**Brief Communication**

Can Eustachian valve be a risk factor for pulmonary embolism?

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**INTRODUCTION:**
The major risk factor for pulmonary embolism is the presence of right atrial thrombi and some of these originate primarily from right atrium and the others from lower extremities. Development of pulmonary embolism related to right atrial thrombus is associated with higher mortality compared to pulmonary embolism cases related to other causes. In this report two cases of pulmonary embolism with thrombus accompanied by right atrial eustachian valve (EV) are presented, one occurring after childbirth and the other one with chronic obstructive pulmonary disease (COPD).

**Case 1:**
A 31-year old woman without any prior complaints experienced progressively worsening shortness of breath, palpitations and fatigue after brief periods of activity during the last trimester of her pregnancy. She admitted to a medical center for her complaints continuing after the delivery and transthoracic echocardiography (TTE) examination revealed normal left ventricle functions, minimal tricuspid failure and symptoms of pulmonary hypertension (pulmonary artery systolic blood pressure 45 mmHg). About one year after the delivery she started experiencing swelling in the abdomen and lower extremities and she admitted to another center; TTE showed marked dilation in right cardiac spaces, third-degree tricuspid failure and advanced pulmonary hypertension (85 mmHg). Following her referral to our center it was found that she had severe shortness of breath, ascites in the abdomen and edema in her legs. Other examination findings included body temperature of 37.4°C, blood pressure 90/60 mmHg, pulse 120 /min, visible neck veins, 3/6 systolic murmur in the tricuspid focus, decreased respiratory sounds in the lungs, tenderness in the upper right quadrant of the abdomen and liver size of 5-6 cm. Laboratory results included total bilirubin 2.1mg/dl, erythrocyte sedimentation rate 22 mm/h, D-dimer 2000 ng FEU/ml and Factor V-Leiden 1691 G>A heterozygous, anticyclolipid IgG, IgM negative, protein-S 61%, homocysteine 12.06 umol/L, antithrombin III 99.91%. The electrocardiogram showed sinus rythm, V1-4 R>S, ST depression and T negativity. Telecardiography revealed cardiomegaly and marked pulmonary conus and non-specific infiltration zones within the pulmonary parenchymal tissues. TTE examination showed normal left cardiac functions and valves, severely dilated right cardiac spaces, tricuspid valve failure (3+), intact interatrial septum, dilation in the right atrium; and a mobile mass extending from the junction of the inferior vena cava and right atrium up to interatrial septum has been identified as EV (Figure 1). Lower extremity venous and hepatic portal vein color-Doppler examination was normal. Dilation in the pulmonary vascular structures and subpleural consolidated zones identified by the thoracic tomography were reported to be consistent with pulmonary embolism. Transesophageal echocardiographic examination identified intact interatrial septum, dilation in the right atrium and echo images consistent with thrombus at the junction of vena cava inferior and right atrium. The mismatch ventilation/perfusion defect identified by the pulmonary ventilation and perfusion scintigraphy was reported as highly probable pulmonary embolism. The patient had a preliminary diagnosis of pulmonary embolism and was started to a treatment regimen consisting of infusional heparin (5000 U loading dose and 1000/h maintenance dose), followed by warfarin 1x5 mg (INR to remain between 2-3), acetyl salicylic acid 1 x 300 mg/day, diltiazem 1x120 mg/day, furosemide 1x20 mg/day and spirinolactone 1x25 mg/day. The patient’s complaints were under control posttreatment. Serial echocardiographic examinations still showed images consistent with thrombus in the proximity of EV in the right atrium and scintigraphy after about 1 year showed persistence of preexisting perfusion defects and new perfusion defects were present.
Case 2:
A 52-year old male patient admitted with generalized swelling in the abdomen and legs, shortness of breath and fatigue after brief periods of activity that were present within the previous year. COPD was identified in his personal history and his physical examination revealed abdominal ascites, venous distension in the neck and bilateral pretibial edema. Laboratory results were within normal range; ECG showed incomplete right branch block and cardiomegaly and non-specific infiltrations were identified by the telecardiography. Protein S,C and factor V-Leiden were in normal range; D-Dimer, anti-phospholipid antibody was negative. Lower extremity Doppler examination was normal; the pulmonary ventilation and perfusion scintigraphy identified multiple mismatch ventilation/perfusion defects in both lungs and highly probable embolism. TTE showed severe dilation in the right cardiac spaces; within the right atrium, membrane extending from coronary sinus orifice up to inferior vena cava and thrombus were identified with diffuse spontaneous echocontrast (Figure 2). Pulmonary artery pressure was 80 mmHg. Left ventricle function and dimensions were within normal range. It was found that tension of the Eustachian membrane was increased due to excessive dilation of the right atrium and its mobility was restricted. The patient was discharged on a treatment regimen including furocemide 1x20 mg/day, spironolactone 1x25 mg/day, acetyl salicyclic acid 1x300mg/day and warfarin 1x5 mg/day.

