The Impact of Small Bowel Endoscopy in Patients with Hereditary Hemorrhagic Telangiectasia

Herediter Hemorajik Telenjiektazi Hastalarında İnce Barsak Endoskopisinin Önemi

To the Editor,

We have read with interest the article entitled “Thalidomide for the Management of Bleeding Episodes in Patients with Hereditary Hemorrhagic Telangiectasia: Effects on Epistaxis Severity Score and Quality of Life” [1].

This article highlights the use of thalidomide in the management of patients with hereditary hemorrhagic telangiectasia (HHT) who present with epistaxis. The prevalence of HHT is thought to be between 1.5 and 2 cases per 10,000 people [2]. HHT can be associated with other bleeding complications such as bleeding from the gastrointestinal tract and in particular the small bowel (SB). The existence of small bowel angioectasias (SBAs) has been reported to vary between 56% and 91% in the literature [3,4,5,6]. The study by Ingrosso et al. [6] also reported that patients with SBAs were considerably older.

We carried out a study at our tertiary center for the management of patients with HHT where 10 patients (60% males) with genetically confirmed HHT were referred for the management of gastrointestinal-related complications. The impact of small bowel capsule endoscopy (SBCE) and double balloon enteroscopy (DBE) was evaluated. The mean age at first SB endoscopy was 62.6±14.4 years (mean ± standard deviation).

Patients had a total of 39 gastroscopies, 16 colonoscopies, and 6 push enteroscopies. Seven patients underwent SBCE: 6 (85.7%) had proximal, 1 (11.1%) had mid, and 3 (33.3%) had distal SBAs. Two patients had a colon capsule that showed angioectasias.

Several DBEs were carried out for 6 patients (median 4; SD ±6) with a mean of 130.5±133.3 days between DBEs. Fifty-seven SBAs were treated with argon plasma coagulation (APC) on average at each DBE. These procedures take an average of 75 minutes. Mean hemoglobin before and after the procedure was 9.8 and 10.2 g/dL, respectively (p=0.1). Six patients were transfusion-dependent initially but 4 improved following intervention.

Need for transfusion resolved in 1 patient when started on lanreotide (a long-acting somatostatin analog), regular endoscopy, and APC, and in 2 patients upon starting DBEs and APC. One patient passed away from pneumonia. Another patient was switched unsuccessfully from octreotide to lanreotide. She stopped being transfusion-dependent with regular gastroscopies and APC. Another patient was unwilling to undergo further endoscopies due to multiple comorbidities. He improved on lanreotide. In 2 patients, anemia remains persistently problematic. One of them is also on dalteparin for superior mesenteric venous thrombosis. The other patient has recurrent epistaxis, which makes it harder for him to have further endoscopies.

SBCE is a useful screening tool in patients with HHT to assess SBAs. Although classified as invasive endoscopy, DBEs and APC can have a significant impact on mortality and quality of life in patients with HHT. Pharmacotherapy such as somatostatin analogs can additionally help to improve transfusion requirements. They have a good safety profile [7], unlike thalidomide, which can result in teratogenicity [8], peripheral neuropathy (50%) [9], and thromboembolism [10].

Keywords: Hereditary hemorrhagic telangiectasia, Small bowel capsule endoscopy, Argon plasma coagulation

Anahtar Sözcükler: Herediter hemorajik telenjiektazi, İnce barsak kapsül endoskopisi, Argon plazma koagülasyonu

Informed Consent: Received.

Conflict of Interest: The authors of this paper have no conflicts of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included.

References

1. Baysal M, Ümit EG, Kırkızlar HO, Özdöver AC, Demir AM. Thalidomide for the management of bleeding episodes in patients with hereditary hemorrhagic telangiectasia: effects on epistaxis severity score and quality of life. Turk J Hematol 2018 (in press).
2. Dakeishi M, Shioya T, Wada Y, Shindo T, Otaka K, Manabe M, Nozaki J, Inoue S, Koizumi A. Genetic epidemiology of hereditary hemorrhagic
Our recent study aimed to investigate the association of Interleukin-2-330T/G and Interleukin-10-1082A/G genetic polymorphisms with B-Cell Non-Hodgkin Lymphoma (B-NHL) in a cohort of Egyptians. The study was designed to understand whether these polymorphisms correlate with the susceptibility to B-NHL in our population. Abdel Rahman et al. [1] concluded that the study highlights the possible involvement of the [interleukin (IL)] IL-2-330T/G genetic polymorphism in the susceptibility to [B-NHL] B-NHL in Egypt, especially indolent subtypes. Moreover, IL-10-1082A/G is not a molecular susceptibility marker for B-NHL in Egyptians [1]. In fact, the role of polymorphism of IL is widely mentioned in relationship to NHL susceptibility [2]. We agree with the observation of Abdel Rahman et al. [1]. The differences of the effects of IL-2-330T/G and IL-10-1082A/G can be explained by molecular quantum calculations of molecular weight changes. This is the same phenomenon as seen in other polymorphisms and it can affect the clinical appearance of many medical disorders, such as the effect of CTLA-4 A49G polymorphism on autoimmune blood disease [3]. For IL-2-330T/G and IL-10-1082A/G, the change of molecular weight is equal to -107.07 and +16 per molecule, respectively. This means that a molecule with IL-2-330T/G requires more molecular mass and a molecule with IL-10-1082A/G requires less molecular mass to complete a biological process compared to a naïve molecule.

Keywords: Interleukin, Lymphoma, Polymorphism

Conflict of Interest: The authors of this paper have no conflicts of interest, including specific financial interests, relationships,