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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Kounis syndrome: is it rare or is it underdiagnosed?

Zespół Kounis: rzadki czy niedostatecznie często rozpoznany?

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A 50-year-old male patient with a history of allergy to crustaceans presented to the emergency department for generalised skin rash after eating a tuna sandwich. Suddenly he presented exacerbation of the cutaneous rash, accompanied by non-irradiated oppressive chest pain and profuse sweating that ceased after the administration of IV corticosteroids. Electrocardiogram (ECG) during chest pain showed a 1-mm generalised ST segment depression (Fig. 1). When the patient was asymptomatic, the ECG had no repolarisation abnormalities (Fig. 2). He was admitted with the diagnosis of non-ST-elevation myocardial infarction. Echocardiogram at admission showed inferolateral hypokinesia. During hospitalisation the patient presented a peak troponin I of 5.25 ng/mL. A coronarography was performed and no lesions were diagnosed (Fig. 3), so the final diagnosis of type I Kounis syndrome was confirmed. The appearance of acute coronary syndrome (ACS) in the context of an allergic reaction was described in 1991 by Kounis and Zavras as a "myocardial allergic infarction". The pathophysiology of this syndrome is not clear. One of the most accepted mechanisms is the degranulation of mast cells that release inflammatory mediators (such as histamine, proteases, and several cytokines) that are responsible for coronary vasospasm and/or rupture of the atheroma plaque. Another mechanism described in the literature is the acute secretion of tryptase, which in turn activates other neuromechanisms that lead to rupture of an atheroma plaque and formation of intravascular thrombus. Kounis syndrome is classically divided into two variants, although in recent years a new variant has been described: (i) Type I: occurs in patients without coronary lesions. It is caused by coronary vasospasm with electrocardiographic changes. Cardiac enzymes may be normal. This can be explained by endothelial dysfunction and/or microvascular angina; (ii) Type II: occurs in patients with pre-existing atherosclerotic plaques. The previously described mediators induce plaque erosion, causing an ACS; (iii) Type III: occurs with thrombosis of drug-eluting stents. Histopathological confirmation of the presence of eosinophils and mast cells in the thrombus is required. The diagnosis is clinical: signs of an acute allergic reaction and a simultaneous ACS. There is no specific diagnostic test for this syndrome and it is probably underdiagnosed because of the variable manifestations of an allergic reaction. The usual treatment of ACS is recommended as an initial treatment, although in type I variant the treatment of the allergic reaction could be enough. In type II patients the administration of beta-blockers may aggravate coronary vasospasm, as well as the administration of adrenaline in case of anaphylactic shock. The prognosis is usually better in patients with the type I variant. However, it depends on the initial allergic response, the patient's sensitivity, the various comorbidities, allergen concentration, and the port of entry. In the long term and after the acute phase, the prognosis is excellent.

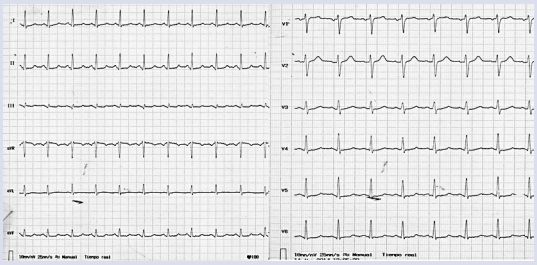

Figure 1. Electrocardiogram during chest pain

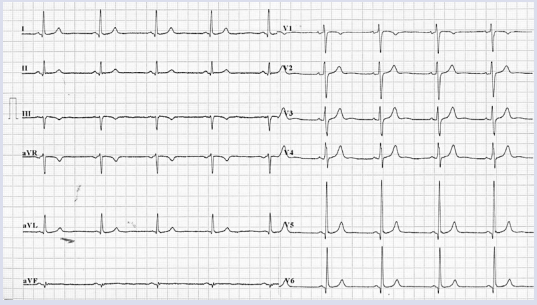

Figure 2. Electrocardiogram without chest pain

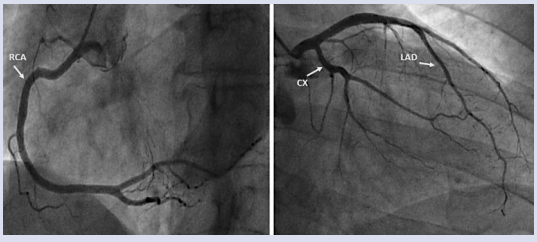

Figure 3. Coronarography; RCA — right coronary artery; LAD — left anterior descending artery; CX — circumflex artery

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