AB1454

DIAGNOSTIC AND THERAPEUTIC PRACTICES IN ADULT CHRONIC NONBACTERIAL OSTEOMYELITIS (CNO)/SAPHO SYNDROME: AN INTERNATIONAL LANDSCAPE

Keywords: Quality of care, Bone diseases, Rare/orphan diseases

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Background: Chronic nonbacterial osteomyelitis (CNO) is a rare auto-inflammatory bone disease. CNO, especially the adult variant, may occur in the broader spectrum of Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis (SAPHO) syndrome. Adult CNO lacks diagnostic or management guidelines in contrast to pediatric disease (1, 2).

Objectives: We mapped current diagnostic and therapeutic practices for CNO/ SAPHO in adults.

Methods: A primary survey was spread among global rheumatological/bone networks and 57 experts were identified from literature (May 2022), covering terminology, diagnostic tools (clinical, radiological, biochemical) and treatment steps. A secondary survey (sent to primary survey responders in August 2022) further queried diagnostic features, treatment motivations, disease activity and treatment response monitoring.

Results: 36 and 23 physicians completed the primary and secondary survey respectively. Diagnosis was mainly based on individual physician assessment, in which the combination of chronic relapsing-remitting bone pain with radiologically-proven osteitis/osteomyelitis, sclerosis, hyperostosis and increased isotope uptake on bone scintigraphy were reported indicative of CNO. Physicians appeared more likely to refer to the condition as SAPHO syndrome in the presence of joint and skin pathology. MRI was most frequently performed as imaging diagnostic, while biochemical and histopathological activity and therapy response in CNO/SAPHO.

REFERENCES:

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Disclosure of Interests: None Declared.

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AB1455

FLUORIDE UPTAKE ON QUANTITATIVE POSITRON EMISSION TOMOGRAPHY/COMPUTED TOMOGRAPHY (NA18F-PET/CT) AS NOVEL BIOMARKER FOR DISEASE ACTIVITY IN ADULT CHRONIC NONBACTERIAL OSTEOMYELITIS/SYNOVITIS ACNE PUSTULOSIS HYPEROSTOSIS OSTEITIS SYNDROME (CNO/SAPHO)

Keywords: Bone diseases, Imaging, Rare/orphan diseases

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Background: Chronic nonbacterial osteomyelitis (CNO) is a rare bone disease characterized by sterile bone inflammation and locally increased bone turnover. CNO that occurs in the rheumatic spectrum of synovitis, acne, pustulosis, hyperostosis, and osteitis is referred to as SAPHO syndrome [1, 2]. To date, there are no objective biomarkers for monitoring disease activity. Recommended imaging tools like technetium radio labelled hydroxyethylidiphosphonate single photon emission computed tomography visualize the increased bone turnover, but fail to reflect the relapsing-remitting disease activity course due to an imprinting pattern [3]. Likewise, magnetic resonance imaging can track the fluctuation of bone marrow edema, but is inferior compared to computed tomography (CT) in capturing subtle inflammatory and accumulated structural changes that reflect disease duration. Sodium fluoride-18 positron emission tomography/CT (Na18F-PET/CT) is another imaging modality which produces quantitative data on the spatial distribution of bone turnover that also correlate with clinical disease activity in other metabolic bone diseases [4].

Objectives: We quantify bone disease activity progression in CNO/SAPHO patients to evaluate its capacities as a disease activity biomarker.

Methods: Cohort study including 43 CNO/SAPHO patients not using immunomodulatory or antiresorptive medications with Na18F-PET/CT performed at our expert clinic between 2019-2022. Images were qualitatively assessed following a systematic reporting format. Maximal standardized uptake values (SUVmax) were determined in the following areas as defined by radiological appearance on CT: osteitic areas (ossesous lesions), inflamed ligaments or joint spaces (soft tissue lesions) and thoracic vertebrae 5 as reference bone (ref-bone).

Results: Qualitative assessment revealed sclerosis and hyperostosis of the manubrium as most prevalent radiologic features (present in 77% and 70%) and costae 1-2 respectively. Diagnosis was mainly based on individual physician assessment, in which the combination of chronic relapsing-remitting bone pain with radiologically-proven osteitis/osteomyelitis, sclerosis, hyperostosis and increased isotope uptake on bone scintigraphy were reported indicative of CNO. Physicians appeared more likely to refer to the condition as SAPHO syndrome in the presence of joint and skin pathology. MRI was most frequently performed as imaging diagnostic, while biochemical and histopathological activity and therapy response in CNO/SAPHO.

REFERENCES:

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AB1456

BIOLOGICS AND JAK INHIBITORS EFFICACY IN VESICULAR SYNDROME FROM FRENCH MULTICENTER CASE SERIES OF 256 PATIENTS

Keywords: Innate immunity, Adaptive immunity, Inflammatory arthritides

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AB1457 MULTIDISCIPLINARY PROSPECTIVE STUDY OF PATIENTS TREATED WITH IMMUNE CHECKPOINT INHIBITORS WHO DEVELOPED RHEUMATIC IMMUNE-RELATED ADVERSE EVENTS

Keywords: Descriptive Studies, Organ damage, Epidemiology

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Background: Immune checkpoint inhibitors (ICIs), by activating the immune system (specifically, T-cells), foster the reaction against tumor cells. However, paradoxically, autoimmune phenomena, known as immune-related adverse effects (ir-AEs) (PMID 33902019), 294924520), can be triggered and manifest in any organ or tissue. The most common rheumatic manifestations are inflammatory arthritis, polymyalgia rheumatica, and myositis, but other inflammatory cases have also been described (PMID 32403289). More data on their frequency and characterization are needed.

