The concurrence of ACS with conditions associated with allergic or hypersensitivity and anaphylactic or anaphylactoid reactions constitutes the Kounis syndrome (1). Two variants of Kounis syndrome have been described (2). The type I variant includes patients with normal coronary arteries without predisposing factors for coronary artery disease. The type II variant includes patients with active or quiescent preexisting atheromatous disease. The type III variant has been proposed recently (3). A number of conditions, several drugs, foods and venom and toxins have been reported as capable of inducing Kounis syndrome (1, 2).

Activation of mast cells and the systemic release of histamine are common side effects of morphine. In addition to other side effects, cutaneous changes may occur as manifested by peripheral vasodilatation and flushing of the skin with urticaria, a response to the histamine releasing properties of the morphine. This case calls attention to the Kounis syndrome which was induced by two other β-lactam antibiotics and aggravated by morphine.

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Video 1, 2: Angiographic views of the left and right coronary arteries

References


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Echocardiographic assessment in children with Gaucher disease receiving enzyme replacement therapy

Gaucher hastalığı olan ve enzim replasman tedavisi alan çocukların ekokardiyografik değerlendirilmesi

Cardiac involvement is rare in Gaucher disease and may be in the form of pulmonary hypertension, constrictive pericarditis, pericardial calcifications, various valvular lesions and infiltration of the myocardium. Pulmonary hypertension in Gaucher disease is not common but it is shown to be secondary to interstitial or perivascular infiltration of Gaucher cells or primary in patients exposed to enzyme replacement therapy (ERT). Valvular lesions are seen as calcifications of aortic and mitral valves and these are mainly reported in patients with D409H homozygosity (1).

We investigated echocardiographic findings in our pediatric patients while receiving ERT. Patients with Gaucher disease who received ERT for at least six months were assessed. A Vingmed (GE, Horten, Norway) Vivid-5 echocardiography equipment with 2.5, 3.5 and 5 MHz transducers were used for echocardiographic evaluation. M-mode, 2-dimensional, color Doppler, pulsed wave (PW) Doppler and continuous wave (CW) Doppler examinations were performed in each patient. Echocardiographic assessment was done by the same pediatric cardiologist and tricuspid regurgitation gradient of 30 mmHg was considered as upper limit of normal as it was known to reflect pulmonary pressure in the absence of ventricular outflow obstruction. Other abnormal findings were also recorded.
Ten of the twelve patients in the study were classified as type I and one as type III who was homozygous for D409H mutation. The mean age was 4.5 years at diagnosis and 11.8 years at the time of this study. None had clinical symptoms or findings of cardiac or pulmonary involvement. Echocardiographic findings were normal in eight of the patients. Only one patient had an abnormal tricuspid regurgitation gradient. However re-examination of this patient after six months was found normal. Two of the patients were found to have mitral valve prolapse and atrial septal defect was found in another patient. No valvular calcifications were encountered in our patients including D409H homozygous one.

Echocardiographic assessment is recommended for both adults and children although no clinical signs and symptoms exist. The study about the outcome of ten years’ echocardiographic findings in children with Gaucher disease stated that follow up echocardiography was not necessary if baseline was normal. The abnormal tricuspid regurgitation gradients of patients in this study turned out to normal after six months (2). In our study, high tricuspid regurgitation gradient of our patient was temporary either. Calcifications of aortic and mitral valves are reported in patients with homozygous D409H mutation (3, 4). These calcifications may not be apparent until early adult life and may increase with age (3, 4). Although the echocardiographic findings of our patient with homozygous D409H mutation were normal, we think it must be reassessed annually. Mitral valve prolapse without calcifications were reported in two siblings with Gaucher disease before. Mitral insufficiency was also reported in those cases (5). In our report we also observed two cases with mitral valve prolapse without mitral insufficiency and calcifications. Incidence of mitral valve prolapse is high in otherwise healthy population so it is difficult to say that mitral valve prolapse is more common in Gaucher disease or not.

In conclusion, this study showed that echocardiographic examination after a mean period of three years of ERT was almost normal in children. Follow up echocardiography should be done in cases with D409H mutations and for the ones with prior abnormality.

References


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