Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

EXPERT: Jakub Podolec, MD PhD, cardiolgist

Affiliation: *Department of Hemodynamics and Angiocardiography, Cardiology Institute, Collegium Medicum, Jagiellonian University at the John Paul II Hospital, Krakow, Poland*

CASE SUMMARY

The 18 year old patient was admitted to the Hospital of Lithuanian University of Health Sciences Kaunas Clinics on 3 Jan 2012 due to the: fewer (38°C), nausea, vomiting, headache, stomach ache. The patient was pale, sluggish, dry. There was bilateral nonexudative conjunctivitis. Erythema of the lips and oral mucosa. Lung auscultation revealed fine wet rales on both sides; respiratory rate was 20 times per minute, SaO2 97%. Heart rate was122 bpm, blood pressure 96/48mmHg, heart auscultation: 3/6 degree systolic murmur at the apex of the heart. Painful stomach palpation. No periferial oedema. Resting electrocardiography showed sinus rhythm and early repolarization but no other abnormality. Blood test revealed high rate of inflammatory markers, mild insufficiency of kidney and liver were found. The findings were differentiated with measles, rubella, infectious mononucleosis, Yersinia enterocolitica infection, bacterial infection - staphylococcal and streptococcal infections causing staphylococcal scalded skin syndrome, toxic shock syndrome and scarlet fever, toxic epidermal necrolysis.

LITERATURE REVIEW

Kawaski disease (KD) is an acute vasculitis typically occuring in young children and was first described by Tomisaku Kawasaki in 1967 [1, 2]. Patients with KD may develop thrombosis or stenotic lesions associated with the aneurysms and are at risk of myocardial infarction, sudden death, and congestive heart failure [2]. Around 4,000 new cases are diagnosed in the USA each year [3]. In Japan the incidence of the disease is 10 times higher. 10,000 new cases of KD are recognized each year [4]. If untreated 25% of patients with KD can develop intra-

coronary aneurysms, while in patients treated with intravenous immunoglobulin the prevalence of aneurysms drops to approximately 5% [5]. Infliximab and steroids are currently the two agents that have been most studied [6]. In some centers clopidogrel is also added to aspirin therapy. The current AHA guidelines suggest discontinuation of anti-platelet therapy when the aneurysm regresses. In patients with aneurysms of ≥8 mm risk for thrombosis is high, systemic anticoagulation with warfarin with an INR between 2.0-2.5 has been associated with improved survival in small series of KD patients from Canada and Japan [7]. If patients present ischaemic symptoms or its equivalents percutaneous coronary intervention in many cases may be complicated due to highly calcified atherosclerotic plaques. In such situation rotablation procedures may be useful [8].

EXPERT'S OPINION

Based on todays knowledge but limited experience, medication and diagnostic methods applied in this patient have been administrated correctly. Although, in this patient with KD typical ischemic symptoms are not present and may not appear at all I would recommend to follow up the patient without intervention at this time, but analyzing ischaemia of the heart with one of the imaging techniques (ex. MRI, PET-CT) in the nearest future. Due to great lumen differences in the aneurysmatic vessels coronary artery bypass grafting may be the treatment method of choice if needed. Aneurysms due to Kawasaki disease may cause acute coronary syndromes in adult patients. In most cases resulting from thrombus formation and artery occlusion [9].

CONCLUSION

The patient was currently qualified for close follow-up. Coronary arteries should be evaluated with the use of MSCT. If symptoms of ischemia occurs CABG is recommended treatment.

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John Paul II Hospital in Kraków
Jagiellonian University, Institute of Cardiology
80 Prądnicka Str., 31-202 Kraków;
tel. +48 (12) 614 33 99; 614 34 88; fax. +48 (12) 614 34 88
e-mail: rarediseases@szpitaljp2.krakow.pl
www.crcd.eu

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John Paul II Hospital in Kraków
Jagiellonian University, Institute of Cardiology
80 Prądnicka Str., 31-202 Kraków;
tel. +48 (12) 614 33 99; 614 34 88; fax. +48 (12) 614 34 88
e-mail: rarediseases@szpitaljp2.krakow.pl
www.crcd.eu