Renal Vein Thrombosis in Vasculitis

It is largely known that renal vein thrombosis is most commonly seen with either malignancy or the nephrotic syndrome. The entity of renal vein thrombosis associated with vasculitis was reported as early as 1969 by Beard ME and Taylor DJ in a case with polyarteritis nodosa and Henoch Schonlein purpura. They hypothesized that obliteration of the arterioles that supply the renal veins was responsible for the same.[1] Although the link of venous thrombosis with vasculitis is a known entity, the association with renal vein thrombosis is rare.

The WeCLOT study, a prospective observational study of patients with Wegener's granulomatosis showed an incidence of VTE of 7 per 100 patient years, which is significantly higher than that described for normal population cohorts or for patients with rheumatoid arthritis or systemic lupus erythematosus.[2] Wlodek *et al*. reported a case with bilateral renal vein thrombosis in ANCA associated vasculitis in a male patient.[3]

A novel contributing mechanism to VTE has been demonstrated in AAV patients, which together with the increased endothelial activation could increase the risk of VTE. Antibodies against plasminogen antibodies and tissue plasminogen activator antibodies have been found in the plasma of AAV patients and cause functional inhibition of fibrinolysis in vitro.[4] Besides, elevated levels of Endogenous Thrombin Generation Potential (ETP) and Factor VIII levels as compared to age and sex-matched healthy individuals, have been reported in patients with AAV in remission, rendering them to a more procoagulant state. High ETP values have been found in both anti-MPO AAV patients and anti-PR3 AAV patients.[5]

It is not clear if AAV itself predisposes to the development of the APLS or if AAV develops as a result of APLS. However, a very small proportion of APLS patients have AAV, so, the former seems more likely.[3] In one single-center series of 218 patients with renal vein thrombosis, 143 had malignancy and 43 had nephrotic syndrome.[6] Only 36 patients in this study underwent thrombophilia screening, making conclusions on a link between renal vein thrombosis and LA or aCL is difficult.

The two cases discussed in the article published show that this is indeed a rare presentation of vasculitis with very few cases reported in the past.[7] Both anti-PR3 and MPO are related to this thrombotic disorder. Albeit postulates of pathogenesis, the true mechanism for the thrombotic tendency is largely enigmatic. However, one must look for the presence of a concomitant thrombotic disorder in a patient with renal vein involvement with vasculitis. Treating the vasculitic disorder with immunosuppressants and anticoagulation for the thrombotic state is generally rewarding.

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