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P082

Clinical and laboratory features of macrophage activation syndrome associated with systemic juvenile idiopathic arthritis receiving tocilizumab and canakinumab

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Introduction: Macrophage activation syndrome (MAS) can still occur in sJIA patients receiving tocilizumab (TCZ) or canakinumab (CAN). Previous reports showed that TCZ and CAN mask the clinical features of MAS, making early diagnosis difficult compared to MAS in biologics (Bio) naïve sJIA patients. Despite these observations, detailed comparative data and the specific clinical differences among them remain fully unknown.
Objectives: The aim of this study is to investigate the clinical and laboratory features of MAS in patients with sJIA receiving TCZ and CAN.
Methods: A total of 14 pediatric rheumatology centers in Japan participated in a study that involved the collection of MAS episodes in patients diagnosed with sJIA and who had received treatment with or without TCZ or CAN. The diagnosis of MAS was based on the criteria proposed by Ravelli A, et al. Patients who did not fulfil all the diagnostic criteria but were clinically considered to have MAS by the attending physician were categorized as possible MAS. A total of 63 episodes of MAS were evaluated according to clinical features, disease courses, laboratory data, and treatment plans. Furthermore, we compared the timing of MAS onset and the point of therapeutic intervention (TI) which means full-blown MAS among the three groups: Bio naïve, TCZ, and CAN groups. These points were determined according to Delphi method of all members.
Results: Thirty MAS episodes in Bio naïve group, 21 episodes in TCZ group, and 12 episodes in CAN group were included in this study. Possible MAS episodes were 1 in Bio naïve, 6 in TCZ, and 3 in CAN group. The incidence of no fever in MAS was 3% in Bio naïve, 66% in TCZ, and 25% in CAN group. Duration from onset to MAS was 4.3 years in TCZ group, and 6.8 years in CAN group. Serum ferritin and CRP, D-dimer levels were significantly decreased in TCZ and CAN group compared to Bio naïve group at MAS onset and TI. AST levels were significantly elevated in TCZ group compared to Bio naïve and CAN group at MAS onset. Platelet counts has no significance among three groups at onset and TI.
Conclusion: MAS in sJIA patients receiving TCZ or CAN often lacks the typical clinical features. Despite showing lower ferritin levels compared to Bio naïve-MAS, sJIA patients receiving TCZ or CAN may have a potential risk for developing MAS.
Disclosure
None declared

P083

Descriptive study of primary versus rheumatologic and infectious secondary hemophagocytic lymphohistiocytosis in pediatric population. value of cholesterol in disease monitoring

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Pediatric Rheumatology, **23(2):** P083

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is a life-threatening hyperinflammatory syndrome, either primary (F-HLH) or secondary to infections or autoimmune diseases, including macrophage activation syndrome (MAS). It is well known that increased triglycerides are prevalent in patients with HLH and is used as one of the diagnostic criteria by the International Society of Histiocytes. However, lipoprotein cholesterol levels have been rarely studied in HLH.
Objectives: 1) To analyze the clinical and laboratory features of pediatric patients with HLH: primary (F-HLH), MAS, and infection-associated HLH (HLH-NOS); 2) To correlate lipid profile with disease activity.
Methods: Retrospective observational study of patients ≤18 years diagnosed with HLH and treated at our center between 2000 and 2023.
Results: A total of 41 patients were included: F-HLH (A) (n=7), MAS (B) (n=27), and HLH-NOS (C) (n=7). Both F-HLH and HLH-NOS presented an early age (years) of onset (A=1.3; B=9.7; C=1.2 *(p=0.043)*; and absence of relevant history prior to HLH (A=100%; B=55.6%; C=100%; *p=0.012*).At diagnosis, F-HLH showed higher rates of hepatomegaly (A=100%; B=37%; C=28.6%; *p=0.007*), splenomegaly (A=100%; B=40.7%; C=85.7%; *p=0.005*), and more severe pancytopenia: hemoglobin (g/dL) (A=8.3; B=10.8; C=8.4; *p=0.002*), platelets (x10^6/mm^3) (A=30; B=141; C=65; *p<0.000*), leukocytes (x10^6/mm^3) (A=2.9; B=9.0; C=3.9; *p=0.008*), neutrophils (x10^6/mm^3) (A=1.0; B=5.8; C=1.2; *p=0.007*), hypertriglyceridemia (mg/dL) (A=401; B=186; C=207; *p*=0.018), elevated AST (U/L) (A=474; B=110; C=99; *p=0.030*), and impaired cytotoxicity (A=83.3%; B=25%; C=20%; *p=0.035*). MAS patients had a more frequent personal history of autoimmunity (B=48.1%; *p=0.007*), arthralgia (B=59.3%; *p*=0.005), and arthritis (B=37%; *p=0.032*). Low HDL cholesterol levels at admission were found in 51.2% of patients (A=3/3; B=13/16; C=2/2; *p=0.037*), and at diagnosis in 65.8% (A=3/3; B=16/22; C=2/2; *p=0.114*). Ferritin/HDL ratio showed a significant inverse correlation (r = - 0.356; *p < 0.001*), persisting throughout HLH episodes in all three groups.
Conclusion: Three distinct clinical-laboratory profiles were identified across pediatric HLH types. HDL cholesterol may serve as a useful biomarker for HLH activity monitoring, inversely correlated with ferritin values
Disclosure
None declared

P085

Ultrasonographic evaluation of exertional leg pain in patients with familial mediterranean fever: a silent component of the disease?

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Pediatric Rheumatology, **23(2):** P085

Introduction: Exertional leg pain (ELP) is a common finding in patients with Familial Mediterranean Fever (FMF). Although its pathophysiology has not been fully elucidated, the most widely accepted hypothesis is that it represents a silent enthesopathy.
Objectives: The aim of this study is to investigate the radiological manifestations and clinical significance of ELP in patients with FMF through ultrasonographic evaluation of lower extremity joints and tendons
Methods: Patients diagnosed with FMF and followed in our clinic between June and September 2024, who were found to carry homozygous or compound heterozygous pathogenic variants in the MEFV gene, were included in the study. Patients with comorbid musculoskeletal diseases, active professional athletes, and those with a follow-up period of less than one year were excluded. All included patients underwent bilateral ultrasonographic (USG) examination of the hip, knee, and ankle joints, as well as assessment and thickness measurements of the patellar tendon, Achilles tendon, and plantar fascia. USG images were obtained by a single blinded investigator and evaluated according to the OMERACT pediatric USG guidelines.
Results: A total of 50 patients were included in the study, comprising 25 FMF patients with ELP and 25 without. No significant differences were observed in demographic characteristics, MEFV variants, disease activity, or treatment regimens between groups. Joint USG revealed grade 1 synovial effusion in nine patients—seven of whom were in the ELP group—synovial hypertrophy in three patients, and loss of the normal fibrillar tendon structure in one patient. Among those with synovial effusion, 6 patients (67%) had elevated acute phase reactants and active disease. The Achilles tendon was found to be significantly thicker in the ELP group compared to those without ELP (right and left median values: 0.50 and 0.42 cm vs. 0.39 and 0.36 cm, p=0.003 and p=0.009, respectively). This difference remained significant even after adjusting for factors known to affect tendon thickness, such as age and sex, using a multivariate linear regression model (B= 0.065, p=0.007). No differences were observed between the groups in other tendon or plantar fascia thickness measurements.
Conclusion: Although the pathophysiology of ELP in FMF remains unclear, this study suggests that it may be associated with increased Achilles tendon thickness. This finding may indicate subclinical enthesopathy and highlight the potential need for further investigation and treatment modification. Moreover, synovial effusion may be present in FMF patients even in the absence of symptoms or physical examination findings and is generally associated with increased disease activity.
Disclosure
None declared

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P087

The contribution of salivary gland ultrasonography to the classification criteria in juvenile sjögren disease: diagnostic value and predictive performance of a weighted scoring model

