Medical Expertise*

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

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CASE SUMMARY

This a case of a 18 year old patient who was admitted to the Hospital of Lithuanian University of Health Sciences Kaunas Clinics on 3 Jan 2012 due to fever, and gastrointestinal symptoms. Additionally he had bilateral nonexudative conjunctivitis, erythema of the lips and oral mucosa. Lung auscultation revealed fine wet rales on both sides and heart auscultation revealed 3/6 degree systolic murmur at the apex of the heart. Blood test revealed increased inflammatory markers, mild insufficiency of kidneys and liver. Echocardiography showed reduced left ventricular ejection fraction with hypokinesis of the posterior wall of the left ventricle, moderate regurgitation of the tricuspid valve and mild regurgitation of the mitral valve. Computed tomography revealed aneurysms of the left main coronary artery with thrombus partially obstructing the lumen, aneurysm of the proximal left anterior descending coronary artery, and proximal part of the right coronary artery. These findings led to the diagnosis of Kawasaki disease. The treatment included intravenous immunoglobulins, aspirin, anticoagulation, steroids and diuretics. At 12 months follow-up the patient was completely asymptomatic and echocardiography was normal. Computed tomography showed no progression of the aneurysmal lesions

LITERATURE REVIEW

Kawasaki disease first described by Tomisaku Kawasaki in 1967 is an acute febrile multisystem vasculitic syndrome involving small and medium-sized arteries, particularly the coronary arteries. It occurs mainly at childhood usually in the first 5 years however adult cases have also been described. The diagnosis is based on clinical features including prolonged fever, cutaneous and mucosal lesions (multiform rash, stomatitis, conjunctivitis, erythema of the hands and feet, with late peeling of the digits), and lymphadenopaty. The acute symptoms

are self limiting and subside in a few weeks time however coronary lesions and myocardial damage can progress over time. Coronary artery aneurysms can develop in some patients and they occur during 7 days before the onset or up to 4 weeks after onset of the acute symptoms and their formation can be prevented with the use of intravenous immmunoglobin. The other cardiac manifestations include pericarditis, myocarditis and myocardial infarction, valvulitis presenting usually as mitral or aortic regurgitation. Low dose aspiring is recommended until the coronary aneurysms subside usually 4-8 weeks after the onset of illness. In giant aneurysms (>8mm) anticoagulation may be warranted due to largely increase risk of thrombosis and myocardial infacrtion.

EXPERT'S OPINION

This is a rare case of an adult patient with Kawasaki disease and typical involvement of coronary arteries with aneurysm formation. Patients with no evidence of coronary artery aneurysm formation generally do not need treatment with aspirin after 6-8 weeks from the onset of symptoms. No exercise restrictions are also needed. Patients with small (<5mm) or medium (5-8 mm) coronary artery aneurysms require treatment with aspirin 3 mg/kg/day which should be used indefinitely. These patients should be evaluated every year. Angiography is recommended if the invasive tests reveal ischeamia. The exercise tests results should be considered when counseling about the appropriate level of physical activity. Similarly in patients with large aneurysms aspirin treatment is required indefinitely. Anticoagulation is usually added to aspirin for the period of at least 2 years. Every 6 month examination should include ECG, echocardiography. Stress test and myocardial perfusion scan should be done annualy. Angiography should be done initially and then according to symptoms. Catheter based interventions or coronary artery bypass grafting can be considered in case of critical lesions however due to severe calcifications of lesions balloon angioplasty can be suboptimal, therefore surgical treatment is usually of choice .

CONCLUSION

This patient requires treatment with aspirin lifelong and regular follow-up assessment with stress tests to screen for possible ischemia.

REFERENCES

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