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Course of Treatment of Chronic Bleeding and Anemia with Systemic Bevacizumab Treatment in Hereditary Hemorrhagic Telangiectasia: A Retrospective Cohort

Herediter Hemorajik Telenjiyektazide Sistemik Bevasizumab Tedavisiyle Kronik Kanama ve Anemi Tedavisinin Seyri: Bir Retrospektif Kohort

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ABSTRACT Objective: Hereditary hemorrhagic telangiectasia (HHT), is an autosomal dominant disorder that causes abnormal angiogenesis. Interest in targeted therapies has been increasing in recent years, especially for the treatment of severe forms of HHT. One of these treatment options is the vascular endothelial growth factor inhibitor bevacizumab. Purpose of this study is to investigate the effect of systemic bevacizumab use on the treatment of chronic bleeding course and anemia in patients diagnosed with HHT. **Material and Methods:** The treatment response and adverse events of patients with bevacizumab were evaluated retrospectively. **Results:** The mean age was 51,5. Mean duration of treatment was 15.4 (4-25 months) months. The first 4 doses of 5 mg/kg intravenous bevacizumab were administered at 2-week intervals in all patients. Bevacizumab maintenance continued at a dose of 5 mg/kg in monthly periods. With bevacizumab treatment, an increase in hemoglobin values, a decrease in epistaxis severity score, parenteral iron and erythrocyte transfusion requirement were achieved. Side effects observed were allergic rash in one patient and arthralgia in one patient. None of the patients required discontinuation of treatment due to side effects. **Conclusion:** Bevacizumab is a promising treatment option in HHT, which can be mortal if not controlled. However, there remains a need for more comprehensive studies in order to achieve a global consensus on treatment protocols and management of adverse events.

Keywords: Bevacizumab; hereditary hemorrhagic telangiectasia; epistaxis

ÖZET Amaç: Kalıtsal hemorajik telenjiyektazi (HHT), anormal anjiyogeneze neden olan otozomal dominant bir hastalıktır. Son yıllarda, özellikle şiddetli HHT formlarının tedavisi için hedefe yönelik tedavilere olan ilgi artmaktadır. Bu tedavi seçeneklerinden biri de vasküler endotelyal büyüme faktörü inhibitörü bevasizumabtır. Bu çalışmanın amacı, HHT tanısı alan hastalarda sistemik bevasizumab kullanımının kronik kanama seyri ve anemi tedavisindeki etkisini araştırmaktır. **Gereç ve Yöntemler:** Bevasizumab kullanan hastaların tedavi yanıtı ve yan etkileri retrospektif olarak değerlendirildi. **Bulgular:** Hastaların yaş ortalaması 51,5 yıl idi. Ortalama tedavi süresi 15,4 (4-25 ay) aydı. Tüm hastalara 2 hafta arayla ilk 4 doz 5 mg/kg intravenöz bevasizumab uygulandı. Bevasizumab idamesi 5 mg/kg dozunda aylık periyotlarla devam etti. Bevasizumab tedavisi ile hemoglobin değerlerinde artış, epistaksis şiddet skorunda parenteral demir ve eritrosit transfüzyon gereksiniminde azalma sağlandı. Gözlenen yan etkiler, 1 hastada alerjik döküntü ve 1 hastada artralji idi. Yan etkiler nedeniyle hiçbir hastada tedavinin kesilmesi gerekmedi. **Sonuç:** Bevasizumab, kontrol edilmediği takdirde ölümcül olabilen HHT'de umut verici bir tedavi seçeneğidir. Bununla birlikte, tedavi protokolleri ve advers olayların yönetimi konusunda global konsensus sağlamak için daha kapsamlı çalışmalara ihtiyaç vardır.

Anahtar Kelimeler: Bevasizumab; herediter hemorajik telenjiyektazi; epistaksis

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is a common autosomal dominant disorder that causes abnormal angiogenesis. Its prevalence has been reported as 1/5,000 in North America. However, since the diagnosis is often missed and some patients may be asymptomatic, the actual prevalence of the disease is estimated to be higher.[1,2] In the pathogenesis of the

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