CASE REPORT

A Rare Case of Left Atrial Mass with Nonhemorrhagic Infarct in a Patient of Inherited Thrombophilia

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Abstract

Inherited thrombophilias are a group of inheritable conditions that predispose individuals to venous or arterial thrombosis like deficiencies of the endogenous anticoagulants protein S, protein C, antithrombin, etc. Protein S deficiency is associated with a well-documented risk of venous thromboembolism but arterial thrombotic and thromboembolic events are very rare. Intracardiac masses are mass lesions present in either ventricles or atrium and include benign or malignant cardiac tumors, thrombus, or vegetation as well as infectious and nonbacterial thrombotic or marantic endocarditis. Cardioembolic stroke has been reported previously in patients with intracardiac masses like atrial myxoma, cardiac sarcoma, mitral stenosis with atrial fibrillation but very rare in Protein S deficiency. We report a case of embolic ischemic stroke due to intracardiac mass in a known case of protein S deficiency.

Keywords: Cardioembolic stroke, Intracardiac mass, Nonhemorrhagic infarct, Protein S.

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Case Description

A 23-year-old nonalcoholic, nonsmoker male presented with swelling and pain in the right lower limb while walking in 2012. Patient gave no history of trauma or prolonged bed rest. Color Doppler showed right-sided lower limb deep vein thrombosis and protein C activity 92.6% (70–140), factor V activity 111% (70–120), and protein S activity 28.0% (60–140). He was diagnosed with protein S deficiency after ruling out other causes and put on warfarin with regular monitoring but had poor compliance. In 2020 he was presented in the emergency ward with acute severe pain abdomen, fever, and generalized weakness. Ultrasonography whole abdomen showed well-defined hypoechoic areas showing internal echoes in region of bilateral adrenal glands measuring approximately 3.6 × 3.4 cm on right side and 3.2 × 2.8 cm on left side s/o hematoma. CT angiography abdomen with CECT abdomen showed bilaterally enlarged adrenals with increased stranding in periadrenal retroperitoneal fat s/o subacute adrenal hemorrhage with acute adrenal infective etiology. He was shifted from warfarin to dabigatran 150 mg twice daily and oral steroid replacement therapy. The patient reported again in July 2021 with weakness of left upper and lower limb since last one day. Patient was conscious, oriented to time, place, and person with blood pressure 150/90 mm Hg, pulse 96 bpm regular, SpO_2 96%, and afebrile. On examination increased tone and power 4/5 were found in the left lower limb with bilateral extensor plantar response and intact higher mental functions. 3D MRI brain revealed multiple scattered hyperintense lesions in the right parietal region in parasagittal location and corpus callosum on right side on T2W and DWI with restriction on ADC maps and appearing as isointense on T1W images, largest lesions measuring approx. 1.4 × 1.2 cm suggestive of acute nonhemorrhagic infarct in the right parietal region (Figs 1A to C). Transthoracic echocardiography showed a small irregular mass on free edge of posterior mitral leaflet measuring 2.1 × 1.6 cm (Fig. 2), dilated left atrium, left ventricle global hypokinesia, global left ventricular ejection fraction 40%, and concentric left ventricular hypertrophy. Timed blood cultures were negative. Carotid Doppler, complete blood count, liver function tests, renal function tests, and thyroid function tests were within normal range. Procalcitonin levels 0.172 ng/mL, INR 1.07, ESR 32 mm/hour, CRP 6.0 mg/L, and homocysteine levels were 12.36 µmol/L. Beta 2 glycoprotein 1 IgG 16.72 SGU (20.00), beta 2 glycoprotein1 IgM 9.57 SMU (20.00), cardiolipin antibody IgG 16.48 GPL (15.00), cardiolipin antibody IgM 18.57 MPL (12.50), lupus anticoagulant 161 second (37 second) which was false positive. The patient was managed conservatively with dual antiplatelet, dabigatran, and supportive care. During the hospital stay, patient sensorium dropped and review MRI showed progression of infarct to the left parietal region as well. The patient was referred for cardiac intervention.

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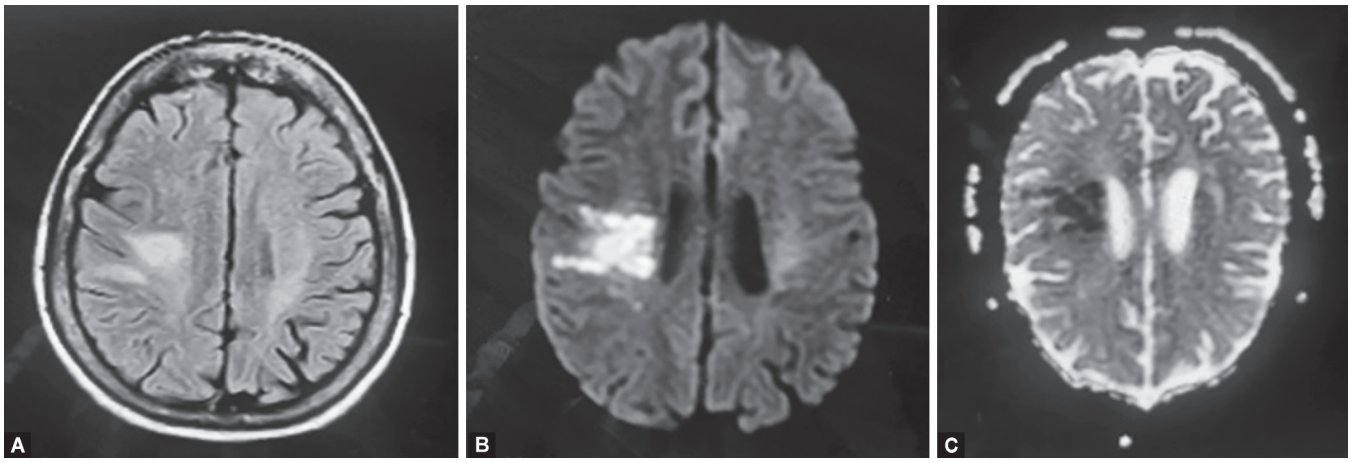
Discussion

