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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

SAPHO syndrome and acute coronary syndrome

Zespół SAPHO i ostry zespół wieńcowy

Paweł Tyczyński[1], Mariusz Kruk[2], Maria Gosk-Przybyłek[3], Łukasz Kalińczuk[2], Adam Witkowski[1]

[1]Department of Interventional Cardiology and Angiology, Institute of Cardiology, Warsaw, Poland
[2]Department of Coronary and Structural Heart Disease, Institute of Cardiology, Warsaw, Poland
[3]Department of Hypertension, Institute of Cardiology, Warsaw, Poland

Chest pain is one of the most common causes for referral to emergency departments, of which cardiac disease accounts for only 8–18% cases. The majority of chest pain seen in primary care is due to more benign conditions. However, musculoskeletal causes of chest pain may present concomitantly with myocardial ischaemia. Inflammatory rheumatoid arthritis (IRA) is associated with earlier development and acceleration of atherosclerosis. SAPHO syndrome is a very rare condition with a prevalence of 0.04%, representing the spectrum of inflammatory bone disorders. Although it was defined as a distinct entity more than 20 years ago, no association of this disease with acute coronary syndrome (ACS) was reported so far. Only a casuistic report of SAPHO syndrome with coronary artery disease (CAD) coexistence was published. We present a patient with SAPHO syndrome, who experienced ACS. A 56-year-old female patient was admitted due to acute retrosternal pain. Her past medical history included SAPHO syndrome, hypercholesterolaemia, and nicotine use. SAPHO syndrome was diagnosed one year previously, although concomitant cutaneous and osteoarticular manifestations (mainly limited to the right-sided mandible and not sterno-costal joints) were present periodically for several years. Whole-body scintigraphy revealed hyperostosis and non-infectious osteitis of costosternal joints and right-sided mandible. Her current pharmacological treatment included methotrexate and atorvastatin. Acute ST-elevation myocardial infarction of the inferior wall was diagnosed. Emergent coronary angiography revealed critical stenosis and thrombus in the mid right coronary artery (RCA), but with TIMI 3 flow (Fig. 1A). After administration of loading dose of ticagrelor and continuous intravenous flow of eptifibatide, percutaneous angioplasty of RCA with implantation of a drug eluting stent was performed. A good angiographic result was achieved (Fig. 1B). Maximal troponin T was 1827 ng/L (UNL < 14). Echocardiography revealed mild hypokinesia of the inferior wall. SAPHO syndrome is predominant in middle-age women, characterised by dermatological and osteoarticular manifestations (synovitis, acne, pustulosis, hyperostosis, osteitis) of unknown aetiology. Some studies suggest certain inflammatory pathways, with markers like C-reactive protein (CRP), interleukin (IL)-6, and IL-18. [Hurtado-Nedelec et al. Rheumatology, 2008; 47: 1160–1167]. From a clinical perspective IL-18 was proven to be a stronger predictor than CRP, Il-6, or fibrinogen of cardiovascular events in healthy, middle-aged European men. Differential diagnosis: Upper sternalis syndrome may result from the sternoclavicular joint pathologies, such as subluxation or other types of arthritis or just SAPHO syndrome. In the latter, the most commonly affected skeletal site is the anterior chest wall region, consisting of the sternocostal joint, sternoclavicular joint, and manubriosternal joint. In one of the largest SAPHO patient cohorts, all of them demonstrated some kind of anterior chest pain. Nevertheless, no ACS or CAD was reported (Li et al. Rheumatology, 2016; 55: 1023––1030). Finally, methotrexate has been shown to play a protective role against cardiovascular disease in IRA-patients. A more than casuistic coexistence of SAPHO syndrome and ACS may not be proven nor excluded in this case. Nonetheless, IRA patients yield increased risk of atherosclerosis and adverse cardiovascular events. Therefore, acute chest or mandibular pain even of a character mimicking patient's usual SAPHO syndrome discomfort must lead to the exclusion of ACS.

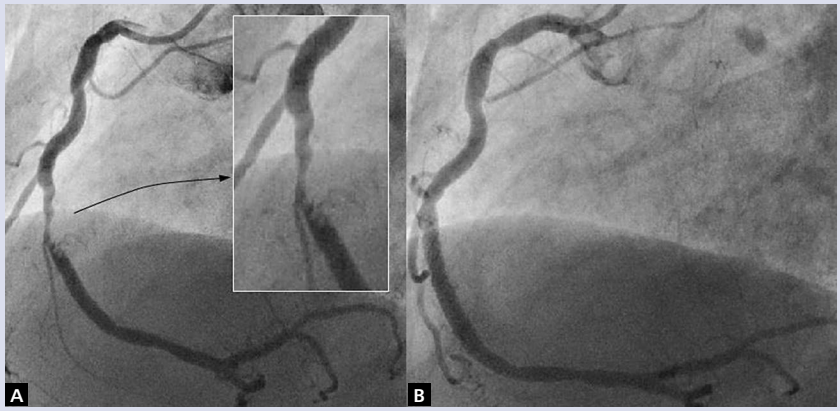

Figure 1. **A.** Tight stenosis in the mid segment of the right coronary artery. Magnification of the narrowing; **B.** Satisfactory angiographic result after implantation of drug eluting stent

Address for correspondence:
Paweł Tyczyński, MD, PhD, Department of Interventional Cardiology and Angiology, Institute of Cardiology, ul. Alpejska 42, 04–628 Warszawa, Poland, e-mail: medykpol@wp.pl

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