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Severe cardiovascular involvement in a patient with rheumatoid arthritis

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A 57-year-old female patient was admitted to the emergency department with angina pectoris. Her past medical history included hypertension and rheumatoid arthritis (RA). Electrocardiography revealed diffuse ST-segment depression. She was hospitalized with a diagnosis of the acute coronary syndrome. Diffuse aortic calcification was detected on posteroanterior and lateral chest radiography (Figure 1A and 1B). Echocardiography revealed severe aortic regurgitation and aortic root calcification (Figure S1A and S1B). In addition, moderate mitral regurgitation and mitral annular calcification were observed (Figure S1C and S1D). The left ventricular ejection fraction was normal. Coronary angiography revealed calcification in left main coronary artery without evident obstructive coronary artery disease (Figure S1E). We investigated the patient for widespread calcification of major vessels and viscera. Pathologic parenchymal punctate calcifications and nodular choroid plexus calcification were observed in the cerebrum on computed tomography (CT) images (Figure S1F). Carotid artery Doppler ultrasound demonstrated atherosclerotic plaques in the left carotid artery and complete occlusion of the right carotid artery. Diffuse atheromatous plaques were detected in the aorta on sagittal and axial plane conventional thoracic CT images (Figure 1C and 1D) and three-dimensional reconstruction thoracic CT images, indicating porcelain aorta (PA) (Figure 1E). It was observed that episodes of bradycardia and tachycardia, detected by bedside monitoring, resulted in severe dyspnea and chest discomfort. She was symptom free other than these attacks during hospitalization. Severe bradycardia led to frequent premature ventricular contractions with long compensatory pauses. Twenty-four hour Holter monitoring recordings were compatible with sick sinus syndrome.

The underlying pathogenesis of cardiovascular disease in RA involves diffuse subclinical atherosclerosis and atherosclerotic plaque calcification caused by chronic inflammation [1]. The extent of systemic inflammation is a predictor of poor cardiovascular outcomes [2]. Aorta is the one of the main target tissue in most of autoimmune diseases [3]. Patients with RA have
higher risk of developing calcification in the aorta, and carotid and coronary arteries [4]. PA is detected incidentally on chest radiography or CT images as extensive calcification of the aorta. It is associated with increased morbidity and mortality. The atherosclerotic PA is associated with RA and seen in individuals with hypertension, hyperlipidemia, and other autoimmune diseases [5]. Chronic inflammatory response leads to immunological vascular damage, which triggers microinfarctions and contributes to dystrophic calcification of soft tissues.

We diagnosed valvular heart disease as mitral and aortic valve regurgitation, cardiac conduction disorder as sick sinus syndrome, and diffuse arterial calcification as PA in our patient. We detected the complete occlusion of the right carotid artery, and dystrophic cerebral and cardiac calcifications. We attributed severe dyspnea, and chest discomfort attacks to increased aortic insufficiency during bradycardia episodes and increased mitral insufficiency to tachycardia episodes. We implanted a dual-chamber pacemaker in the patient to prevent bradycardia episodes. To prevent tachycardia attacks, we administered the maximum well-tolerated dose of a beta-blocker to the patient, which resulted in prominent symptomatic relief in 1 week.

In conclusion, advanced cardiovascular and cerebral investigations should be performed during the clinical evaluation of patients with RA.

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Figure 1.

**Figure 1A and B.** Chest radiography demonstrating diffuse aortic calcification.

**Figure 1C and D.** Conventional thoracic computed tomography images demonstrating diffuse aortic calcification.

**Figure 1E.** Three-dimensional reconstruction computed tomography images of the porcelain aorta.