Right atrial anomalous muscle bundle. Clinical implications

Maciej K Michałowski[1], Maria Sudomir[1], Tomasz Oleksiuk[2], Paweł Tyczyński[1], Ilona Michałowska[3]

[1]Department of Interventional Cardiology and Angiology, National Institute of Cardiology, Warszawa, Poland
[2]Department of Radiology, Mazowian Bródnowski Hospital, Warszawa, Poland
[3]Department of Radiology, National Institute of Cardiology, Warszawa, Poland

Correspondence to:
Paweł Tyczyński, MD, PhD, Department of Interventional Cardiology and Angiology, National Institute of Cardiology, Alpejska 42, 04–628 Warszawa, Poland,
phone: +48 22 343 42 72,
e-mail: medykpol@wp.pl

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Right atrial muscle bundle (RAMB) is a congenital anomaly of unknown origin, probably deriving from the right venous valve [1]. So far, only four patients with RAMB have been reported, including two children [1–4]. Similar malformations were found much more frequently in the left atrium. In an impressive study by Yamashita et al. (Supplementary material, [5]) of 1100 autopsies, left atrial muscle bundle (LAMB) was observed in 2% of cases and coexisted with other abnormalities, such as premature atrial complexes (41%), Chari's network (27%), or patent foramen ovale (23%). LAMB may also cause mitral valve prolapse and its regurgitation. Potential arrhythmogenicity of LAMB remains unknown.

A 60-year-old male with a 3-month history of hemoptysis, rest dyspnea, and deterioration of exercise tolerance was admitted with suspicion of pulmonary embolism (PE). His previous medical history included heart failure and stable coronary artery disease. He also underwent implantation of a double-chamber pacemaker due to tachycardia-bradycardia syndrome 6 years earlier, aortic valve replacement (St. Jude prosthesis) with coronary artery bypass grafting twelve years earlier, and supracoronary prosthesis implantation due to aneurysm of the ascending aorta with two re-do operations due to post-operative complications 6 years earlier. On admission, there were no signs of local or generalized infection (normal device pocket, body temperature within normal limits, no elevated inflammatory biomarkers). Transthoracic echocardiography (TTE) showed dilated both ventricles, significantly reduced left ventricular ejection fraction (20%), and elevated pulmonary artery systolic pressure (90 mm Hg). Moreover, an additional echo was seen on the ventricular electrode in the right atrium, suggesting an old organized thrombus. Cardiac computed tomography (cCT) showed a structure with dimensions of 55 × 5 mm, resembling a thrombus and located between the superior vena cava (SVC) and the right atrial appendage (Figure 1). No thrombi in the pulmonary arteries were visualized.

The patient was treated with an intravenous infusion of tissue plasminogen activator, heparin (initially unfractionated and subsequently low-molecular weight), and diuretics, which led to clinical improvement.

On repeated cCT examination one month later, the additional structure looked virtually identical. Therefore, a strong suspicion was raised that it was not a thrombus but RAMB. Three years later, the patient underwent an uneventful pacemaker upgrade to cardiac resynchronization therapy-defibrillator. The clinical follow-up two years later was also unremarkable.

The clinical significance of RAMB remains largely unclear. Philip et al. reported that RAMB precluded percutaneous closure of atrial septal defect (ASD) [2]. Madjarov et al. [3] presented a patient with RAMB, who developed SVC syndrome with pulmonary embolism. Recently, Kofler et al. [4] described a fourth case of a patient with RAMB, who presented with two ASD and supraventricular tachycardia (both were successfully treated percutaneously).

In our case, we wish to emphasize that visualization of RAMB may be challenging, especially in patients with pacemakers (none of the previously reported patients had a pacemaker). Cardiac device-related infective endocarditis, lead-thrombosis, or another thrombus [5] must be excluded. Thus, careful evaluation of inflammatory biomarkers (like white blood cells, C-reactive protein, and procalcitonin)

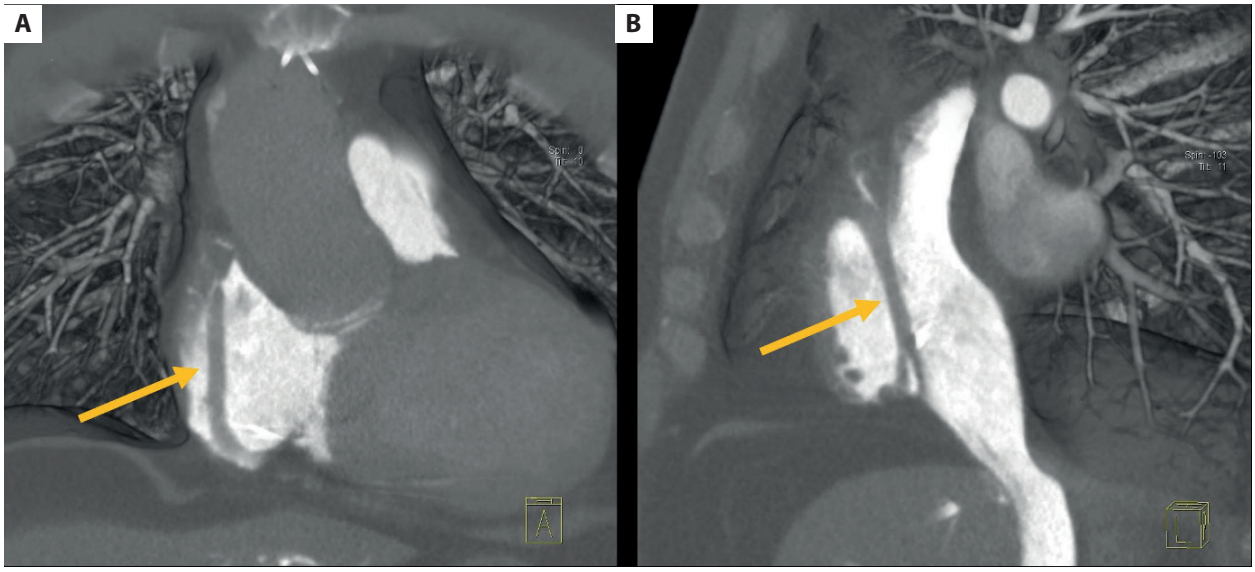

Figure 1. Cardiac computed tomography. Muscle bundle in the right atrium (yellow arrow): Coronal (**A**) and sagital (**B**) view

and blood cultures, as well as multimodality visualization by TTE, transesophageal echocardiography, cCT, or even positron emission tomography, may be indispensable in dubious cases. In previously reported cases, RAMB was visualized by TTE/transesophageal echocardiography, not by cCT or positron emission tomography.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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