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GLOMERULAR MICROANGIOPATHY WITH MARKED SYSTEMIC THROMBOTIC MICROANGIOPATHY SHORTLY AFTER BORTEZOMIB IN A NEWLY DIAGNOSED POEMS SYNDROME PATIENT

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Abstract: The dipeptide boronic acid analogue bortezomib as a potent and selective inhibitor of the proteasome is used for the treatment of plasma cell dyscrasias such as multiple myeloma (MM). Bortezomib may induce glomerular microangiopathy (GMA) with or without systemic thrombotic microangiopathy (TMA) in which vascular endothelial growth factor-nuclear factor (VEGF) -κ B pathway could be involved. MM itself can cause TMA but primarily at presentation. **Case report:** We present a case with GMA associated with clinical features supporting systemic TMA shortly after bortezomib. **Case:** A 54-year-old woman has been diagnosed as having POEMS syndrome. She had symmetric mild degree of peripheral neuropathy, scleroatrophic skin lesions, Raynaud's phenomenon, and retinopathy. IgG kappa type paraproteinemia with a monotypic increase of plasma cells and increased pulmonary artery pressure contributed to the diagnosis. Bortezomib based treatment was started. **Methodology:** At the 20th day she developed severe dyspnea. Bilateral pleural effusion and acute kidney failure with thrombocytopenia and microangiopathic hemolytic anemia were documented. Urgent steroid and plasmapheresis were started. ADAMTS13 level proved to be within normal and plasmapheresis did not contribute to improvement. She commenced on hemodialysis and kidney biopsy was decided. Light microscopy findings revealed glomerular capillary thrombus, basement membrane thickening and segmental **Results:** duplication. Hyperplastic arteriolar changes were present. No immune deposits were detected by immunofluorescence microscopy. Biopsy findings were diagnostic for thrombotic microangiopathy. The clinical picture deteriorated as sleepiness and confusion. Cranial imaging and cerebrospinal fluid analysis showed no abnormality. Eculizumab with off-label approval contributed to stabilization but no improvement. **Conclusion:** Conclusion: Proteasome inhibitors associated with TMA may be life-threatening along with organ dysfunction due to microangiopathy-related ischemia. Membrane attack complex (C5b-9) deposition was found on endothelial cells in culture exposed to plasma from patients during the acute phase of the disease which may point to complement blockade benefit.

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PEDIATRIC HEMATOLOGY ABSTRACT CATEGORIES

COAGULATION AND FIBRINOLYSIS DISORDERS

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THE SUCCESSFUL MAJOR SURGERY IN A PATIENT WITH INHERITED FVII DEFICIENCY AND A HUGE NASOPHARYNGEAL ANGIOFIBROMA

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Objective: The bleeding phenotype of patients with inherited FVII deficiency is variable, and epistaxis is one of the most frequent symptoms. Interestingly, the bleeding risk does not correlate with the level of FVII activity. The severity of FVII deficiency and the type of surgery are not determinants of the optimal management of surgery, the doses and the duration of rFVIIa therapy are widely variable. The aim of this study is to present our successful experience in a 16-year-old boy with inherited FVII deficiency and a huge nasopharyngeal angiofibroma with a very high risk of bleeding **Case report:** The patient was referred with recurrent epistaxis in the last 6 months and he was diagnosed as an inherited FVII deficiency (FVIIC:29%, FVII inhibitor negative with positive family history). Tranexamic acid (10days) and rFVIIa (2doses) were used with success in the management of this surgery. Since this surgery may cause life-threatening bleeding, endovascular particle embolization was done to the important vessels feeding the mass one day before surgery without rFVIIa support. No bleeding or thrombosis were observed in our patient. **Conclusion:** In conclusion, a life-threatening major surgery was successfully done for a patient with inherited FVII deficiency and a huge angiofibroma. However, perioperative management of patients with FVII deficiency still remains a major challenge and clinical trials are needed to provide evidence-based optimal management of surgeries. And, angiofibroma should be thought in the differential diagnosis of epistaxis.

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