Spontaneous Echo Contrast in the Right Heart Chambers of a Patient with Autoimmune Hemolytic Anemia

Spontaneous echo contrast (SEC) is an echocardiographic phenomenon characterized by slow intracavitary contrast movement, like a "smoke cloud". It is caused by the aggregation of red blood cells, which provides blood with echogenic capacity.

We report a case of intense SEC in a rare location. The case corresponds to a 24-year-old male patient with history of systemic lupus erythematosus (SLE), who consulted for a 48-hour history of arthritis in the left knee. On admission, his blood pressure was 95/60 mmHg, his heart rate 120 bpm, and axillary temperature 38 °C.

Lab tests: Hematocrit 25%, hemoglobin 8 g/dl; WBC 12,000/mm3; platelets 120.000/mm3; total bilirubin 1 mg/dl; positive direct and indirect Coombs tests with cold agglutinins in serum.

Streptococcus agalactiae was isolated from blood cultures and synovial fluid. The patient was hospitalized with diagnosis of septic arthritis associated with hemolytic anemia. Antibiotic therapy with imipenem/vancomycin was started. The patient progressed to septic shock, requiring mechanical ventilation. Hydrocortisone and IV gamma globulin therapy was initiated.

Transthoracic echocardiography showed left heart chambers with normal diameters and left ventricular systolic function and presence of dense SEC in non-enlarged right chambers, inferior vena cava and suprahepatic veins (Figure 1A and B). Moderate pericardial effusion and mild mitral, aortic, and tricuspid regurgitation was also observed. Echocardiography was performed without echographic contrast, and no other clear bubble source was found.

An Echo-Doppler of the lower limbs showed no signs of venous thrombosis, and a chest CT scan with intravenous contrast ruled out pulmonary thromboembolism (PTE).

Shock improved and the hematocrit was stabilized without transfusion or anticoagulant therapy.

Transthoracic echocardiography was repeated one week after the first study showing SEC absence (Figure 2A and B). Subsequently, a transesophageal echocardiography confirmed disappearance of SEC and absence of vegetation.

Normally, during echocardiography, cardiac chambers are free from echoes because ultrasound reflection on blood is not intense enough to generate images. However, under blood congestion, red cells stack together (rouleaux), which provides echogenic capacity that can be observed as a "smoke cloud". (1)

Spontaneous echo contrast depends on the concentration of red blood cells, blood velocity, and presence of plasma proteins. (2) Individual red blood cells are normally prevented from aggregating by the repulsive electrostatic effects of their negatively charged surface caused by plasma proteins, particularly fibrinogen. (3) Spontaneous echo contrast generation is more common with increased fibrinogen, which would act as a screen on which erythrocyte aggregation occurs more easily. (2, 4)

Spontaneous echo contrast is observed by transthoracic echocardiography in left chambers in 0.1% to 3.5% of studies in patients with conditions causing blood stasis, such as chronic atrial fibrillation, severe mitral stenosis, and dilated cardiomyopathy with low cardiac output. It is considered to predict embolism, so patients should be considered for anticoagulant therapy. (2)

Although most often detected in the left atrium and left atrial appendage, SEC has also been described within the left ventricle, right heart chambers, descending aorta, pulmonary artery, and inferior vena cava. (1) Cases of SEC in the right atrium and ventricle have been reported in patients with autoimmune hemolytic anemia and erythrocyte aggregation mediated by auto-antibodies. (5, 6)

Miller et al. reported the case of a 9-year-old girl with a history of Evans syndrome (thrombocytopenia and autoimmune hemolytic anemia), who was admitted due to fulminant disseminated aspergillosis. During hospitalization, the echocardiogram showed SEC in the right heart chambers (not in the left chambers) resulting from red blood cell agglutination mediated by 'warm' IgM antibodies. Autopsy revealed pulmonary thromboembolism (PTE) and infarction in lungs, brain, liver, and kidneys. (5)

Similarly, Dogan et al. reported the case of a 67-year-old male patient with SEC in the right heart

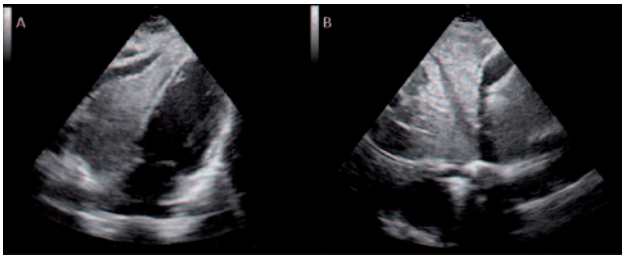


Fig. 1. Transthoracic echocardiography, subcostal view. Spontaneous echo contrast in the right chambers (**A**) and in the inferior vena cava and suprahepatic veins (**B**).

