中文題目：肺部黏膜組織淋巴瘤併S蛋白缺乏症以免疫性血小板低下紫斑症與肺栓塞為起初臨床表現：一病例報告
英文題目：Protein S deficiency and pulmonary maltoma with initial presentation of immune thrombocytopenia, deep vein thrombosis, and pulmonary embolism
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Case Report:

A 78-year-old man with a history of idiopathic thrombocytopenic purpura (ITP) was admitted to our emergency department because of hemoptysis.

Two years before presentation, thrombocytopenia was noted at blood test before cholecystectomy for gall stones. Thrombocytopenia persist for two years and bone marrow study later revealed adequate megakaryocyte. He was treated with prednisolone and azathioprine, thrombocytopenia slightly improved and around 20000~30000/ul.

Five days before presentation, hemoptysis occurred. Blood-tinged sputum was cough out intermittently. He visited chest clinic at NTUH. The patient reported mild dyspnea and right ankle swelling but no fever, rhinorrhea, sore throat, night sweats or body weight loss. Chest radiography revealed bilateral basal and LUL consolidation. He was referred to emergency due to suspect of pneumonia.

Trace back his family history, his mother had an ischemic stroke at 50 years. His mother had bed ridden for 5 years at home before expired. There were no other thrombotic event in his other family members.

Hemogram showed thrombocytopenia (43000/ul). Chest CT scan showed consolidative patches in bilateral lungs. Right pulmonary artery embolism and aortic wall mural thrombus were incidentally noted. Three sets of sputum stain for Mycobacteria showed no acid-fast bacillus. Vascular duplex sonography confirmed the diagnosis of right lower limb deep vein thrombosis. He has thrombocytopenia but pulmonary embolism, deep vein thrombosis concurrently. He was treated with Eltrombopag 25mg qd and platelet count gradually increased to 80000~100000/ul. Low molecular weight heparin (Enoxaparin) was administered for pulmonary embolism and deep vein thrombosis after hemoptysis improved and was bridged to oral warfarin gradually.

Blood tests for the etiology of thrombosis were performed, including antiphospholipid, anticardiolipin, and β2-glycoprotein antibodies were reportedly negative, as were antinuclear antibody, rheumatic faster, C3 and C4; the blood level of protein S was reportedly low 33%, especially compared with protein C. Serum IgA/IgM/IgG were checked and IgA was reported monoclonal gammopathy.

Bronchoscope was performed after thrombocytopenia improved and biopsy over left apical-posterior bronchus (LB1/2) and left superior lingual bronchus (LB4). The pathology report revealed B-cell lymphoma, compatible with extra-nodal marginal zone lymphoma. He was alive and well, follow up at NTUH clinic.

Discussion:

We presented a case of ITP with venous thromboembolism (VTE), who was later be diagnosed with protein S deficiency and extra-nodal marginal zone lymphoma of lung.

We checked his sibling, son and daughter. We found her daughter also had mild protein S deficiency and confirmed congenital protein S deficiency.

A risk factor for thrombosis can now be identified in over 80 percent of patients with venous thrombosis. Furthermore, there is often more than one factor at play in a given patient. In our patient, ITP, protein S deficiency and underlying malignancy all may contribute to his VTE events. Recent studies have demonstrated that patients with ITP have an increased risk of thrombosis. These studies provide evidence that the incidence of thromboembolism is increased in patients with ITP, even in the presence of very low platelet counts.

The management of thrombosis in thrombocytopenic patients is challenging and not addressed by current guidelines. Likewise, there are no evidence-based data on which to draw from for recommendations. Treatment should be personalized in each patient and close monitoring of hemorrhagic event is important.