sarcoidosis was more frequently observed in those patients with a previous history of neoplasia, while the association with hematological neoplasms was linked to a higher frequency of sarcoidosis involving ENT and bone marrow.
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FRI0616 EYE MANIFESTATIONS OF PATIENTS WITH MUCKLE-WELLS SYNDROME

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Background: CAPS is a rare autoinflammatory disease associated with mutations in the *CIAS1* gene, encoding for NLRP3 that result in overactivation of the inflammasome and systemic inflammation. Muckle-Wells syndrome (MWS) is a rare autosomal dominant disease which causes episodic fever attacks, sensorineural deafness, recurrent hives, arthritis and eye involvement.
Objectives: Here we present the findings of eye involvement in a family whose 11 members have MWS.
Methods: Clinical data was collected during the course of ongoing patient care.
Results: We evaluated the clinical features of 11 patients who were referred to a tertiary care center. The median age of the patients was 25 years (range: 9–65). The ratio of females /males was 1.2 (6/5). All patients had arthritis with exacerbation on exposure to cold and recurrent episodes of pink eye. The median age of onset of ocular involvement was 8 years (2–45). We observed severe eye involvement in 36% of our cases, including band keratopathy, severe damage of corneal stroma and neovascularization. Corneal involvement and clouding was detected in four patient. Two of those had the diagnosis of keratoconus as well. Patients with keratoconus had corneal scarring due to corneal hydrops verified with corneal topography. The other two patients with corneal clouding had bant keratopathy. One of those patient was a 17 year old girl who had recurrent uveitis with hypopyon which necessiated the use of intravitreal dexamethasone implant. She also had posterior synechia of the iris to the lens. The other eye of that patient had signs of phthisis bulbi. The other patient with bant keraopathy was 46 years old male who had optic atrophy and tractional fibrovascular membranes at the posterior pole of the eye. Anakinra was used for treatment of 5 cases, and canakinumab of 3 cases. It was observed that the frequency of conjunctivitis decreased after anti IL-1 therapy. There was no mutation detected in the study of MEFV (all exons), TNFRSF1A (exons 2 to 7), MVK (all exons), NLRP3 (all exons), NOD2 (exons 4, 8 and 9) and PSTPIP1 (exons 10 and 11) genes.
Conclusions: In this study, it has been shown that eye findings related to MWS can vary from conjunctivitis to severe uveitis. We want to emphasize that ocular involvement in MWS should be carefully assessed, since it can lead to visual impairment.
Disclosure of Interest: None declared
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FRI0617 DIAGNOSTIC SENSITIVITY OF CUTOFF VALUES OF IGG4-POSITIVE PLASMA CELL NUMBER AND IGG4-POSITIVE/CD138-POSITIVE CELL RATIO IN TYPICAL MULTIPLE LESIONS OF PATIENTS WITH IGG4-RELATED DISEASE

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Background: Immunoglobulin G4-related disease (IgG4-RD) is a recently recognized systemic inflammatory disease with multi-organ involvement [1]. Diagnostically, two of the most important hallmarks of IgG4-RD are high IgG4-positive plasma cell (PC) counts and high IgG4/IgG ratios in affected organs. Although the International consensus statement (ICS) on the pathology of IgG4-RD adopted different IgG4-positive PC counts among affected organs for the diagnosis to differentiate IgG4-RD mimickers from IgG4-RD [2], histological and immunohistochemical findings of the specimens from not only one but multiple organs in the same patient has not been evaluated.
Objectives: This study aimed to investigate the diagnostic sensitivity of the cutoff values of IgG4-positive PC number and IgG4-positive/CD138-positive cell ratio proposed by the International consensus statement (ICS) on the pathology of IgG4-RD in typical multiple lesions of patients with IgG4-RD.
Methods: We evaluated IgG4-positive PC number and IgG4-positive/CD138-positive cell ratio in 35 samples from 16 IgG4-RD patients having more than two typical lesions of IgG4-RD.
Results: We evaluated twelve submandibular, eleven ophthalmic, four skin, four kidney, two pancreatic, and one bronchus and prostate lesion each in 16 patients with typical clinical, serological, and radiographic features. Concerning IgG4+ PC number per high power field, most ophthalmic (8/11), kidney (4/4), pancreatic (2/2), and bronchial lesions (1/1) fulfilled the cutoff value of ICS, whereas many of the submandibular (5/12) and skin lesions (0/4) did not. In contrast to the absolute number, almost all lesions fulfilled the cutoff value of IgG4+/CD138+ cell ratio. In five cases, only one or two lesions in the same patient fulfilled the cutoff value of ICS, while the others did not.
Conclusions: These results suggest that ICS criteria have different sensitivities among the affected organs in diagnosing IgG4-RD.
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FRI0618 ADALIMUMAB IN NON-INFECTIOUS UVEITIS – EFFICACY ACROSS DIFFERENT ETIOLOGIES IN THE VISUAL I AND VISUAL II TRIALS

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Background: There is increasing interest in understanding the efficacy of adalimumab across different etiologies of uveitis. No prospective analysis has been conducted to date to determine the efficacy of adalimumab among non-infectious uveitis patients with different etiologies.
Objectives: To assess adalimumab (ADA) efficacy in active and inactive, non-infectious uveitis across different etiologies in patients who were recruited as part of the VISUAL program.
Methods: Exploratory data analyses from two global phase 3, double-masked trials: VISUAL I (patients with active uveitis despite ≥2 weeks of prednisone 10–60 mg/day) and VISUAL II (patients with inactive disease dependent on 10–35 mg/day of prednisone to maintain inactivity) were performed. Patients received placebo (PBO) or ADA subcutaneously (80 mg week 0, followed by 40 mg every other week from week 1 up to 80 weeks). In VISUAL I, all patients received a prednisone burst followed by taper to 0 mg by week 15. In VISUAL II, prednisone taper to 0 mg was mandatory by week 19. The primary endpoint was time to treatment failure (TF) at or after week 6 for VISUAL I; and at or after week 2 for VISUAL II[1,2]. For this analysis, patients were categorized into different uveitis etiologies which they presented at study entry. Hazard ratios (HR) for time to TF were obtained for each uveitis etiology.
Results: The efficacy of ADA was significantly greater than PBO in the largest subgroup of patients with Idiopathic/other uveitis (VISUAL I: 103 and VISUAL II: 90) etiology in both VISUAL I[1] and VISUAL II trials. All other subgroups showed

Figure: Hazard ratios of time to treatment failure by uveitis etiologies.
VISUAL I (A) and VISUAL II* (B) clinical trials.