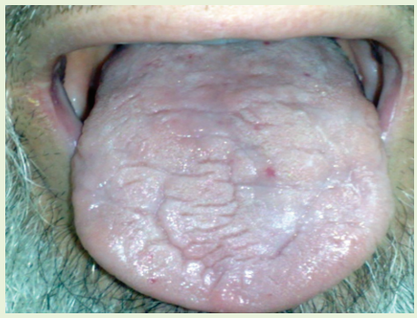
Hereditary hemorrhagic telangiectasia

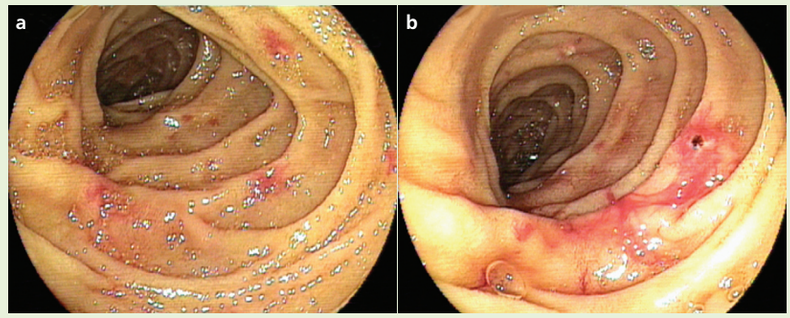
Herediter hemorajik telenjiektazi

Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome) is an autosomal dominant disorder. It is characterized by dermal, mucosal and visceral telangiectasias and recurrent bleeding symptoms. The incidence of the disease was estimated to be approximately 1 in 5000-8000. The most profound presentation is epistaxis, and this complaint was encountered in 90% of the patients. Diagnosis is based on three criteria among: epistaxis, gastrointestinal telangiectasia, visceral arteriovenous malformation, or family history of hereditary hemorrhagic telangiectasia.

Herein, we present the pictures of two patients diagnosed with hereditary hemorrhagic telangiectasia. Bevacizumab, which is a recombinant IgG1 monoclonal antibody, was used in the first patient, and argon plasma coagulation was applied to the duodenal telangiectasias in the second. In the first patient, there were multiple telangiectasias on his tongue (Figure 1). The other patient's gastroscopy revealed widespread telangiectasias in his duodenum after the bulbus (Figure 2a), and he was treated with argon plasma coagulation (Figure 2b).



Resim 1. Multiple telangiectasias are seen on the 65-year-old-patient's tongue.



Resim 2. a. Widespread telangiectasias in the duodenum before APC treatment. **b.** Duodenum image of the patient after APC application.

Fatih ERMİŞ[1], Ahmet UYANIKOĞLU[2], Filiz AKYÜZ[3], Fatih BEŞIŞIK[3]

Department of [1]Internal Medicine, Division of Gastroenterology, Düzce Medical Faculty, Düzce University, Düzce
Department of [2]Internal Medicine, Division of Gastroenterology, Harran Medical Faculty, Harran University, Şanlıurfa
Department of [3]Gastroenterology, İstanbul Medical Faculty, Istanbul University, İstanbul

YORUM:

HHT gerçekten nadir görülen fakat bir kere görüldüğünde de unutulmayacak vasküler bir patoloji. Tüm vücudu tutabilen telenjektaziler ile karakterli bu hastalıkta aile öyküsü ve tekrarlayan epistaksislerin bulunması tanıya ulaşmakta çok yardımcıdır. Gastrointestinal sistemin tutulduğu olgularda ulaşılabilen yerlerde lokal ablatif tedaviler yararlı olmaktadır. Sistemik tedavisi halen sorun olup östrojen praparatlarından da çok fayda bulunamamaktadır.

Prof. Dr. Orhan SEZGİN
Özgün Görüntüler Editörü

İletişim: Fatih ERMİŞ
Department of Internal Medicine, Division of Gastroenterology, Düzce Medical Faculty, Duzce University, Düzce, Turkey • Phone: +90 380 514 88 30
Fax: +90 0 380 542 13 02 • E-mail: fatihermis2@hotmail.com

Geliş Tarihi: 16.08.2012 • **Kabul Tarihi:** 25.08.2012