

Is there any difference in clinical features of Turkish children with Kawasaki disease?

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Kawasaki disease (KD) has replaced acute rheumatic fever as the most common cause of acquired heart disease in children in developed countries and occurs in 9% of childhood vasculitis in our country (1). There are few studies concerning the characteristic features of Turkish children diagnosed with KD (1-4). We aimed to identify the clinical characteristics and potential risk factors for coronary artery abnormalities (CAAs) in Turkish children diagnosed with KD.

We evaluated clinical, demographic and laboratory data of 49 patients diagnosed with KD in our Pediatric Cardiology clinic between January 2005 and May 2013, retrospectively. The median age was 27 months and male to female ratio was 1.08 eighteen cases (36.7%) had incomplete presentation. 43 cases were admitted at the acute phase of the disease, they were treated with intravenous immunoglobulin (IVIG) and unresponsiveness to the first dose of IVIG was observed in five cases (11.6%). CAAs were detected in 13 cases (26.5%) by echocardiography. Most cases had mild (6 cases, 46.2%) or moderate (5 cases, 38.5%) size aneurysms and the giant aneurysm occurred in two cases (15.4%). CAAs were detected at the median of 9th day (between 5th and 18th days). CAAs were detected in seven of 13 cases (54%) within the first ten days of febrile illness. CAAs were seen more frequently in patients with incomplete presentation (33.3%) than patients with complete presentation (25%) ($p=0.023$). The resolution of CAAs was observed in 11 patients within six months, but the giant CAAs didn't resolve in two cases during follow up period. One of them was a 4-month old infant and he had presented with multiple giant coronary aneurysms and massive pericardial effusion. Despite intensive anticoagulation, intramural coronary thrombosis developed in this case and he suddenly died because of myocardial ischemia at the 51th day. The giant aneurysm was partially regressed in the other case within three years. We identified that the longer time interval from disease onset to treatment ($p=0.005$), higher platelet

counts ($p=0.04$) and male gender ($p=0.02$) were the risk factors for coronary abnormalities.

In this study, we noticed that a small number of cases with KD was under one year old at admission and there was balanced in gender distribution. These two features were the main clinical differences in our study group. Like our study, Gülhan et al. (2) were noticed that there was no gender predominance in Turkish children with KD, and they also mentioned that the earlier occurrence of desquamation and cardiovascular disease than expected by conventional teaching. Likewise previous Turkish patient series (1-4), the most common clinical finding was changes in the lips and oral cavity in our study. We also observed that most of our patients were irritable at the time of the diagnosis.

The other important feature of the Turkish children with KD was a relatively high rate of coronary artery abnormalities which occurs in up to 44.2% of patient series (1-5). Approximately, 10-33% of cases with KD have incomplete presentation and they have also had an increasing risk for development of CAA (1-5). The reported risk factors for CAA are male gender, race, younger patient age, lower albumin and hemoglobin level, higher platelet count. Similar to previous study, longer interval from disease onset to treatment, higher platelet count and male gender were identified as risk factors in our study.

Approximately 5-25% of children with KD doesn't respond to a first dose of immunoglobulin and have a risk of CAAs (5). The risk factors for unresponsiveness to steroid treatment are lower albumin and hemoglobin level, and higher platelet count (5). A half of our cases who needed the second dose of IVIG treatment had also at least three risk factors.

Anticoagulation is another important issue for children with giant aneurysm. In our series, myocardial ischemia related to intracoronary thrombosis occurred in only one case with giant aneurysm. Tissue plasminogen activator (t-PA), fractionated or

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unfractionated heparin can use in case of giant CAAs and associated intracoronary thrombosis. Anticoagulation had been given rapidly in our cases and provided a rapid resolution of the coronary thrombosis. Therefore, to prevent ischemic complication, the close monitoring of coronary arteries by echocardiography is mandatory in patients with giant aneurysm and the management of intracoronary thrombosis may be possible by pharmacologic treatment.

Prognosis of coronary abnormality depends on the size of the aneurysms (1-5). Kayıran et al. (3) reported that coronary lesions resolved in all cases within eight months, in accordance with our study. Erdoğan et al. (5) evaluated 22 coronary aneurysms in patients with incomplete form of KD and reported that the half of these abnormalities disappeared after median of 2.8 years of follow up.

Based on our study population and literature data, there are some differences in the demographic and clinical features of Turkish children with KD. We have noticed that there were a balanced gender distribution and the small number of case had diagnosed under one year old. The earlier occurrence of desquamation, and the increased number of cases with incomplete clinical findings were the other findings in Turkish children with KD. The rate of CAAs was high in our population and developed in most of cases at the first ten days of illness. We need multi-center studies which reflecting clinical characteristics of KD in Turkish children.

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