SAT0539  MUCKLE-WELLS SYNDROME IN RHEUMATOLOGY PRACTICE, FAMILY CASES: FEDERAL CENTER EXPERIENCE.

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Background: Muckle-Wells syndrome (MWS) is a monogenic autoinflammatory disease caused by a NLRP3 gene mutation. It is the most common variant of cryopyrin-associated periodic syndromes (CAPSs) and can be observed in rheumatology practice. It manifests itself in fever, urticaria-like rash, arthralgias/arthritides, conjunctivitis/uveitis, sensorineural hearing loss, acute-phase markers of cryopyrin-associated periodic syndromes (CAPSs) and can be observed in rheumatology practice. It is very important to acquire family medical history to identify affected family members and prescribe therapy in a timely manner. IL-1 inhibitors are an effective and safe treatment option for MWS patients.

Methods: This was a single-centre prospective observational study including patients referred for musculoskeletal symptoms while treated with ICI. After baseline rheumatological evaluation defining the clinical entity presented, follow-up visits were organised according to the type and severity of irAE. At one year, persistence of irAE, ongoing treatment, as well as cancer outcomes were assessed.

Results: 63 patients were included between September 2015 and June 2016. 24 patients (38%) presented with non-inflammatory musculoskeletal conditions managed with short-term symptomatic treatment and did not require specific follow-up. 39 patients (62%) experienced inflammatory manifestations, mimicking either rheumatoid arthritis (RA, n=19), polymyalgia rheumatica (PMR, n=16), psoriatic arthritis (PsA, n=3) and one flare of a pre-existing axial spondyloarthritis. Overall, 32 patients (52%) received systemic glucocorticoids, with a median dosage of 15mg/day (range: 5-60mg/day). None of the patients had to permanently discontinue ICI therapy for rheumatic irAE. 20 patients (67%) were still receiving glucocorticoids at one year, with a median dosage of 5mg/day (range: 2-20mg/day). Glucocorticoids were more frequently discontinued for patients with RA-like condition (44%) than PMR-like condition (23%), but no other predictive factor of glucocorticoids withdrawal could be identified. At one year, overall survival and progression-free survival were comparable between patients who were still receiving glucocorticoids for rheumatic irAE and patients who had discontinued. Eight patients required csDMARDs. 8 patients referred for musculoskeletal symptoms while treated with ICI. After baseline rheumatological evaluation defining the clinical entity presented, follow-up visits were organised according to the type and severity of irAE. At one year, persistence of irAE, ongoing treatment, as well as cancer outcomes were assessed.

Disclosure of Interests: None declared

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SAT0540  ONE-YEAR OUTCOMES AFTER RHEUMATIC IMMUNE-RELATED ADVERSE EVENTS FROM CHECKPOINT INHIBITORS

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Background: Description and initial management of rheumatic immune-related adverse events (irAEs) from cancer immunotherapies have been reported by several groups but to date, few studies have evaluated the long-term outcomes and management of rheumatic irAEs (1).

Objectives: To describe the long-term management and assess the one-year outcomes of patients who experienced rheumatic immune-related adverse events (irAEs) due to immune checkpoint inhibitors (ICI).

Methods: Within a retrospective cohort of 112 patients referred for musculoskeletal symptoms while treated with ICI. After baseline rheumatological evaluation defining the clinical entity presented, follow-up visits were organised according to the type and severity of irAE. At one year, persistence of irAE, ongoing treatment, as well as cancer outcomes were assessed.

Results: Sixty-three patients were included between September 2015 and June 2016. Twenty-four patients (38%) presented with non-inflammatory musculoskeletal conditions managed with short-term symptomatic treatment and did not require specific follow-up. Thirty-nine patients (62%) experienced inflammatory manifestations, mimicking either rheumatoid arthritis (RA), polymyalgia rheumatica (PMR), psoriatic arthritis (PsA), and one flare of a pre-existing axial spondyloarthritis. Overall, thirty-two patients (52%) received systemic glucocorticoids, with a median dosage of 15mg/day (range: 5-60mg/day). None of the patients had to permanently discontinue ICI therapy for rheumatic irAE. Twenty patients (67%) were still receiving glucocorticoids at one year, with a median dosage of 5mg/day (range: 2-20mg/day). Glucocorticoids were more frequently discontinued for patients with RA-like condition (44%) than PMR-like condition (23%), but no other predictive factor of glucocorticoids withdrawal could be identified. At one year, overall survival and progression-free survival were comparable between patients who were still receiving glucocorticoids for rheumatic irAE and patients who had discontinued. Eight patients required csDMARDs. Ten patients referred for musculoskeletal symptoms while treated with ICI. After baseline rheumatological evaluation defining the clinical entity presented, follow-up visits were organised according to the type and severity of irAE. At one year, persistence of irAE, ongoing treatment, as well as cancer outcomes were assessed.

Disclosure of Interests: None declared

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