Conclusion: According to MRI findings, hip damage was characterized for all ages of FOP patients. Sacroilitis we found mostly in patients older 10 y.o. Based on our experience of JAK-inhibitor therapy we can suggest about positive effect and advantages of TOFA over other therapy approaches. Dynamics of MRI imaging may be good option for the confirmation of therapeutic anti-inflammatory effects.

Disclosure of Interests: None declared


CRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS (CRMO): NEW INSIGHTS INTO EXTRA-OSSEOUS MANIFESTATIONS.

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Background: Chronic recurrent multifocal osteomyelitis (CRMO) is a rare inflammatory disease of the skeleton characterized by chronic and recurrent episodes of osteo-articular inflammation. The median age of onset is 10 years old. Clinical manifestations include musculoskeletal symptoms that are well described (pain, tenderness, swelling). Sometimes, skin lesions or digestive manifestations occur. Whole body magnetic resonance imaging (MRI) is the gold standard for assessing the multifocal pattern of the CRMO. Treatment is still empirical and mainly relies on non-steroidal anti-inflammatory drugs (NSAIDs). Bisphosphonates and biologics are used as second-line treatments. To date, the focus has been set on bone involvement and very few data are available on extra-osseous manifestations in CRMO.

Objectives: This study aims to further describe these extra-osseous clinical manifestations in CRMO.

Methods: A historical cohort was designed using 61 CRMO patients at the Pediatrie du Nord, Aix-en-Provence, in a tertiary university hospital in Paris (Hôpital Bicêtre, France). All patients underwent a MRI that confirmed the diagnosis according to the criteria of Jansson. Skeletal involvement was characterized with 1/axial, 2/ peripheral, 3/ axial and peripheral lesions. Extra-osseous manifestations were divided into 1/ skin lesions, 2/ gastro-intestinal manifestations, 3/ enthesitis and 4/ others. Pain was evaluated thanks to the Visual Analog Scale (VAS, from 0 to 10). Treatments used were recorded. The study complied with ethical requirements.

Results: Forty one patients were included in the study, with 31 females (75.6%). The mean ± SD age at onset was 79.1 ± 59.8 months, with a delay at diagnosis beyond six months (6.71 ± 6.96). Twenty-one patients had a familial history of inflammatory diseases (51.2%), with a majority of psoriasis or ankylosing spondylitis (n=13/21, 61.9%). At diagnosis, the level of pain was 5.71 ± 3.24. Eleven patients (42.3%) had blood inflammation. Bone lesions were reviewed thanks to whole body MRI: four patients had isolated axial involvement (10.0%), nine had peripheral involvement (22.5%) and 27 patients harbored both types of lesions (67.5%). The mean number of lesions was 6.65 ± 4.23. After 12 months of follow-up, all parameters regarding disease’s activity decreased (pain, blood inflammation, number of lesions). Regarding extra-osseous symptoms, fever occurred in seven patients (17.1%). Twenty-four patients had skin manifestations (58.5%) with palmpalantar lesions (n=3, 12.5%), acne (n=6, 25.0%), psoriasis (n=5, 20.8%) and aphthous (n=10, 41.7%). Four patients (9.76%) had gastro-intestinal symptoms and seven (17.1%) had enthesitis. One patient had uveitis. Almost all patients received NSAIDs (n=39/41, 95.1%) and half of the cohort were treated with bisphosphonates (n=21/41, 51.2%). Nine patients (22.0%) received biologics with a majority of TNF inhibitors. All patients that received TNF inhibitors had either a cutaneous involvement or digestive symptoms or enthesitis (Figure 1).

Conclusion: Extra-osseous manifestations have to be carefully searched in CRMO, especially in the presence of familial history of inflammatory diseases. While the severity of bone involvement can lead to use bisphosphonates, the introduction of biological treatment rely on extra-osseous symptoms. These conclusions are drawn on a retrospective study and need to be confirmed in larger cohort.

REFERENCES:

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PIMS THROUGH THE WAVES OF COVID 19: DATA FROM THE JIR COHORT

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Objectives: To describe the clinical, immunological and genetic aspects in 89 patients.

Methods: A prospective registry collecting demographic, clinical and paraclinical data on patients with pediatric inflammatory diseases. Two groups were distinguished: from March 2020 to July 2020 for patients in the first wave, from July 2020 to June 2021 for patients in the second and 3rd waves. These two groups were compared using a Fischer test for categorical data and a Mann-Whitney test for quantitative data.

Results: 138 patients meeting the PIMS criteria were included (64 patients in the 1st wave, 72 patients after). Patients had less frequent myocarditis (51 patients in wave 1 vs. 36 patients after, p=0.0003) and respiratory distress (34 patients vs 10 patients, p<0.0001). Corticosteroids were used more frequently in the second wave (32 patients in wave 1 vs. 67 patients after PIMS, p=0.0001). Intravenous immunoglobulins were used as much over the waves (58 patients in wave 1 vs 69 patients after, p<0.5). Antibiotics were less used since the second wave (53 patients received antibiotics before July 2020 vs 11 after, p<0.0001). During the duration of hospitalization decreased significantly (p<0.0001) with a median duration of 9 days during the first wave (interquartile range, 7-12) and 7 days (interquartile range, 5-10) after the first wave.

Conclusion: This is a decrease in the number of complications of PIMS, particularly cardiac and respiratory complications, and a decrease in the length of hospitalization over time. The treatment of PIMS has also evolved, with a clear increase in the use of corticosteroids and a decrease in the use of antibiotics.

REFERENCES:

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