Inherited thrombophilias are inheritable blood coagulation disorder that increases the risk of thrombosis. Protein S is a vitamin K-dependent glycoprotein that acts as a cofactor for protein C and phospholipids to inactivate factors Va and VIIIa and likely due to a defect in the *PROS1* gene.[1] Both protein C and S deficiencies are autosomal-dominant genetic disorders, usually manifesting as venous thromboembolism with deep and superficial vein thrombosis, pulmonary embolism but rarely associated with an arterial thrombosis as well as embolic or thrombotic



Figs 1A to C: Noncontrast 3D MRI showing hyperintense lesions in the right parietal region on (A) T2W, (B) DWI with restriction on ADC, and (C) Suggestive of acute nonhemorrhagic infarct in the right parietal region

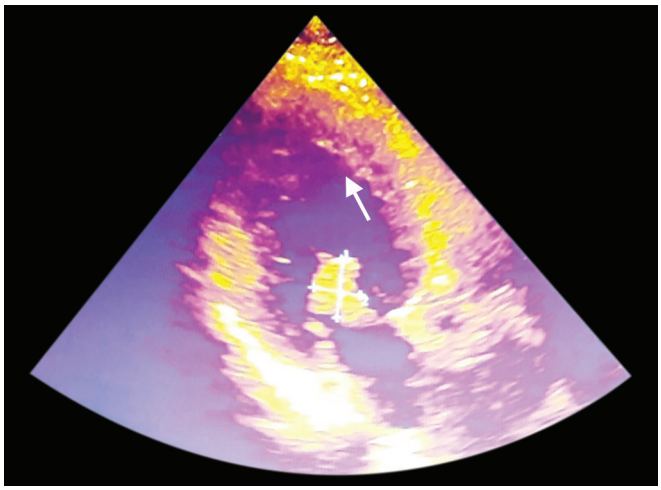


Fig. 2: Transthoracic echocardiography showing a small irregular mass on free edge of posterior mitral leaflet measuring 2.1 × 1.6 cm

nonhemorrhagic stroke. Protein C and S deficiencies have also been linked to adverse pregnancy outcomes like fetal loss, preeclampsia, intrauterine growth retardation.[2] It is primarily a hereditary disorder but age of onset is different in homozygous or heterozygous state and present mostly in three or four decades.

When our patient presented with young stroke, firstly thrombotic stroke was suspected due to preexisting thrombophilic condition. The combined deficiency of protein S and C leading to middle cerebral artery infarct was reported by Patel et al.[3] Moreover, Pantam et al. reported left posterior cerebral artery thrombosis with protein S deficiency.[4] But multiple hyperintensities raised suspicion of embolic stroke. Transthoracic echocardiography revealed left atrial mass on posterior mitral leaflet. Asymptomatic type B right atrial thrombus in a case with protein S deficiency was reported by Rawat et al.[5] Multiple thrombi in right atrium, right ventricle, and inferior vena cava in a patient with dysfunction of protein S was reported by Ok et al.[6] The most common underlying pathologic predisposition to atrial thrombus is atrial dilatation, decreased cardiac output, intracardiac catheter insertion, peripheral deep vein thromboembolism, valvular abnormalities, atrial fibrillation, malignant tumor, amyloidosis, and nephritic syndrome, etc. Pahuja et al. had reported large left ventricular thrombus with systemic and venous thromboembolism in combined protein S and C deficiency.[7] The index patient also had left atrial dilatation with left ventricular hypokinesia which makes him more prone to develop intracardiac thrombus along with protein S deficiency which he was already having. Exact treatment guidelines for the management of atrial mass in inherited thrombophilias are not standardized. The use of thrombolysis was very controversial due to the past history of adrenal hemorrhage, large infarct, and moreover, possibility of atrial myxoma could not be ruled out.

Take-home Message

Protein S deficiency is a well-known predisposing factor for venous thrombosis but should be kept as a differential diagnosis for arterial thrombosis too. Although exact mechanism and its role in the formation of intracardiac mass or thrombus are not known.

Left atrial mass presenting with embolic ischemic stroke in patients with protein S deficiency is extremely rare. The management of such patients with a history of life-threatening hemorrhage, like adrenal hemorrhage in our patient, makes it more difficult and systemic relapsing thromboses can be fatal.

Direct oral anticoagulants (DOACs) efficacy is not well studied in recurrent venous or arterial thromboembolism due to thrombophilia such as protein S deficiency and requires further evaluation.

Patients with protein S deficiency should undergo regular yearly echocardiography examination to rule out thrombus in the heart in order to prevent life-threatening embolism.

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