Conclusion: Hip involvement is a major and highly prevalent issue in our JIA patients.

Disclosure of Interests: None declared

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AB0731 CERVICAL SPINE INVOLVEMENT IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in children. In contrast to adult rheumatoid arthritis, where numerous studies have shown a high prevalence of involvement of the cervical spine, few studies have been published examining this entity in JIA.

Objectives: We aimed to analyze clinical and radiological findings of cervical spine involvement in patients with JIA.

Methods: A retrospective study including 35 patients was conducted between 2010 and 2019. The patients enrolled met the ILAR criteria for the diagnosis of JIA. Clinical, biological, and radiological data were collected. Patients had a radiological evaluation that included cervical spine x-rays in antero-posterior, lateral and lateral views with flexion.

Results: Thirty-five patients were enrolled. The mean age of the disease onset was 9 years [3-15]. The mean age of the patients at the time of the study was 37.8 years [17-69]. The mean duration of the disease was 27 years [2-56]. These patients were assigned to discrete JIA categories: rheumatoid factor positive polyarthritis (43.5%), rheumatoid factor negative polyarthritis (21.7%), enthesitis-related arthritis (17.4%), oligoarthritis (13%) and psoriatic arthritis (4.4%). Sixteen patients (45%) reported neck pain. Cervical spine involvement occurred on average 7 years [0-13] after the JIA onset. Cervical spine radiographs showed anterior atlantoaxial subluxation (> 5mm) in 8 patients. Magnetic-resonance imaging was performed in 9 patients that had abnormal neurological examination showing a pannus formation of C1-C2 junction (3 cases), a basilar invagination (4 cases) and erosions of the odontoid process (2 cases). A cervical collar has been used for immobilization in patients with significant cervical spine damage. A C1-C2 arthrodesis was proposed to 4 patients.

Conclusion: These findings suggest that the presence of cervical involvement in JIA patients is frequent. Radiologic assessment of cervical spine should be systematically performed for early detection and to prevent its complications.

Disclosure of Interests: None declared

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AB0732 JUVENILE IDIOPATHIC ARTHRITIS IN ADULTHOOD

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease of unknown aetiology in childhood. Approximately 40- 60% of patients with juvenile idiopathic arthritis (JIA) have continuous or recurrent disease activity extending into adulthood.

Objectives: The aim of this study was to evaluate the clinical, biological and radiological course of JIA in adulthood.

Methods: A retrospective study including 35 patients was conducted between 2010 and 2019. The patients enrolled met the ILAR criteria for the diagnosis of JIA. Patients with JIA older than 20 years were included. Data regarding sociodemographic features, Physical growth, disease activity, biological and radiological parameters were analysed.

Results: Thirty one patients were recruited. The sex ratio was 0.53. The mean age at the time of the study was 38.9 years [20-69]. The mean age of the disease onset was 9 years [3-16]. These patients were assigned to discrete JIA categories: rheumatoid factor positive polyarthritis (47.7%), rheumatoid factor negative polyarthritis (23.9%), oligoarthritis (14.2%), enthesitis-related arthritis (9.5%), and psoriatic arthritis (4.7%). A failure to thrive was seen in 33.3% of patients. Overweight and obesity were found in 38% and 19% of patients, respectively. Biological inflammatory syndrome was noted in 52% of patients, and 65.2% had active disease. Hip involvement was noted in 43.5% of patients and 17.4% of them had a total hip replacement. Sixteen patients had neck pain and the imaging showed an atlanto-axial dislocation in 50% of them. Radiographs showed a joint destruction in 60.9% of patients and a wrist arthritis was the most frequent involvement.

Conclusion: Most of our included patients maintain active disease and have functional impairment in adulthood.

Disclosure of Interests: None declared

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AB0733 DADA2 VASCULITIS PRESENTING AS POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES): AN ATYPICAL PRESENTATION

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Background: Deficiency of Adenosine Aminase deficiency 2 is monogenic disease presenting with multisystem involvement of vasculitis, Stroke¹, hematological manifestations. We hereby present a young child who presented with PRES initially and later diagnosed as DADA2. Only one case of DADA2 presenting as PRES is reported so far². This case highlights the atypical presentation of DADA2.

Objectives: A case report to highlight the rare presentation of DADA2 vasculitis.

Methods: 8 year old developmentally normal male child, born out of consanguineous marriage presented with fever, abdominal pain, seizures 1 year back. Ultrasound of abdomen revealed mesenteric lymphadenitis and MRI Brain was suggestive of Meningoencephalitis. He was managed with antivirals and antibiotics then.

6 months later, he had fever, skin rash, pain abdomen, status epilepticus, hypertension followed by gangrene of fingers and toes. APLA, ANA, ANCA workup was negative. Complete Hemogram was normal. CRP was 130mg/dl. CT Abdomen was normal and no evidence of aneurysms. Renal Doppler Ultrasound was normal. CT upper limb showed left radial and ulnar artery significant narrowing with patchy occlusion. MRI Brain showed bilateral fronto parietal, occipital, putamen, left insula and subcortical and cortical T2W FLAIR hyperintensity without diffusion restriction suggestive of PRES. He was diagnosed as Childhood Polyarteritis Nodosa and treated with cyclophosphamide. Genetic Analysis of ADA2 (CECR1) gene mutation by Sanger Sequencing of exons 2 to 10 showed a known variant (rs202134424) in ADA2 gene.

Figure 1. Gangrene of left 1-5th digits and right 4th and 5th digit resorption

Figure 2. T2W FLAIR image showing bilateral fronto parietal, occipital, putamen, left insula and subcortical and cortical hyperintensity without diffusion restriction suggestive of PRES.

Results: He was started on Infliximab and is on follow up. No further events noted so far. Rehabilitation of left upper hand was done.

Conclusion: DADA2 can present with varied CNS manifestations like infarct, hemorrhage, aneurysms and PRES. High index of suspicion and early recognition can help in maintaining vascular integrity.

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