DELAY IN DIAGNOSIS OF KAWASAKI DISEASE IS THE COMMONEST PROXIMATE REASON FOR DEVELOPMENT OF GIANT CORONARY ARTERY ANEURYSMS—OUR EXPERIENCE AT CHANDIGARH, NORTH INDIA

KUMAR RAKESH1, Ankur Jindal1, Anuj Gupta1, Deepi Suri1, Manphool Singh2, Surjit Singh1.1 Post Graduate Institute of Medical Education and Research, Chandigarh, Allergy Immunology Unit, Department of Pediatrics, Chandigarh, India; 2Post Graduate Institute of Medical Education and Research, Chandigarh, Department of Radiodiagnosis and Imaging, Chandigarh, India

Background: Long-term effects of Kawasaki disease (KD) depend primarily on development of coronary artery abnormalities. Giant coronary aneurysm (GCA) is one of the most severe sequelae in KD. Regression of giant aneurysm is rare. Data from Indian subcontinent is limited. Herein, we review patients with KD who had GCA.

Objectives: To describe the profile of patients with KD who developed GCA from a cohort of KD patients at Advanced Pediatric Centre, Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh.

Methods: Records of all children diagnosed to have KD during 1994-2017 were analysed. Out of the 680 patients with KD, clinical details of 17 (2.5%) children with GCA were retrieved.

Results: Diagnosis of GCA was based on coronary artery diameter >8 mm or >10 z score. Six of 17 children (boys 13; girls 4) with GCA had incomplete KD. Diagnosis of KD was made at a mean of 17.2±12.2 days of fever. Eight (47%) children were <1 year. Median age of diagnosis was 18 months (range 1.5 months-12 years). Left anterior descending artery (LAD) coronary artery was affected in 82% followed by right coronary artery (RCA) in 59%. Multiple GCA >1 were seen in 65% patients. All patients had received first line therapy as IVIg. Median day of IVIg administration of IVIg appears to be the commonest proximate cause of development of GCA.

REFERENCES

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LUPUS LIKE SYNDROME ASSOCIATED WITH INFlixIMAB TREATMENT IN CHILDHOOD: CASE SERIES OF TWO PATIENT

divyaj kurthela1, Ferhat Demir2, Betül Sözeri2, 1University of Health Sciences, Umraniye Training and Research Hospital, Physical medicine and rehabilitation, Istanbul, Turkey; 2University of Health Sciences, Umraniye Training and Research Hospital, Pediatric Rheumatology, Istanbul, Turkey

Background: Drug-induced lupus (DIL) caused by medication is a form of lupus erythematosus. When the medication is stopped, disease symptoms generally disappear in days or months. Infliximab is an anti-TNF chimeric antibody widely used and approved for the treatment of many diseases. TNF-α antagonist induced lupus-like syndrome (TAILS) is among the uncommon side effects of infliximab. TAILS is a clinical syndrome with features similar to SLE, but with some differences in laboratory and clinical findings.

Objectives: We have presented our experience with lupus like syndrome emerged after infliximab treatment in two children.

Methods: We evaluated the clinical properties of 2 patients who were monitored with diagnoses of juvenile idiopathic arthritis and uveitis symptoms and developed TAILS after infliximab treatment.

Results: First patient applied to our pediatric rheumatology outpatient clinic with a complaint of swelling in the left knee continuing for 1 month. During the physical examination, we found arthritis on the left knee. During the eye examination, it was discovered that he had bilateral pars planitas. Anti-nuclear antibodies were found positive at 1/320 titration, and anti-dsDNA was negative. Following JIA and accompanying uveitis diagnoses, methylprednisolone and methotrexate treatment was started for the patient. After treatment, the patient whose joint and eye symptoms started to regress, was subjected to lesser dose of methylprednisolone, but the uveitis attack of the patient started to occur again. We added infliximab treatment to its treatment that was resistant to uveitis. At the 6-month of infliximab treatment, there was no uveitis attack observed. During remission, after 6. dose of infliximab treatment, there were complaints of tiredness and polyarthralgia. It was observed that the patient had arthritis in several joints. After the re-studied laboratory tests, there was anti-dsDNA antibody positivity. We thought the patient developed TAILS, and stopped the infliximab treatment. After stopping the treatment, complaints of tiredness and polyarthralgia, as well as arthritis symptoms were improved, and anti-dsDNA antibody levels returned back to normal. The second, 15-year-old patient was being monitored by pediatric rheumatology outpatient clinic with oligoarticular JIA diagnosis since the age of 1. The patient had also uveitis on the right eye since the age of 6. During methylprednisolone, methotrexate, and azathioprine treatment, there were uveitis attacks that developed at different periods. We started infliximab treatment for uveitis of the patient. After 2. dose of infliximab, the patient started to complain about migraine style headaches that occurred once or twice a week. In redone laboratory tests, ANA (at 1/320 titration), anti-cardiolipin antibody, and anti-beta2 glyco-protein antibody were found positive. There was also low levels of C3 and C4 detected. The infliximab treatment was stopped for the patient who was diagnosed with TAILS. After stopping treatment, the headache complaints regressed, and auto-antibody positivity's turned back to normal.

