients developed major organ involvement earlier than female patients (p=0.042).
  - A median of 12 (6-84) months for males vs. a median of 84 (60-108) months for females.
- Of all patients, 33.3% had ocular involvement, 27.5% had vascular involvement, 17.6% had CNS involvement, and 3.9% had GIS involvement (Table 1).

Results

Table 1: Distribution of disease characteristics of pediatric onset Behçet's Disease patients.

Clinical findings	Male (n=31)	Female (n=20)	p	All (n=51)
Oral aphthae	30 (96.8%)	20 (100%)	1	50 (98%)
Genital ulcers	26 (83.9%)	17 (89.5%)	0.695	43 (84.3%)
Pathergy test positivity	21 (80.8%)	10 (66.7%)	0.453	31 (75.6%)
Papulopustular lesions	21 (67.7%)	10 (50%)	0.205	31 (60.8%)
Erythema nodosum	15 (48.4%)	17 (85%)	*0.008*	32 (62.7%)
Family history	4 (13.3%)	6 (30%)	0.171	10 (20%)
Arthritis/ arthralgia	-	2	0.149	2 (3.9%)
Major organ involvement	22 (71%)	10 (50%)	0.131	32 (62.7%)
Vascular	12 (38.7%)	2 (10%)	*0.025*	14 (27.5%)
Ocular	11 (35.5%)	6 (35.3%)	0.685	17 (33.3%)
Neurologic	6 (19.4%)	3 (15%)	1	9 (17.6%)
Gastrointestinal	1 (50%)	1 (50%)	1	2 (3.9%)

- 47 (M:27/F:20) patients had a follow-up with median of 50 (20-82) months in adulthood period.
  - Thirty-one (65.6%) patients had major organ involvement.
    - 20 (64.5%) patients had major organ involvement in the pediatric period,
    - 11 (35.5%) patients developed major organ involvement in adulthood.
- Overall, 19 (40.4%) patients had active disease manifestations (relapse and/or new major organ involvement) in adulthood follow-up.
  - 11 (57.9%) had new major organ involvement,
  - 7 (36.8%) had a relapse of the same organ,
  - one (5.3%) had both new major organ involvement and a relapse.
- The disease course of patients are seen in Table 2.

Conclusion

- Our results show that, about half of the pediatric-onset BD patients has still active disease manifestations (mainly new major organ involvement) in adulthood period.

References: 1. Yıldız, M., et al., Pediatric Behçet's disease-clinical aspects and current concepts. European journal of rheumatology, 2020. ***7****(Suppl 1): p. S38.*

Table 2: Disease course of patients followed in adulthood period.

Patients followed in adulthood	n=47 (%)
Major organ involvement in childhood	20 (42.6%)
Relapse or/and new major organ involvement	8 (17%)
No relapse or major organ involvement	12 (25.5%)
Mucocutaneous disease in childhood	27 (57.4%)
New major organ involvement	11 (23.4%)
No major organ involvement	16 (34%)