Objectives: To prospectively evaluate the incidence of rheumatic ir-AEs during ICIs treatment, along with clinical characterization, management required, and outcomes.

Methods: An observational, prospective study was conducted at a tertiary center in Spain, led by the oncology department with the participation of several specialties, to evaluate the occurrence of ir-AEs in patients starting ICIs between January 2019 and April 2022. Participants were routinely followed at oncology clinics to detect ir-AEs through pre-specified clinical and laboratory assessments. For rheumatic symptoms, ir-AEs were studied by records review and evaluated in person at rheumatology clinics for those with a degree of involvement of ≥2, according to the ASCO guidelines [3]. The incidence - with a 95% confidence interval (CI) - and characterization of defined rheumatic ir-AEs are presented here.

Results: Of 181 patients, 21 (11.6%, 95%CI 7.7-17.1%) developed rheumatic ir-AEs, 13 men (61.9%) with a median age of 62.3 years (p25-75 51.8-75.0). The median time from the start of ICIs to the development of rheumatic ir-AEs was 85 days (p25-75 51.5-165). Blood tests for autoimmunity were positive in 69.2% of available cases (9/13), but at all a low titer (table 1). According to ASCO guidelines, most patients had a toxicity grade 1-2, but 3 (14.3%) patients presented with severe manifestations (grade 3): 1 (4.8%) case of inflammatory arthritis, 1 (4.8%) of xerostomia, and 1 (4.8%) of Raynaud’s phenomenon with ulcers. 9 (42.9%) patients also presented with concurrent ir-AEs of different types. Rheumatic ir-AEs were successfully settled, though infliximab and intravenous vasodilators were required for some cases. While discontinuing ICIs was mostly due to neoplasm progression (47.6%), in 3 cases (14.3%) it was due to grade-3 rheumatic manifestations. 7 (33.3%) patients died during follow-up due to the oncological disease, no case due to rheumatic toxicity.

Table 1

<table>
<thead>
<tr>
<th>Type of tumor/Drug</th>
<th>n (%)</th>
<th>Clinical presentation (%)</th>
<th>Antibodies n (%)</th>
<th>Treatment n (%)</th>
<th>Outcome n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lung</td>
<td>9 (42.9)</td>
<td>8 (88.9)</td>
<td>8 (88.9)</td>
<td>No rheumatological</td>
<td>Discontinuation of ICIs</td>
</tr>
<tr>
<td>Melanoma</td>
<td>5 (22.7)</td>
<td>7 (77.8)</td>
<td>5 (55.6)</td>
<td>Anti-Teconomic</td>
<td>Discontinuation</td>
</tr>
<tr>
<td>Hodgkin’s lymphoma</td>
<td>2 (9.5)</td>
<td>2 (100)</td>
<td>1 (50)</td>
<td>Anti-CD20</td>
<td>Discontinuation</td>
</tr>
<tr>
<td>Kidney</td>
<td>2 (9.5)</td>
<td>2 (100)</td>
<td>1 (50)</td>
<td>Anti-CD20</td>
<td>Discontinuation</td>
</tr>
<tr>
<td>Esophagus</td>
<td>1 (4.8)</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>Anti-CD20</td>
<td>Discontinuation</td>
</tr>
<tr>
<td>Bladder</td>
<td>1 (4.8)</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>Anti-CD20</td>
<td>Discontinuation</td>
</tr>
<tr>
<td>Mesothelioma</td>
<td>1 (4.8)</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>Anti-CD20</td>
<td>Discontinuation</td>
</tr>
</tbody>
</table>

Conclusion: Through a prospective and multidisciplinary study, we estimated an 11.6% occurrence of rheumatic ir-AEs in patients under ICIs. Most presented with mild or moderate involvements, though severe cases were also seen. A coordinated approach with the oncologists is thus essential for patients treated with ICIs at risk of developing ir-AEs.

REFERENCES: NIL.

Acknowledgements: NIL.

Disclosure of Interests: None Declared.

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AB1458 TREATMENT UTILIZATION, SYMPTOMS, AND COMORBIDITIES IN DERMATOMYOSITIS: AN ANALYSIS OF ELECTRONIC MEDICAL RECORDS IN THE UNITED STATES

Keywords: Comorbidities, Myositis, Real-world evidence

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Background: Dermatomyositis (DM) is a rare, chronic inflammatory disease characterized by skin manifestations and/or progressive muscle weakness and other systemic manifestations. Prior analyses have reported higher morbidity, hospitalization rates, and mortality among patients with DM compared with matched controls[1-3]; however, more recent data on the disease burden and treatments for DM are warranted.

Objectives: We conducted this analysis to provide updated data on the treatment utilization, symptoms, and comorbidities recorded for patients with myositis DM in the United States.

Methods: This was a descriptive, retrospective cohort analysis that used TriNetX US electronic medical records (EMR) of adults who had been diagnosed myopathic DM (ICD-9 codes: 710.3/ICD-10: M33.1x, M33.9x, M36.0) between 1 January 2007 and 1 September 2020. Key inclusion criteria were: age ≥18 years at index date (i.e., date of DM diagnosis), ≤6 months of baseline data before the index date, and ≥6 months of follow-up after the index date. Assessments included utilization rates for medications of interest prior to the index date, percentage of patients who received ≥2 consecutive unique non-steroidal immunosuppressive therapies within 12 months of the index date, classification and qualification of the most common post-index symptoms and comorbidities, along with their comparison to pre-index values.

Results: The TriNetX database contained 1097 patients with DM (mean age: 54.6 years; sex: 77% female). The mean observation period was 9.7 years (6.0 years pre-index; 3.7 years post-index). Prior to the DM index date, 60% of patients...