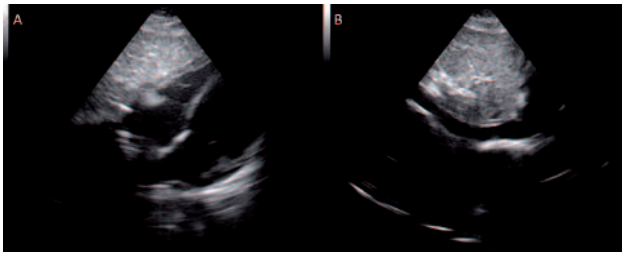


Fig. 2. Transthoracic echocardiography, subcostal view. Disappearance of spontaneous echo contrast in the right chambers (**A**) and in the inferior vena cava (**B**).

chambers and chronic obstructive pulmonary disease, who was admitted due to pneumonia and autoimmune hemolytic anemia. In addition to PTE, this patient had pulmonary hypertension and right ventricular enlargement. Spontaneous echo contrast in the right heart chambers disappeared after a week with corticoid therapy and anticoagulation with low-molecular-weight heparin and warfarin. (6)

We report the case of a patient with SLE, septic shock, and autoimmune hemolytic anemia, with intense SEC detected in a rare location (right heart chambers, inferior vena cava, and suprahepatic veins). Different from the cases described above, no signs of PTE were found in our patient. Moreover, SEC disappeared with the remission of shock and "hemolytic crisis" under corticoid and gamma globulin therapy, and without anticoagulant therapy. Therefore, it would be reasonable to assume that the major SEC mechanism was erythrocyte aggregation caused by the interaction between red blood cells and auto-antibodies. The lower shear stress in the venous circulation (especially in septic shock) would contribute to SEC production in the right heart circulation. The absence of SEC in the left heart chambers would occur because the erythrocyte aggregation in "stacks of coins" or rouleaux disintegrate in the pulmonary microcirculation or are retained by the pulmonary capillaries acting as filter.

According to our review of the literature, this would be the first report of spontaneous echo contrast caused by this mechanism, in the absence of PTE, and disappearing without anticoagulant therapy.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/Supplementary material).

Juan C. Ruffino, Miguel Amor[MTSAC], Elías G. Benchuga, Hugo A. Mosto, Rita Tepper, Rafael S. Acunzo[MTSAC]

Department of Cardiology, Hospital Ramos Mejía

Juan C. Ruffino

Department of Cardiology, Hospital Ramos Mejía

Urquiza 609 - (1221) Buenos Aires, Argentina.

e-mail: ruffinojuancarlos@gmail.com

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Recurrent Syncope in Patient with Multinodular Goiter

Syncope is a rare clinical manifestation of cervical tumors. (1) Its pathophysiology lies in the mechanical compression or irritation caused by the tumor on the carotid sinus and the IX cranial nerve (glossopharyngeal nerve). As a result, an exaggerated baroreflex response is triggered as the precursor of the so called neuromediated syncope. (2) We report a rare case of cervical goiter first manifested as syncope.

The case corresponds to a 67-year-old hypertensive and diabetic female patient, with chronic anemia secondary to atrophic gastritis. She had daily syncopal episodes of 2-month evolution, associated with movements of the neck and upper limbs. Physical examination revealed a palpable cervical tumor. Baseline ECG showed a sinus rhythm of 65 bpm, first-degree atrioventricular (AV) block (PR interval 220 ms), narrow QRS, and no other abnormalities (Figure 1A). Electrocardiographic monitoring during syncopes revealed a paroxysmal AV block with QRS intervals >3.5 seconds (Figure 1B and C). A thyroid ultrasound followed by a thorax CT scan confirmed the presence of a large left-sided multinodular goiter with bilateral

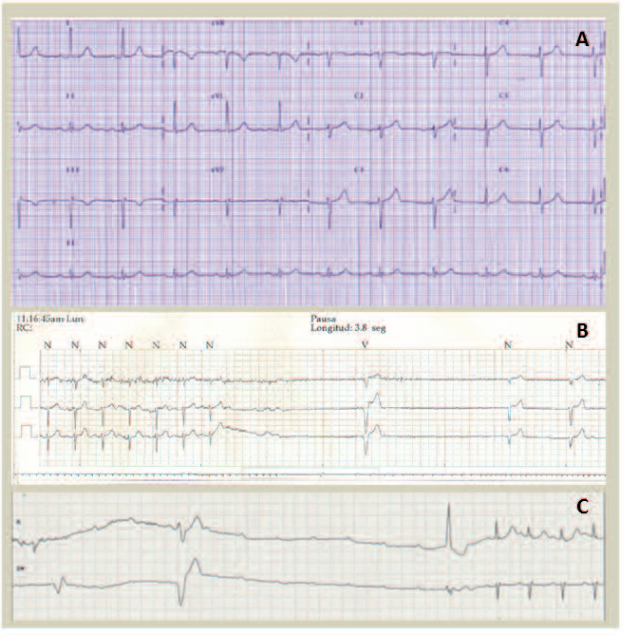


Fig. 1. A. Baseline ECG showing sinus rhythm with prolonged PR interval. **B** and **C**. Complete paroxysmal AV block during two syncopal episodes.