The evaluation of the static and dynamic balance disorders in patients with familial Mediterranean fever.

Keywords: Rehabilitation, Enthesitis

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Background: Familial Mediterranean fever (FMF) is an autoinflammatory disease characterized by self-limiting recurrent episodes/attacks of fever, serositis, arthritis, and erysipelas-like erythema[1]. Musculoskeletal manifestations such as arthralgia, enthesitis, exertional leg pain, myalgia, sacroiliitis, and lower extremity synovitis are also common in FMF [2]. The fact that the disease progresses with lifelong persistent inflammation, as well as attacks of arthritis, synovitis, and enthesitis in the lower extremities, decreased lower extremity proprioception, muscle weakness, osteopenia, and erythema may further contribute to deteriorated balance.

Objectives: The aim of this study is to examine the relationship between static and dynamic balance in patients with Familial Mediterranean fever (FMF).

Methods: FMF patients who met the modified Tel Hashomer criteria and healthy volunteers were included in the study. The demographic data, clinical characteristics, assessments of familial Mediterranean fever in children genetically diagnosed with the syndrome. The most widely used treatment in 80% of patients was methotrexate and 62.5% continued with this medication until the end of the observation. In 30% of the patients the treatment had to be modified to control the disease and the most used drugs were mycophenolate, azathioprine, cyclophosphamide and rituximab. When patients were discriminated by current treatments according to Foster’s stage (n: 120) patients with earlier Foster’s stages used methotrexate more frequently, reaching 60% considering Foster’s stages 0, I and II. Mycophenolate, azathioprine, and cyclophosphamide were used in patients with Foster’s stage II or higher, while rituximab was used mainly in Foster’s stage 4. Systemic steroids were used in 58% of the patients and 23% continued using it at the last visit. Adverse events that led to stopping treatment were observed in 12% of all treated patients (There were: dapsone, methotrexate, azathioprine, cyclophosphamide and mycophenolate). In Addition, one death due to sepsis secondary to methotrexate was recorded. Foster’s stage progressed in 31% of patients despite therapy and 8.1% (n:12) finished with blindness.

Conclusion: Most of the patients were diagnosed in recent years and three quarters present initial stages (Foster II or less). Due to the inflammatory, autoimmune, progressive and scarring nature of OMMP, early initiation of immunosuppressive drugs/immuno-suppressive therapy is essential, in order to suppress inflammation and avoid scarring and sequelae. Methotrexate seems to be a good starting alternative and requires fluid compliance according to interdisciplinary approach and follow-up.

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OCULAR MUCOUS MEMBRANE PEMPIGOID: A RHEUMATOLOGY MULTICENTER REPORT

Keywords: Descriptive Studies, Rare/orphan diseases

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Background: Mucous membrane pemphigoid is a systemic, chronic, cicatrical inflammatory disease of autoimmune etiology of low incidence, characterized by the appearance of subepithelial bullous and scarring lesions that detach from the mucous membranes throughout the body. Isolated ocular involvement has classically been called Ocular Cicatral Pemphigoid but currently guides prefer the term Ocular Mucous Membrane Pemphigoid (OMMP). This involvement can lead to irreversible blindness if not treated early. The goal of therapy is to suppress inflammation and prevent scarring and sequelae, and step therapy with systemic immunomodulatory/immunosuppressive drugs is often used to achieve this. The autoimmune systemic nature of this pathology and the lack of familiarity of ophthalmologists with the use of immunosuppressive drugs motivates the referral of these patients to specialists used to their management, for interdisciplinary approach and follow-up.

Objectives: To analyze the clinical/epidemiological data and the established treatments of patients with OMMP in different Argentinian rheumatology centers.

Methods: Observational and multicenter study. Medical records of patients diagnosed and re with OMMP by an ophthalmologist, from different rheumatology centers in the Argentine Republic, were reviewed from May 2006 to July 2022.

Results: One hundred forty-seven medical records of patients diagnosed with OMMP were analyzed, mostly with confirmatory biopsy. Seventy-two percent were female. The mean age at diagnosis was 64 years (SD13). The time from the first symptoms to diagnosis was 30 months (0-240). Most of the patients (72%) were diagnosed in the last 5 years. On the analysis of 120 patients who had reported Foster’s stage, 73% had stage II or less. (Fosterin: 0-9; 1/43; 2/63; 3/18; 4/14). Another associated autoimmune disease was presented in 29% and the most frequent was Sjogren’s Syndrome. The most widely used treatment in 80% of patients was methotrexate and 62.5% continued with this medication until the end of the observation. In 30% of the patients the treatment had to be modified to control the disease and the most used drugs were mycophenolate, azathioprine, cyclophosphamide and rituximab. When patients were discriminated by current treatments according to Foster’s stage (n: 120) patients with earlier Foster’s stages used methotrexate more frequently, reaching 60% considering Foster’s stages 0, I and II. Mycophenolate, azathioprine, and cyclophosphamide were used in patients with Foster’s stage II or higher, while rituximab was used mainly in Foster’s stage 4. Systemic steroids were used in 58% of the patients and 23% continued using it at the last visit. Adverse events that led to stopping treatment were observed in 12% of all treated patients (There were: dapsone, methotrexate, azathioprine, cyclophosphamide and mycophenolate). In Addition, one death due to sepsis secondary to cyclophosphamide was recorded. Foster’s stage progressed in 31% of patients despite treatment and 8.1% (n:12) finished with blindness.

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