Methods: A retrospective single-center hospital-based cohort study was performed to analyze cancer occurrence among JIA patients treated with biologic agents over the observation period between January 2004 and January 2018. The medical patient records were reviewed to obtain information about the clinical follow-up. As reference data for direct standardization; age, sex and calendar-year-specific incidence rates from Turkish cancer registry were used. The standardized incidence ratio (SIR, ratio of cancers observed to expected) was generated, with 95% confidence intervals.

Results: The study sample consisted of 504 JIA patients, who had been started their first biologic treatment between 2004 and 2018. Mean age was 17.1 years (SD 5.6) with 56% of female proportion. The mean disease duration was 10.3±5.1 years. Median time from baseline to start of the first biological was 17.5 (IQR:43) months. Mean age of initiation of biological treatment was 9.8 ± 4.2 years. Etanercept was the most commonly preferred drug to initiate an anti-TNF therapy first-line biologic treatment (n=361,72%). 172 (34.1%) patients in the cohort required a switch to a second biological agent. Main reason for switching to another biologic agent was due to lack of response (16.6%). Median duration of biologic use was 35 (IQR:41) months. One cancer occurred within observation period, compared with 0.095 expected (SIR:10.53, 95% CI 0.526 to 51.91). The patient was 18-year-old male, who had previously received etanercept and tocilizumab up until diagnosis of the hematological malignancy.

Conclusion: In our JIA cohort, patients treated with biologic agents appeared to have an increased rate of incident malignancy compared to children of the same sex and age group in the general population in Turkey. However, before mentioning a clear causal relationship, other potential contributing factors such as inflammatory process of the underlying disease itself and the use of concomitant immunosuppressants should be taken into consideration. Additional long-term studies with larger populations are needed to be able to draw definite conclusions.

REFERENCES

Disclosure of Interests: None declared


SAT0504 IGG4 RELATED DISEASE IN CHILDREN: A SINGLE CENTRE EXPERIENCE FROM NORTH-WEST INDIA

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Background: Immunoglobulin G4 related disease (IgG4RD) is a multisystemic disorder characterized by elevated serum IgG4 levels and infiltration of IgG4 positive plasma cells accompanied by fibrosis. It is mostly considered a disease of adults and elderly people. There is paucity of literature on pediatric IgG4RD. A recent systematic review has found only 25 pediatric cases of IgG4RD

Objectives: To report broad patterns of organ involvement in IgG4RD in children and also to create awareness among treating pediatrics about this new entity.

Methods: The study is based on a review of the hospital records of children with IgG4RD at tertiary centre from North-West India. Diagnosis was based on clinical features, IgG4 levels and characteristic histopathology findings.

Results: Six patients had IgG4RD. Pt-1: 10-year-old boy presented with fever for 3 months and significant hepatomegaly. Investigations showed anemia, thrombocytopenia, elevated erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP). Ultrasonography (USG) revealed a hepatic mass (10x5x5 cm) that was confirmed on computed tomography. Liver biopsy showed increased plasma cells (>50 IgG4 positive plasma cells/HPF) and storiform fibrosis suggestive of IgG4 related hepatic mass. Serum IgG4 level was 420 mg/dl (N: 6-28).

Pt-2: 12-year-old girl presented with an abdominal lump. Upper gastrointestinal endoscopy showed an intragastric mass with exophytic component. Histopathology of the abdominal mass was consistent with IgG4RD. Serum IgG4 level was >170 mg/dl (N: 6-28).

Pt-3: 21-year-old male symptomatic since age of 14 years with recurrent erythematous swellings over dorsum of the left hand, forearm and chest. Investigations showed anemia, elevated ESR, CRP, and hypergammaglobulinemia. IgG4 levels were 211 mg/dl (N: 7-26).

Histopathology of this unrecognized disease in pediatrics. Pediatr Rheumatol Online J. 2016;14(1):18

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Disclosure of Interests: None declared


SAT0505 LONG-TERM FOLLOW-UP IN KAWASAKI SYNDROME: EVIDENCE FROM RETROSPECTIVE MONOCENTRIC DATA IN REAL LIFE

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Background: Kawasaki syndrome (KS) is a severe medium size vasculitis with predilection for coronary arteries, mainly affecting young children. The identification of risk factors for coronary artery lesions (CAL) is the aim of the scientific society in order to early detect patients who need a more aggressive therapy.

Objectives: To report clinical data of a monocentric cohort of children with KS over a long-term follow-up.

Methods: A monocentric retrospective study between Feb 1982 and Aug 2018, involving children with KS diagnosis followed at the Rheumatology and Immunology Unit of Meyer Children’s Hospital. According to cardiac involvement, patients (pts) have been stratified in subject with absence of CAL (no-CAL) and presence of CAL. This latter has been divided in coronary ectasia (CE) and presence of coronary aneurysm (CAN). Data analysis was conducted performing a descriptive analysis and assessing risk factors for CAL with analysis of variance (Tukey, LSD and Bonferroni test) and correlation test (Spearman rho test). Each pt, regardless coronary involvement, underwent a scheduled cardiological follow-up after 6-8 weeks (6 months, 1 year and ergometric test at 8 years old). Pts with CAL received additionally cardiac evaluations on clinical need. Pts with a follow-up of less than 1 year were excluded from follow-up analyses.

Results: Data from 361 pts (219 M, 142 F, aged 1 month-24 years and 5 months) have been reported. Median age of disease onset was 2 years and 1 month; 90% of pts developed the disease < 5 yrs. Disease onset at < 6 months was in 32 patients (8.9%), at < 1 year in 94 (26%). Full clinical data were available in 267 pts. Beside fever,
We set up an observational study of patients with KD in a cohort of patients with KD diagnosis from northern Spain. A) To describe demographic, clinical and analytical features of patients with predilection for coronary arteries. Due to the high probability of aneurysms were used for diagnosis. B) To study the long-term follow-up assesses, in real life, the benign course of KD in our cohort was 0.37 cases/100,000/year, 8-10 times lower than other national series. However, mean age at onset, presence of previous viral infections and aneurysms development, were similar to countries with higher prevalence.

Conclusion: Although the incidence of KD in our population is lower than other territories, it is still the most frequent cause of acquired heart disease in childhood. Early recognition and treatment with IVIG improve prognosis leading to a decrease in the rate of long-term cardiovascular outcomes.

<table>
<thead>
<tr>
<th>N pts</th>
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<th>CAn</th>
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<tbody>
<tr>
<td></td>
<td>288</td>
<td>58</td>
<td>15</td>
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<tr>
<td>Median age at disease onset</td>
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<td>2y 5m</td>
<td>5m</td>
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<tr>
<td>Median duration of fever</td>
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<td>Average value of CRP (mg/dl)</td>
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<td>Day of first dose of IVIG</td>
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<td>11d</td>
</tr>
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Table 1

Table 2

REFERENCES