AB1003

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

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Background: Long-term effects of Kawasaki disease (KD) depend primarily on development of coronary artery abnormalities. Giant coronary aneurysm (GCA) is the 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, Post-graduate 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 $\geq +10$ 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 (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 was 15.5 days. Twelve had received additional therapy with infliximab. Thromboses developed in 4 (23.5%) and most common coronary affected was LAD. All patients were started on anticoagulation therapy and there were no significant complications related to anticoagulation.

Conclusion: Results of this study suggest that GCA develop more commonly in infants and young children. Delay in diagnosis and consequent administration of IVIg appears to be the commonest proximate cause of development of GCA.

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AB1004

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

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Background: Drug-induced lupus (DIL) caused by medication is a form of lupus erythematosus. When the medication is stopped, disease symptoms generally disappears 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 emerge 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 planitis. 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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AB1005

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

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Background: Biological therapy in patients with pediatric rheumatology disease may be associated with adverse events up to demyelinating 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 polyarthritis variant of juvenile idiopathic arthritis (JIA) since 2006.

Results: The first symptoms (at the age of 1,5) were polyarthritis 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 intraarticular injections, methylprednisolone pulse therapy with initial response. By the 2012 the child deteriorated with progressive polyarthritis, deformation of wrist joints, bilateral camptodactyly 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 4th 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: