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.

Methods.– Single centre (Great Ormond Street Hospital, London) retrospective case note review of children with BD seen between 1987–2012. Demographics, clinical features, treatment, relapses were recorded. Continuous variables were summarised as median and range. Categorical variables were presented as percentages. To explore gender differences in presenting symptoms a Fisher’s exact test was applied.

Results.– Forty-six 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 [1].

Discussion.– 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 cases. A range of drugs was used including biologic therapy for severe cases.

Conclusion.– Large multicentre prospective studies of paediatric BD are needed to facilitate development of diagnostic criteria, standardization of treatment protocols and inform therapeutic clinical trials.

Reference

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Refractory cutaneous vasculitis associated with MAGIC syndrome: Case report

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Introduction.– An overlap syndrome of relapsing polychondritis (RP) and Behçet’s disease (BD) has been described in which patients present with features resembling both conditions. This syndrome is termed the “MAGIC” syndrome, an acronym for mouth and genital ulcers with inflamed cartilage. A variety of skin findings can occur in MAGIC syndrome. Herein, we report a case of refractory cutaneous vasculitis associated with MAGIC syndrome.

Methods.– We have retrospectively reviewed the clinical, laboratory, and histological data of this case.

Results.– A 17-year-old woman presented with a 5-year history of recurrent oral aphthous ulcers, intermittent fever, malaise, sore throat, arthralgia, and a relapsing nonpruritic rash. Four years after the first symptoms she presented with painful inflammation of both ears and nose, reddened left eye, and recurrent erythematous ulcerative lesions in the skin of the scalp, face, trunk and limbs. Laboratory studies revealed no abnormalities. Pathergy test was positive. Her HLA-typing revealed B27 positive. Chest and abdominal computed tomography were normal. An autoimmune, neoplastic and infectious screening showed no abnormalities. Histologic examination of skin lesion specimens showed lymphocytic vasculitis. A presumed diagnosis of MAGIC syndrome was made. After initial treatment with corticosteroid, colchicine, and intravenous immunoglobulin, she received several immunosuppressive drugs with only partial control of the aphthous ulcers and the cutaneous lesions.

Discussion.– This patient met the classification criteria for relapsing polychondritis and Behçet disease, therefore we suggest she had a MAGIC syndrome. This interesting presentation of two rare diseases occurring together suggests a shared pathogenic mechanism. However, whether this is indeed a bona fide syndrome, or simply BD complicated by RP manifestations, has been debated in the literature.

Conclusion.– Patients with MAGIC syndrome may present with a refractory cutaneous vasculitis.

Further reading

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Behçet’s disease and pregnancy

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Introduction.– Behçet’s disease (BD) can affect young women during their procreation period, and the outcome of BD during pregnancy is a concern for rheumatologists in daily practice. Some studies have reported flares of BD during pregnancy. A higher rate of miscarriage has been reported once, thus raising the question whether the outcome of the pregnancy and of the newborn can be affected by BD. The question of treatment of BD during pregnancy is an important issue because of adverse effects that may affect the course of pregnancy. We aimed to describe the interplay between BD and pregnancy in a large cohort.

Methods.– A retrospective study including 76 pregnancies in 46 patients [median age (IQR): 28.4 (22.8–30.9)] fulfilling the international criteria for BD. Factor associated with the occurrence of complications during pregnancy were assessed.

Results.– Twenty-seven out of 76 pregnancies (35.5%) pregnancies were complicated by worsening of BD symptoms, of which oral and genital ulcerations (78.4 and 67.6%), as well as ocular complications (32.4%) were the most frequent. The annual rates of BD flares were

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