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Pediatric Rheumatology, **23(2):** P087

Introduction: Juvenile Sjögren's disease (jSjD) is a rare and diagnostically challenging autoimmune condition that lacks pediatric-specific classification criteria.
Objectives: This study evaluated the diagnostic utility of salivary gland ultrasonography (US)—not currently included in the 2016 ACR/EULAR criteria—in jSjD, and assessed its performance within a weighted scoring model (WS) incorporating conventional parameters.
Methods: Seventeen patients diagnosed with jSjD between 2017 and 2024 were retrospectively analyzed. All underwent B-mode parotid US scored using the OMERACT system. Diagnostic parameters included anti-SSA (Ro) positivity, Schirmer test (≤5 mm/5 min), labial salivary gland biopsy with focus score ≥1, and salivary gland US score ≥1. The WS (range 0–8) was calculated as:

$$WS = (\text{anti–SSA positivity} \times 3) + (\text{labial salivary gland biopsy positivity} \times 3) + (\text{Schirmer test positivity} \times 1) + (\text{parotid gland ultra sound positivity} \times 1)$$

ROC analyses evaluated the WS's ability to predict patients exceeding the ACR/EULAR threshold (>4 points), and the predictive value of US alone in identifying patients with ≥2 conventional criteria (Composite Score ≥2). WS and the classic model (excluding US) were compared by AUC and diagnostic metrics.
Results: Among 17 patients (94.1% female, n=16), salivary gland US score ≥1 was present in 64.7% (n=11), Schirmer test was positive in 41.2% (n=7), anti-SSA was positive in 35.3% (n=6), and biopsy was positive in 88.2% (n=15). The median US score was 1.0 (IQR: 0.0–2.0), median biopsy focus score was 2.0 (IQR: 1.0–2.0), and median WS was 4.0 (IQR: 4.0–5.0). The WS yielded an AUC of 0.72 for predicting ACR/EULAR positivity, indicating an acceptable level of discriminative performance. US alone showed strong alignment with classification components (AUC: 0.84 for Composite Score ≥2). ROC comparison revealed similar overall performance between WS and the classic model (AUC: 0.85 vs. 0.87; p = 0.76). However, WS had superior specificity (92% vs. 77%) and PPV (86% vs. 73%), enhancing its diagnostic value in borderline cases.
Conclusion: Although adding US to the classification model did not significantly increase overall AUC, it improved specificity and PPV. Its strong concordance with core criteria, noninvasive nature, and feasibility in pediatric settings support salivary gland US as a promising adjunct to jSjD classification—potentially substituting more impractical tests such as unstimulated salivary flow rate and ocular staining score. However, prospective validation in larger, multicenter pediatric cohorts is essential before its integration into formal classification systems.
Keywords: Classification criteria, Juvenile Sjögren, salivary gland ultrasonography
Disclosure
None declared

P089

The role of PET/CT in evaluating rheumatologic conditions in children with fever of unknown origin - a multicenter study

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Introduction: Fever of Unknown Origin (FUO) is a relatively common clinical challenge that is frequently referred for evaluation by pediatric rheumatologists. In many cases, it is necessary to rule out other medical conditions before establishing a rheumatologic diagnosis. 18F-FDG PET/CT is a valuable diagnostic tool that may help identify the fever's source. However, its use in children involves risks, including radiation exposure and the need for sedation.
Objectives: To assess the contribution of PET/CT to FUO diagnosis in hospitalized children, and to identify rheumatologic conditions that are either detected through the evaluation or excluded by it. In addition, to identify predictive factors for its diagnostic utility.
Methods: A retrospective cohort study reviewing medical records of children (0-18 years) diagnosed with FUO who underwent PET/CT

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Pediatric Rheumatology

MEETING ABSTRACTS

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O01

Reconstitution of the B cell compartment following anti-CD19 CAR-T cell therapy in refractory patients with Juvenile Dermatomyositis (JDM)

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Pediatric Rheumatology, **23(2):** O01

