The Official Journal of the Italian Society for the Study of Hemostasis and Thrombosis
SISET
Società Italiana per lo Studio dell'Emostasi e della Trombosi
Corso Nazionale SISET
Bergamo | 23-25 ottobre 2025

29TH NATIONAL CONFERENCE OF THE ITALIAN SOCIETY FOR THE STUDY OF HEMOSTASIS AND THROMBOSIS, 2025

ABSTRACT N. PO74

COMBINED THERAPEUTIC APPROACHES IN A PATIENT WITH HEREDITARY HEMORRHAGIC TELANGIECTASIA (HHT) AND CARDIOVASCULAR COMORBIDITIES.

M. Romeo, E. Cimino , P. Conca, I.L. Calcaterra , G. Iannuzzo , C. Caputo , M. Aversano , E. Franco , S. Donnarumma , C. De Luca , M. Di Minno , A. Tufano.

Department of Clinical Medicine and Surgery, Federico II University, Naples.

Background: Hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is a rare autosomal dominant genetic disorder characterized by vascular dysplasia. The condition involves mutations in genes known as ALK1, which play critical roles in the regulation of angiogenesis and vascular integrity.

Case report: A 77-year-old female patient presented with a history of recurrent massive epistaxis, since childhood, worsening in recent years. The clinical examination revealed telangiectasias on the fingertips, tongue, lips, and sublingual mucosa, in addition to systemic AVMs involving the nasal cavity, pancreas, liver, kidneys, and duodenum. Comorbidities included severe arterial hypertension, moderate aortic stenosis, type 2 diabetes mellitus, and obesity.Previous epistaxis treatments included nasal Argon plasma coagulation (2021, 2024) and right sphenopalatine artery embolization (2021), with only temporary relief. Recent and recurrent episodes of severe anterior epistaxis resulted in progressive anemia, necessitating blood cell transfusions. Medical therapy was started with antifibrinolytics (tranexamic acid at a dosage of 10 mg/kg/dose, administered intravenously -IV-every 8 hours), and vasoactive treatment with short-acting octreotide (0.1 mg subcutaneously three times daily for the initial 15 days), followed by the administration of long-acting release octreotide (octreotide LAR 20 mg intramuscularly-IM-, every 28 days), which had not been used previously. Although the literature reports benefits from antiangiogenic agents (e.g., bevacizumab) and desmopressin in managing HHT-related bleeding, these options were deemed unsuitable in this case. Desmopressin was contraindicated due to uncontrolled hypertension, while the predominant nasal bleeding phenotype limited the potential benefit of bevacizumab, which is more effective in gastrointestinal hemorrhages. Diagnostic examinations were carried out to review the distribution of angiodysplasias: brain MRI with angiographic sequences excluded cerebral AVMs, thoraco-abdominal CT angiography confirmed the multiple abdominal angiodysplasias, none bleeding, with accessory hypodensity at the left ventricular apex and myocardial scintigraphy with pharmacological stress (technetium-99m) highlighted an area of reduced coronary reserve of approximately 10% in the mid-apical segments and the lower-lateral region of the left ventricle. Although therapy with oral antifibrinolytics (tranexamic acid 15 mg/Kg/dose/8 h) and octreotide 20 mg every 28 days, and optimization of blood pressure control, a new relapse occurred with massive posterior epistaxis associated with hematemesis, resolved with octreotide 0.5 mg in IV infusion for 24 hours and parenteral antifibrinolytics (tranexamic acid 10 mg/Kg/8 h), and blood transfusions. After a new therapeutic failure, angiography was then performed with embolization of the right and left sphenopalatine and ethmoidal arteries, complicated by subacute occipital ischemia with transient cortical blindness. The patient showed progressive clinical improvement with recovery of vision and significant reduction of hemorrhagic manifestations without the need for further transfusion support.

Conclusions: HHT is a rare and poorly recognized genetic bleeding disorder. The treatment must be individualized, considering all aspects of the condition, including local and systemic manifestations, and the patient's comorbidities.

Email: *19martyrom95@gmail.com*