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VEXAS SYNDROME AS A NEW SEVERE THROMBOPHILIC CLONAL CONDITION: A SINGLE CENTRE EXPERIENCE

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Introduction. VEXAS syndrome is an autoinflammatory condition first described in 2020. Reported VTE incidence is from 35 to 57%. Little is known about the best anticoagulant duration, intensity and risk of recurrence. [b]Aim[/b] To describe the incidence of VTE and its characteristics in patients with VEXAS followed at our Institution.
Materials and Methods. All patients had UBA1 mutation and clinical features of VEXAS. We analyzed data on superficial vein thrombosis (SVT) and VTE defined as pulmonary embolism (PE) or deep vein thrombosis (DVT).
Results. Seven male patients have been followed. One patient deceased, with no thrombotic events; two patients, in remission after bone marrow transplant, have no history of thrombosis. 4 patients experienced thrombotic events. Case 1. Age 69, in 2017 DVT of internal jugular was associated to piriform sinus abscess; warfarin was started with INR range 2-3. In 2018 during INR in range, SVT and DVT of the right leg occurred; INR range was increased to 2.5-3.5. VEXAS was confirmed in 2021. In 2022 bone marrow transplant was performed, then warfarin was stopped without recurrence. Case 2. Age 74, diagnosis of VEXAS made in 2023 due to eosinophilia, macrocytic anemia, dermatosis. Steroids were started. In 2024 due to DVT of the left popliteal and femoral vein, apixaban 5 mg bid was started indefinitely. Case 3. Age 82, in 2024 diagnosis of VEXAS due to fevers, macrocytic anemia, MGUS, pulmonary involvement; tocilizumab and steroids were started. Three months later he developed SVT and right superficial femoral DVT. Apixaban 5 mg bid is still ongoing. Case 4. Age 76, in July 2024 first SVT of the left leg treated with Fondaparinux 2.5 mg od for 60 days; in December SVT of the right arm was treated with Fondaparinux 2.5 mg od until he had a third event and was switched to Rivaroxaban 10 mg od. In April 2025 during regular anticoagulation a third SVT of the left leg and femoropopliteal DVT occurred, Dabigatran 150 mg bid was started. VEXAS was diagnosed in September 2025 due to interstitial pneumonia and macrocytosis.
Conclusions. VEXAS-associated VTE incidence at our centre is 57%, in line with literature. Our cases' clinical history highlights different presentations of thrombotic events. The anticoagulant or anti-inflammatory therapy alone may not be sufficient to protect from thrombosis and its recurrence, possibly their association may be more efficacious. Alternative treatments for long-term indications are to be investigated.