Case Report

Protein C deficiency

Biatrial thrombus presentation

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ABSTRACT

Protein C deficiency is an inherited thrombophilia presented in adults with venous thrombosis at different sites. Symptomatic biatrial thrombus presentation of protein C deficiency has not, to my knowledge, been described. This report investigates a man with protein C deficiency who presented with dyspnea and recurrent attacks of dizziness associated with biatrial thrombus. Complete disappearance of the symptoms and thrombi was observed within less than 3 weeks of anticoagulation.

Keywords: Atrial thrombus, protein C, echocardiogram.

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Protein C is a vitamin k-dependent protein synthesized in the liver. It acts as anticoagulant after activation to the serin proteinases which inactivate factors Va, V111a which are required for thrombin formation and factor X activation.[1,2] Protein C deficiency is one type of thrombophilia inherited as autosomal dominant. The overall prevalence has been estimated to average between 1 per 200 to one per 500 in healthy population.[3-5] The main presentation in the adult is venous thromboembolism, which is estimated to be present in 2%-5% of these patients.[6,7] The most common sites are deep veins of the leg, iliofemoral and mesenteric veins. Approximately 60% of affected individuals develop recurrent thrombosis and 40% have signs of pulmonary embolism.[8]

Case Report. A 17-year-old male, student, complaining of recurrent attacks of dizziness and easy fatigability of 2 weeks duration. No chest pain, orthopnea, paroxysmal nocturnal dyspnea, palpitation, cough, hemoptysis, fever, arthralgia, weight loss or limb weakness were reported. Past medical history was negative. Physical examination revealed a good general condition, stable vital signs, normal carotid and jugular venous pressure. The apex beat was normal, no parasternal lift, and no precordial thrill. Cardiac auscultation revealed variables first heart sound intensity, intermittent diastolic plop sounds over mitral area, and no murmurs. The physical examination was negative for lymphadenopathy, hepatosplenomegaly, finger clubbing, splinter hemorrhages, and peripheral pulse deficit. Laboratory investigation revealed normal complete blood count (hemoglobulin 12.5 g/dl, white blood cells 8000, platelet 150,000, and erythrocyte sedimentation rate 12mm/hr), biochemistry, and urinalysis. Echocardiogram (ECG) and chest x-ray were normal. Transthoracic ECG shows biatrial masses protruded intermittently through mitral and tricuspid valves, no valvular incompetence, good left ventricular function.

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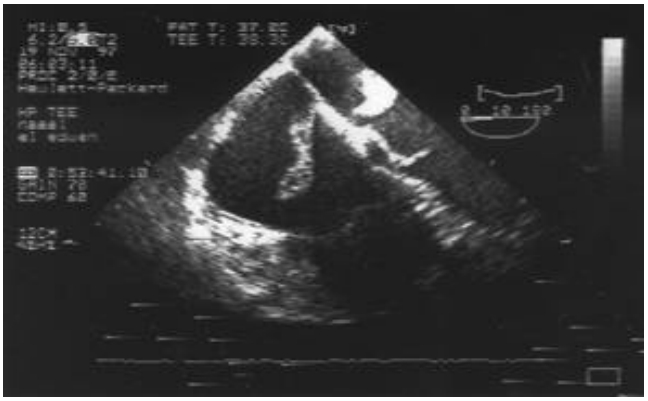

Figure 1 - Transesophageal echocardiogram, shows pedunculated biatrial large masses floating freely in both atrium with intermittent protrusion through mitral and tricuspid valves attached to the interatrial septum at area of fossa ovali, no other masses, left atrial appendage is clear, no valvular incompetence, and good left ventricular function.

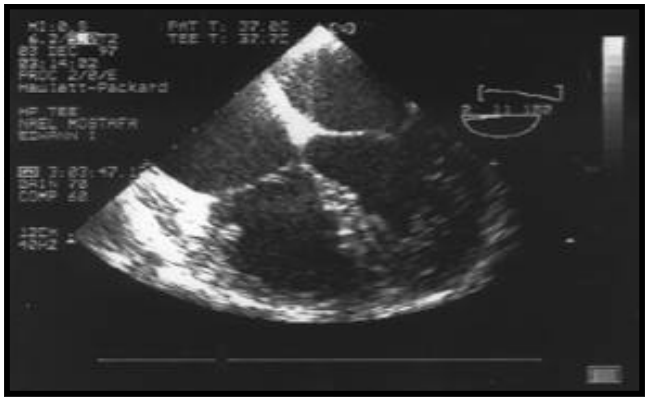

Figure 2 - Eighteen days later transesophageal echocardiogram, showed complete disapperance of biatrial masses with no evidence of patent foramen ovale and normal mitral and tricuspid valves.

Transesophageal ECG, at presentation showed pedunculated biatrial large masses floating freely in both atrium with intermittent protrusion through mitral and tricuspid valves attached to the interatrial septum at area of fossa ovali, no other masses, left atrial appendage is clear, no valvular incompetence, and good left ventricular function **(Figure 1)**. Lower limb ultrasound was negative for deep veins or arterial thrombosis. The differential diagnosis was in favor of biatrial thrombus more than myxomas. Thrombophilia screening was carried out before any treatment and cardiac surgical consultation was arranged. The patient was unable to undergo the operation at that time, and started on heparin and warfarin to keep international normalized ratio (INR) 2.5-3. One week later thrombophilia screening result showed normal antithrombin 111, protein S level, and protein C level of 30% (normal value 60-120%). Sixteen days later the patient reported one attack of severe right leg pain lasting for a few minutes resolving spontaneously. Eighteen days later the patient was asymptomatic, no dizziness, dyspnea, no history of neurological deficit, and on examination all peripheral pulses were intact, and normal heart auscultation. Eighteen days later transesophageal ECG showed complete disappearance of biatrial masses with no evidence of patent foramen ovale and normal mitral and tricuspid valves **(Figure 2)**. Repeated lower limb ultrasound was normal. Patient was doing well and followed in outpatient clinic over the last 30-months on warfarin to keep INR 2.5-3 with no complaints. Family screening for protein C level was advised.

Discussion. This case represents, for the first time, the presentation of protein C deficiency as biatrial thrombus that completely disappeared within less than 3-weeks of anticoagulation. In the literature, intracardiac thrombus caused by protein C deficiency has not been reported. Atrial thrombus may be seen in different clinical situations in the right or the left atrium. Right atrial thrombus mostly originates from venous thromboemboli trapped in the right atrium.[8-11] In situ thrombus is found in conditions associated with blood stasis as right atrial dilatation[12,13] atrial arrhythmias, cardiomyopathy, or both[12-14] or foreign bodies in right atrium as central veinous catheter,[10,15,16] transveinous electrodes[17,18] and swan-ganz catheter.[19] Left atrial thrombus is associated with blood stasis in the left atrium as atrial fibrillation, mitral valve disease and low cardiac output.[20] Occasionally free floating ball thrombi in the atrium can intermittently be seen to drift into atrioventricular orifices during diastole completely or partially obstructing flow across the atrioventricular valve.

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