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AUTO-IMMUNE AND INFLAMMATORY DISEASES IN CHILDREN WITH SICKLE CELL DISEASE: DIAGNOSTIC AND THERAPEUTIC ISSUES

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Background: Coexistent auto-immune and inflammatory diseases (AIID) and sickle cell disease (SCD) have been recently described in adults and children, however its frequency and physiopathology remain unclear (1–6).

Objectives: The aim of this study is the analysis of clinical and biological characteristics at AIID diagnosis and the evolution under treatment in children with SCD

Methods: Between May 1991 and March 2018, 35 of 3,800 SCD children diagnosed with AID in seven hospitals in Paris and suburb were analyzed in a retrospective survey.

Results: Thirty-five patients reported 44 AIID: auto-immune liver disease (AILD, n=13), inflammatory bowel disease (IBD, n=7), juvenile idiopathic arthritis (JIA, n=6), systemic lupus erythematosus (n=5), autoimmune hemolytic anemia (n=3), Sjögren's syndrome (n=2), histiocytic necrotizing lymphadenitis (n=2), vasculitis (n=2), myasthenia gravis (n=2), sarcoidosis (n=2), inflammatory uveitis (n=1), scleroderma/juvenile dermatomyositis (n=1). Median age at diagnosis was 10 [2 – 18] years. The mean delay between first symptom and diagnosis was 15.5 (± 29) months. The time of diagnostic was significantly longer for patients with JIA compared to other AID (63 versus 10 days, p=0.004). Sixteen patients (45.7%) had hypergammaglobulinemia > 20 g/L at diagnosis. AILD had a hypergammaglobulinemia at the time of diagnosis (30.0g/L), with a statically significant decrease at the end of follow-up (18.2g/L, p=0.0048). Among 21 patients (60%) treated with systemic steroids, it triggered vaso-occlusive crisis in 14 (66.7%), one acute chest syndrome, one transient ischemic attack. Thirteen of 35 patients (37.1%) were managed with biotherapy for AIID, well tolerated. Three patients (8.6%) underwent stem cell transplantation, one died of a cortico-resistant and multipolar graft versus host reaction, two were cured of both AIID and SCD. Nine severe infections were reported, four under steroids, five under biotherapy.

Conclusion: Diagnosis and therapeutic care of coexistent auto-immune and inflammatory diseases are difficult and challenging in children with SCD. Annual monitoring of inflammatory markers could be recommended to detect AIID earlier and prevent diagnostic delay in case of high ascension in SCD patients.

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UVEITIS IN PEDIATRIC RHEUMATOLOGY: TERTIARY CENTER EXPERIENCE IN TURKEY

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Background: Since pediatric uveitis is generally asymptomatic, the diagnosis and treatment may be mostly delayed. Severe complications and visual loss may be observed even at the initial visit. Pediatric uveitis is tend to be chronic, persistent, recurrent, and the management may be complex (1).

Objectives: The aim of this study is to report epidemiology, etiology, clinical features, management and the outcomes of non infectious pediatric uveitis at a tertiary pediatric rheumatology center in Turkey.

Methods: The clinical records of the patients with non infectious uveitis who were followed up by department of pediatric rheumatology and ophthalmology were reviewed, from January 2013 to June 2018, retrospectively. The inclusion criteria were as follows being age ≤ 16 years, following up at least 6 months in both the ophthalmology and pediatric rheumatology clinics. Uveitis was categorized anatomically according to the Standardization of Uveitis Nomenclature criteria (2).

Results: Of 37 patients (67 eyes), 45.9% were female. Mean age of onset was 8, 5 ± 4, 4 years (1.6 - 15.6), mean follow-up was 60 ± 42 months (6 - 191). The general features of uveitis were anterior, idiopathic and bilateral in this study similar to literature (Table 1). The most common systemic diseases associated with uveitis were juvenile idiopathic arthritis (JIA). Two patients improved with local medications, while the remaining 35 patients required systemic treatments such as short-time (oral/iv) corticosteroids (CS) in 94.5% of them, methotrexate (MTX) in 86.4%, azathioprine (AZA) in 5.4%, adalimumab (ADA) in 67.5%, tocilizumab (TCB) in 2.7%. In 26.1% of patients receiving ADA who did not respond to standard dose of ADA, we had to shorten the dosage intervals of ADA from every 2 weeks to every week. At least 1 ocular complication was observed in 83.7% of the patients, such as cataract, glaucoma, band keratopathy, synechiae, macular edema and retinal detachment. Four (10.8%) patients had moderate visual loss and 6 (16.2%) patients severe visual loss (3). The prevalence of surgery in our study was 18.9% for cataract and glaucoma treatment.

Conclusion: Diagnosis and management of uveitis in childhood is complicated. Despite the new medication options, the advancements in diagnosis and surgical techniques, the complications are still high. Usage of shorter dose interval of ADA may be an alternative to control of the disease in patients with unresponsive to standard dosage of ADA. However large-scale clinical trials are required to assess the efficacy and safety of this treatment.

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