Conclusion: Although the development of ANA anti-dsDNA antibodies during infliximab therapy is common, TAILS is rare, especially in the pediatric population. In patients develop SLE findings under infliximab infusion, TAILS must be considered and the drug should be discontinued.

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THE UNUSUAL ADVERSE EVENTS INCLUDING MULTIPLE SCLEROSIS DEVELOPED UNDER ABATACEPT THERAPY IN A PATIENT WITH JIA

Alia Latypova, Anna Shapirovalenko, Irina Nikishina. V.A.Nasonova Research Institute of Rheumatology, Pediatric, Moscow, Russian Federation

Background: Biological therapy in patients with pediatric rheumatology disease may be associated with adverse events up to demyelinative lesion of CNS including multiple sclerosis (MS). It is a rare condition in the pediatric practice.

Objectives: to describe the clinical case of newly diagnosed psoriasis and subsequently developing of multiple sclerosis in pediatric patient revealed in JIA who has been followed-up in Pediatric department of V. A. Nasonova Research Institute of Rheumatology for 8 years.

Methods: Description of clinical case of 13 y.o. male Caucasian pts presented with psoriasis variant of juvenile idiopathic arthritis (JIA) since 2006.

Results: The first symptoms (at the age of 1.5) were polyarthrits with fever, morning stiffness and inflammatory activity in laboratory tests in chronological link of DTP – re-vaccination. The patient admitted to the regional hospital where JIA was diagnosed and he received traditional therapy including NSAIDS, methotrexate, numerous of intrarticular injections, methylprednisolone pulse therapy with initial response. By the 2012 the child deteriorated with progressive polyartthritis, deformation of wrist joints, bilateral camptodactyla with progressive destruction on X-ray. Under the administration of abatacept therapy in our clinic in 2012 the significant improvement (70-90% ACR response) was achieved. 2 years later the plaque psoriasis developed. The cutaneous changes were impaired by the regular local application of GC so infusions of abatacept were continued. In 4 years of regular therapy (in the age of 13) sudden neurological symptoms such as headache, loss of sensitivity, ataxia, visual field defect appeared. He was admitted to neurological clinic. Multiple sclerosis was diagnosed based on the presenting signs and symptoms supporting cranial MRI – multiple focal areas of demyelination.

Conclusion: We presented a patient with JIA who developed two different immunological related adverse events under long-term abatacept using:
AB1006
THE CLINICAL SPECTRUM OF TWO HETEROZYGOUS MUTATIONS IN THE MVK GENE CONFIRMING HYPERIMMUNOGLOBULIN D SYNDROME
Dragana Lazevic1, Jelena Vojnovic2, 1Clinical Center Niš, Clinic of Pediatrics, Niš, Serbia; 2University of Niš, Faculty of Medicine, Niš, Serbia

Background: Autoinflammatory syndromes represent the wide spectrum of rare diseases (associated with genetic disturbances) characterized by the presence of chronic or recurrent systemic inflammation with diverse clinical presentation.

Objectives: We report a case report of child with confirmed two heterozygous mutations in the MVK gene with clinical features associated hyperimmunoglobulin D syndrome.

