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 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 (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 IV Ig. Median day of IV Ig emergence after infliximab diagnosis of KD was 15.5 years. 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 IV Ig appears to be the commonest proximate cause of development of GCA.

REFERENCES

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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. Nasonov 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 polyarthritics 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 polyarthritics, deformation of wrist joints, bilateral camptodactylia 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: