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PReS-FINAL-2363: Behçet's disease in children: the Great Ormond Street Hospital experience

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Introduction

Behçet's disease (BD) is rare in childhood and remains challenging in diagnosis and lack of evidence-based data for its treatment. Hence there is an urgent need to understand the scope of the disease in children.

Objectives

The aim of this study is to describe the clinical spectrum and the therapies used to treat children with Behçet's disease (BD) in children.

Methods

46 patients (22 male) were identified with a positive family history of BD in 6 cases. Age of onset was 4.87 (0.04-15.71) years with a time to diagnosis of 3.74 (0.25-13.48) years. The main clinical features at presentation were: recurrent oral ulceration (87%), genital ulceration (20%), cutaneous symptoms (11%), fever (30%), gastrointestinal symptoms (26%), musculoskeletal (22%). uveitis (2%). Recurrent genital ulceration was significantly more common in female patients (P = 0.044). The majority of children were treated with colchicine (74%) and corticosteroid (41%). Anti TNF-a treatment was reserved for severe and/or refractory cases (15%). There was a median of 2 (range 0-12) episodes of oral ulceration per year after the treatment. Interestingly only 10 patients fulfilled The International Study Group (ISG) BD diagnostic criteria.

Results

46 patients (22 male) were identified with a positive family history of BD in 6 cases. Age of onset was 4.87 (0.04-15.71) years with a time to diagnosis of 3.74 (0.25-13.48) years. The main clinical features at presentation were: recurrent oral ulceration (87%), genital ulceration (20%), cutaneous symptoms (11%), fever (30%), gastrointestinal symptoms (26%), musculoskeletal (22%). uveitis (2%). Recurrent genital ulceration was significantly more common in female patients (P = 0.044). The majority of children were treated with colchicine (74%) and corticosteroid (41%). Anti TNF-a treatment was reserved for severe and/or refractory cases (15%). There was a median of 2 (range 0-12) episodes of oral ulceration per year after the treatment. Interestingly only 10 patients fulfilled The International Study Group (ISG) BD diagnostic criteria.

Conclusion

Although most cases were diagnosed in late childhood the first presentation was as early as 1 month old. Delay in diagnosis due to incomplete presentation in certain cases. Oral ulceration was the most common presenting symptom. Uveitis was less frequent than previous series. A range of drugs was used including biologic therapy for severe cases.

Disclosure of interest

None declared.

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