Results: We present girl with clinical symptoms starting just after birth when in the first days of life she was examined at gastroenterology department in Belgrade due to elevated parameters of inflammation, anemia, direct hyperbilirubinemia, abdominal bloating and hepatosplenomegaly. Breastfeeding was discontinued due to galactosuria and lactose free diet was advised. Due to the maintenance of hepatomegaly, a detailed hematological, virological and gastroenterological diagnostic testing was performed. Since liver biopsy has showed portal and lobular hepatitis with cholestasis without fibrosis she was on ursosalk treatment with partial response. At five months of age she started to have recurrent episodes of fever every month for few days with no associated infection, but always followed with digestive symptomatology (abdominal pain and abdominal flatulence) and elevation of inflammatory parameters. Despite antibiotics, episodes of fever continued to repeat twice monthly with accompanying occurrence of hypersalivation, small ulcers in the mouth, skin rash, cervical lymphadenopathy, hepatosplenomegaly, abdominal pain and abdominal bloating with elevated inflammatory markers. One year later she was admitted for the first time at our department and detailed differential diagnostic testing was performed. Clinical spectrum of presenting symptoms with specific phenotypic aspect (hypertelorismus and frontal bossing) was without IgG increase. Genetic testing have revealed presence of two heterozygous mutations (heterozygous variant c.790del p.(Leu264Serfs*2)) and heterozygous variant c.1129G>A p. (Val377Ile) in the mevalonate kinase deficiency gene (MVK gene) confirming hyperimmunoglobulin D syndrome.

Conclusion: Diverse clinical manifestations of some patients with autoinflammatory diseases can provoke differential diagnostic and treatment dilemmas. Genetic testing is of great importance for establishing the final diagnosis and starting accurate treatment in order to prevent potential serious complications seen in these patients.

Disclosure of Interests: None declared


AB1007
PREVALENCE AND CHARACTERISTICS OF TEMPOROMANDIBULAR JOINT (TMJ) INVOLVEMENT IN A COHORT OF YOUNG ADULT PATIENTS WITH DIAGNOSIS OF JUVENILE IDIOPATHIC ARTHRITIS (JIA) AND NON-JIA CHRONIC INFLAMMATORY ARTHROPATHIES
Adriano Lercara1, Gloria Crepaldi2, Francesco Liciardi3, Marco Davico3, Stefano Cintio4, Sarah Marouen5, Enrica Vandelli4, Yves-Marie Pers4, Claudia Lomater1

Background: Mandibular joint (TMJ) is involved in about 50% of JIA cases, often bilateral and asymptomatic in up to 71% of cases. Adult patients with JIA have been shown to have, compared to healthy individuals, higher rate of dysfunction and anatomical abnormalities. Clinical examination has been shown to have high specificity but low sensitivity in revealing TMJ inflammation. To date MRI is the gold standard to assess TMJ involvement.

Objectives: To investigate the prevalence of TMJ involvement in young adults with JIA and young adults with non-JIA inflammatory rheumatisms.

Methods: Patients were recruited prospectively in 2 clinical centers. Inclusion criteria were: patients <35 years diagnosed with JIA who had undergone transition from the pediatric to the adult rheumatologist, and patients diagnosed with non-JIA inflammatory arthropathies. All patients were assessed for joint count, clinical examination for TMJs (tenderness to palpation, swelling, signs of damage such as joint crepitations, lateral deviation, retrougnathia and decreased mouth opening), evaluation of global disease activity with composite indexes and underwent MRI of the TMJs to detect inflammation (bone marrow edema, effusion, synovial thickening) or damage (condylar flattening, erosions, disk abnormalities); MRIs with either inflammation or damage were considered pathological. Demographic and clinical characteristics were described using frequency and median and interquartile range (IQR), depending on the distribution of the variable.

Results: 19 patients were included in the JIA group and 8 patients in the non-JIA group. Patients’ demographic and disease characteristics were reported in Table 1.

MRI results are collected in Table 2.

There are no statistically significant differences between groups for the presence of inflammation on MRI, while damage (in particular, disk abnormalities) is more likely in JIA rather than non-JIA patients (p = 0.02).

Conclusion: It was found that it is more likely to find damage on MRI in patients of both groups rather than inflammation. Both groups show the same frequency of TMJ involvement, suggesting that TMJ involvement must be sought by the adult rheumatologist as it not only affects JIA patients, but also those with adult-onset inflammatory arthropathies.

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