Figure 1. Transthoracic echocardiographic view in the subcostal position showing between inferior vena cava (IVC) and right atrium (RA) trombus (Tro), the eustachian valv (EV), Ao=Aort, IVC=inferior vena cava
DISCUSSION:
Right atrium may be a possible source of both pulmonary and systemic embolisms. Some of the masses localized in the right atrium are congenital and in several articles, congenital right atrial structures have been reported to predispose to formation of thrombus and vegetations [1,2]. The function of the Eustachian valve, an embryonic remnant localized in the right atrium is to direct some of the venous blood coming to the heart from vena cava inferior to patent foramen ovale during the intrauterine period. However, most of this structure regresses during adulthood. Wide use of echocardiography facilitated identification of masses localized in the right atrial space and defining associated structures. EV could be erroneously considered as structures such as vegetations [1] and thrombi [2] localized in the right atrium. Also, mixomas and fibroelastomas, common primary tumors of the heart, could originate from EV and appear as pseudothrombus [3-5]. In some cases, the extension of EV localized in the right atrial space up to vena cava inferior and right atrial segment could be clearly identified by using TTE. Also, TEE provides additional information on the presence of right atrial thrombus and integrity of interatrial septum and localization of thrombus and its association with EV. Combined use of TTE and TEE allows localization of EV and associated structures such as thrombus or vegetation and also helps diagnosing complications caused by thrombus including pulmonary embolism. Congenital structures localized in the right atrium may predispose to formation of vegetation and thrombus [6] by yet unknown mechanisms. In several publications, endocarditis cases associated with EV have been described [1,3]. Neither clinical findings nor laboratory tests were consistent with endocarditis in our cases. Although echocardiography is commonly used for the evaluation of cases with pulmonary embolism, definite diagnosis cannot be made in the majority of such cases. Echocardiographic examinations of our cases with pulmonary embolism have revealed presence of EV and thrombus in the right atrium and also, predisposing factors such as COPD and pregnancy were present. In a study investigating the contribution of right atrial thrombus in cases with pulmonary embolism, proportion of right atrial thrombus was found to be comparable in cases with or without COPD [7]. This suggests the contribution of additional factors besides COPD causing development of right atrial thrombus per se. Dilated right cardiac spaces may predispose to thrombus formation in the right atrium due to stasis. In conditions where clotting tendency is increased such as pregnancy, presence of a mechanic barrier in the right atrium that causes stasis of blood flow might predispose to thrombus formation. Some of the right atrial thrombi originate from the venous system in the lower extremity. Some of the embolized thrombi deriving from the deep veins in the lower extremity may attach to EV within the right atrium [8]. No thrombus was identified in our patients by the bilateral lower extremity Doppler examination. The identification of new perfusion defects at the pulmonary scintigraphy one year later in the segments previously ventilated without prior perfusion defects in our first case suggests that right atrial thrombi cause recurrent pulmonary embolisms. In addition, absence of Behçet’s disease and other rheumatologic disorders, negativity of factor V-Leiden mutation and tests for clotting tendency such as protein S and C and localization of the thrombus between the junction of vena cava inferior and right atrium and EV in all cases suggests that the pulmonary embolism originates from EV. On the other hand concomitance of EV, right atrial thrombus and pulmonary embolism in our patient with COPD supports the argument that routine use of echocardiography would be beneficial with respect to the risk of pulmonary embolism.

Figure 2. TTE shows severe dilation in the right cardiac spaces; within the right atrium (RA), membrane extending from coronary sinus orifice up to inferior vena cava and thrombus. Eustachian valve (EV), RA thrombus (Tro), LA=left atrium, IAS=interatrial septum
IN CONCLUSION:
In conditions where clotting tendency is increased such as pregnancy and COPD where dilation occurs in the right cardiac spaces, presence of embryonic remnants (e.g. EV) in the right atrium might predispose to thrombus formation by behaving as a mechanic barrier. Concomitance of EV and thrombus in the right atrium may also increase the risk for pulmonary embolism. We believe that larger case studies are needed to confirm this association and to determine the predispositional role of EV in the development of right atrial thrombus and pulmonary embolism.

References: