Kounis syndrome: A forgotten cause of chest pain/ Cardiac chest pain in children

Kounis sendromu: Göğüs ağrısının unutulan bir sebebi/ Çocuklarda kardiyak göğüs ağrısı

Dear Editor,

I read with interest the article "Cardiac chest pain in children" by Çağdaş et al. (1) which has retrospectively evaluated 120 children admitted to a pediatric cardiology clinic with chest pain. Although chest pain in children is rarely reported to be associated with cardiac diseases in the literature (2) authors have found that 52 of the patients (42.5%) had cardiac diseases and 28 (23.3%) of these patients' cardiac diseases were thought to directly cause their chest pain. These higher ratios may result experience of a tertiary referral centre. Although a variety of diseases is described as the causes of the chest pain in the present study, I want to remind a forgotten cause of chest pain in children.

Kounis syndrome (allergic angina or allergic myocardial infarction) is the coincidental occurrence of chest pain and allergic reactions accompanied by clinical and laboratory findings of classical angina pectoris caused by inflammatory mediators released during an allergic insult (3). There are several factors, which have been reported as capable of inducing Kounis syndrome. These include a number of conditions, several drugs, foods, insect stings and etc. The mechanism of Kounis syndrome is coronary artery spasm due to mast cells degranulation and the subsequent release of vasoactive mediators (3).

Three variants of Kounis syndrome have been described (3-5). Type I, occurring in patients with angiographically normal coronary arteries, and type II, occurring in patients in whom concomitant atheromatous lesions are found. Type III variant includes patients with coronary artery stent thrombosis in whom aspirated thrombus specimens stained with hematoxylin-eosin and Giemsa demonstrate the presence of eosinophils and mast cells. However, since type II and type III variants include patients with preexisting atheromatous disease, I think that it is almost impossible to encounter the type II and type III variants in childhood.

After the first report of Kounis syndrome in a 13-year-old boy (6), we have recently published first children with Kounis syndrome in the literature (7). Although nearly 300 cases with Kounis syndrome have been reported in the literature I think that many of the cases are misdiagnosed or unreported.

I want to emphasize that pediatricians should be aware of the allergic myocardial infarction. The diagnosis of this unique disease should be entertained when acute-onset chest pain is accompanied by allergic symptoms, electrocardiographic changes and elevated cardiac enzymes. All adults and children admitted to the emergency departments with chest pain and ST elevation on electrocardiography, should be interrogated for allergic insults. Kounis syndrome is not a very rare disease but a 'very rarely diagnosed' disease.

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Author's Reply

Dear Editor,

We thank author of the letter for their valuable comments for our study. In our study, we found that the risks of structural cardiac disease and arrhythmia and the admission with chest pain increase with increasing age in children. In addition, we found a high ratio of cardiac disease in children with chest pain. First reason of this may be the high frequency of some cardiac diseases in children such as MVP or ventricular/supraventricular ectopies. Second, some of our patients were referred patients. Third and the most important reason is the standard evaluation of patients, using our algorithm for chest pain, with standard questions for evaluation and standard diagnostic tests.

In the study of Selbst et al. (1), the frequency of cardiac pathology was found to be 4%, but they reported in their study that fifty three percent of the patients were not evaluated by electrocardiography (ECG). So, the frequency of cardiac pathology in their study may not reflect the real frequency. In our study, all patients were evaluated with ECG. Kounis syndrome, which is the result of ischemic changes caused by inflammatory mediators released during allergic reactions, may be suspected only, if the patient is evaluated with ECG. In our study, our patients did not have both electrocardiographic findings of ischemia and allergic symptoms.

There are many different causes of chest pain in children like Kounis syndrome (2, 3), so there is need for further investigations in the evaluation of chest pain in children. Using an algorithm with standard workup will make the evaluation easier in the outpatient clinics.

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Swyer-James syndrome or destroyed lung?/ Coronary artery bypass in a patient with Swyer-James syndrome due to pulmonary tuberculosis

Swyer James sendromu mu yoksa harap akciğer mi?/ Pulmoner tüberküloza bağlı Swyer-James sendromlu bir hastada koroner arter baypas olgusu

Dear Editor,

We read with great interest the recent article of Özcan et al. (1) about coronary artery bypass in a patient with Swyer-James Syndrome (SJS) secondary to pulmonary tuberculosis (TB). However, we do not agree with the authors regarding their diagnosis of Swyer-James Syndrome, but feel that it is more consistent with destroyed left lung secondary to pulmonary TB and compensatory left sided- mediastinal shift of right lung. It is well documented that SJS is a rare syndrome characterized by unilateral hyperlucency of one lung, lobe or part of a lobe which was first described in 1953 by Swyer and James (2) and further detailed by MacLeod (and called Swyer-James-MacLeod Syndrome (SJMS)). SJMS is considered a postinfectious manifestation of childhood bronchiolitis obliterans (BO). These respiratory infections include measles, whooping cough, TB, Mycoplasma pneumonia and influenza A (3, 4). BO results in inflammation and fibrosis in the walls and contiguous tissues of respiratory bronchioles with narrowing of their lumens (4). Fibrosis of the interalveolar septa results in obliteration of the pulmonary capillary bed secondarily diminishes blood flow to the major pulmonary artery segments, causing the hypoplastic arterial development. Because of the decreased parenchymal perfusion, this syndrome is often mentioned as translucent or hyperlucent lung (5, 6).

The chest X- ray finding of SJMS is a noticeable one-sided hyperlucency caused by oligemia of the involved lung segments. A mediastinal shift toward the affected side may occur on inspiration. Air trapping or a shift of the mediastinum towards the unaffected side may be demonstrated by expiratory chest X- ray. On thorax computed tomography (CT) and high resolution computed tomography (HRCT), SJMS appears as hyperlucent areas due to decreased pulmonary perfusion of the lung without an anteroposterior gradient attenuation (6, 7). In addition, ventilation perfusion (V/Q) scans are important modality in the diagnosis of SJMS. V/Q scans document matched ventilation and perfusion defect (3-7).

As a result, we thought that chest X-ray and thorax CT demonstrated in the article by Özcan et al. (1) fit into the characteristic radiographic appearance of destroyed lung secondary to pulmonary TB rather than SJMS.

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Author's Reply

Dear Editor,

In our country, open heart surgery operation procedures include cost management in fixed prices determined by social security association (SGK) so that some expensive tests cannot be applied like VPS as precious reviewer mentioned in 3th and 4th topic of his/her critic. Original presentation formation also included restriction of 250 words. Our real aim was to emphasize cardiopulmonary bypass operation can be made in such situations of destroyed lung. Under the constriction of these limitations and conditions, the case named as Swyer-James Syndrome.

We thank authors of the Letter to the Editor for their comments on our article.