中文題目：一位反覆流鼻血但是血小板 PT/aPTT 正常的病患

英文題目：A case of recurrent nasal bleeding with normal platelet count and PT/aPTT

作　　者：馬冠鈞[1]，張鐳王欝[4]，林子皓[3]，紀韶軒[3]，戴啟明[1]，蘇裕傑[2,3]

服務單位：[1]義大醫院內科部，[2]義大醫院內科部血液腫瘤科，[3]義守大學醫學系，[4]義大醫院藥劑部

Introduction: Nasal bleeding (epistaxis) is a common clinical condition. Most episodes are minor in nature and cause by local factors. Systemic factors like renal failure, alcoholism, and vascular abnormalities, coagulopathies should also be considered in these patients, especially refractory epistaxis.

Case presentation: Herein, we report a 61-year-old male presenting as refractory recurrent epistaxis. The epistaxis was spontaneous without trauma history. Initial examination with endoscope examination showed no gross tumor but blood oozing around posterior ends of the inferior and middle turbinate in each nasal cavity. The physical examination found no hepatosplenomegaly. The laboratory data showed mild anemia, normal white blood cell (WBC) count (5,430/μl), normal platelet count and PT/aPTT. Lab data of renal and liver functions were within normal range. Initial management with blood transfusion and oral tranexamic acid were offered for hemodynamic stabilization when he was in the emergency department. Endoscopic sphenopalatine artery ligation was then performed. The intensity of the epistaxis episode improved. However, the patient presented to our emergency department with severe headache and sudden diminution of vision in both eyes one week later after discharge. Contrast enhanced MRI of nose and paranasal sinus revealed no abnormality. Ophthalmoscopic examination found bilateral multiple retinal hemorrhages and dilated, segmented, and tortuous retinal vein. Based on fluorescein angiography, a diagnosis of bilateral nonischemic central retinal vein occlusion (CRVO) was made. Further PB smear showed rouleaux formation and many lymphoplasmacytic cells were found. A bone marrow biopsy revealed a hypercellular bone marrow with diffuse infiltration of little small lymphocytes admixed with predominantly variable numbers of plasma cells and plasmacytoid lymphocytes. So hyerviscosity syndrome was suspected and protein electrophoresis showed monoclonal gammopathy (IgM-kappa). Further surveys were done and the patient was diagnosed with Waldenström's macroglobulinemia. Then further chemotherapy was arranged. The immunoglobulin M level decreased from 11,729.1 mg/dl to

1,544.80 mg/dl (normal range 57-288 mg/dl). After these treatments, the epistaxis and loss of vision resolved gradually. Patient then received regular follow up at out-patient-department.

Discussion: Bleeding diathesis refers to the increased tendency to bleed or bruise. Bleeding diathesis may result from acquired or congenital causes, most of which result in an impaired clotting process as vascular defect, platelet defect and coagulation factors defect. Hyperviscosity is a rare cause of bleeding and should be considered in differentiate diagnosis. Hyperviscosity may arise from the presence of high-molecular weight protein (like monoclonal IgM), which leads to the increase of osmotic pressure and resistance to blood flow. Furthermore, the presence of the IgM component in the blood may interact with several circulating proteins such as coagulation factors and platelets. Consequently, hyperviscosity can cause prolongation of the bleeding and clotting times. We present this case and will also describe how to make the differentiate diagnosis of bleeding tendency in clinical setting.

Conclusion: The most common cause of hyperviscosity syndrome is Hypergammaglobulinemia. Waldenström's macroglobulinemia results in monoclonal hypergammaglobulinemia, which leads to increasing of serum viscosity. It should be considered as a cause of recurrent nasal bleeding and be included in the differential diagnosis. Accurate diagnosis and early intervention of this disease is crucial.