Introduction: CD19 CAR T-cell therapy is an innovative strategy to induce profound B cell depletion in patients with refractory idiopathic inflammatory myopathies (IIM). However, its effects on B cell phenotypes in pediatric patients remain largely unknown.
Objectives: To characterize the reconstitution profile of B cell subsets in three patients with juvenile dermatomyositis (JDM) treated with anti-CD19 CAR T-cell therapy.
Methods: Three patients with refractory JDM received a single infusion of autologous, second-generation anti-CD19 CAR T cells (1×10^6 CAR T cells/kg; lentiviral vector), manufactured on the CliniMACS Prodigy system. Lymphodepletion was achieved using cyclophosphamide (1000 mg/m^2 over 2 days) and fludarabine (90 mg/m^2 over 3 days). B cell immunophenotyping was performed according to standard operating procedures (SOPs) before and after CAR T-cell infusion. A comparator group of five JDM patients treated with rituximab (RTX) was included.
Results: B cell reconstitution occurred at a median of 8 weeks (IQR: 8.0-12.0) post-CAR T therapy, with an immature transitional B cell phenotype. B cell subsets distribution was assessed at a median of 27.3 weeks (IQR: 22.5–40.2) after treatment, when all B cell subsets were present for each patient, and compared to pre-treatment profiles. In the RTX group, B cell phenotyping was performed at a median of 47 weeks (IQR: 33.4–50.7) post-treatment. The proportion of CD19$^+$ B cells among lymphocytes was similar before and after CAR T-cell treatment (pre: 32% [IQR: 23–40], post: 21% [IQR: 20–24]). B cell subset analysis showed comparable levels of transitional, naïve and plasmablast populations pre- and post-treatment. However, there was a notable reduction in total memory B cells (MBCs) post-treatment (pre: 6.4% [IQR: 6.3–9.1], post: 3.9% [IQR: 3.1–4.0]), particularly in the switched MBC subset. Interestingly, JDM patients treated with RTX showed higher frequencies of switched MBCs during B cell reconstitution compared to those treated with CAR T cells: CAR T treated-patients 16.0 % [IQR: 8.5-18.0], RTX group 52.0% [IQR: 28.0, 60.0].
Conclusion: These data show that following CAR T cell treatment, the memory B cell compartment presents a lower frequency of switched MBCs when compared to pre-treatment levels and to patients treated with RTX, suggesting a specific effect of CAR T cell treatment in regulating the memory B cell compartment in JDM patients.
Disclosure
None declared

O02

Anti-CD19 Car-T cell therapy in refractory juvenile dermatomyositis (JDM): a single-center case series

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Pediatric Rheumatology, **23(2):** O02

Introduction: CD19 CAR T-cell therapy represents an innovative approach emerging as a promising treatment option in patients with refractory idiopathic inflammatory myopathies (IIM), but the experience in children so far is limited.
Objectives: To report on the safety and efficacy of anti-CD19 CAR T-cell therapy in 3 JDM patients.
Methods: All 3 patients received a single infusion of fresh, autologous second-generation anti-CD19 CAR T product (lentiviral vector) manufactured on the Prodigy device ($1x10^6$ CAR T cells/kg), after lymphodepletion with cyclophosphamide (1000 mg/m^2 over 2 days) and fludarabine (90 mg/m^2 over 3 days).
Results: Maximum follow-up after CAR T-cell therapy was 20 months. Patients 1 and 2 had a history of severe chronically active disease with extensive calcinosis and ulcerations. Patient 3 presented interstitial lung disease (ILD) and persistently active muscle and skin disease with ulcerations. Short-term effects of CAR T-cell therapy on Patient 1 have been previously reported (Nicolai A&R 2024). All patients were refractory to multiple immunosuppressive treatment lines and presented significant side effects from chronic glucocorticoid treatment. After infusion, CAR T cells rapidly expanded, reaching peak concentrations after a median of 7 days (range 7-9). At peak, the median number of circulating CAR T cells was 32.69 per microliter (range 31.04-535.02). CD19+ B cells were undetectable in peripheral blood after a mean of

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