

HEMOLİTİK ÜREMİK SENDROM

36.

BÖLÜM

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GİRİŞ

Hemolitik üremik sendrom (HÜS), endotel hücre zedelenmesi, trombüs oluşumu ve mikroanjiopati sonucu gelişen mikroanjiyopatik hemolitik anemi, trombositopeni ve akut böbrek hasarı ile karakterize, trombotik mikroanjiopatilerin (TMA) heterojen bir grubudur ^(1,2). Trombotik mikroanjiopati, mikroanjiyopatik hemolitik anemi (MAHA), trombositopeni ve mikrovasküler tromboz ile karakterize bir grup hastalığı tanımlamak için kullanılmaktadır ⁽³⁾. Böbrekler, beyin, kas, gastrointestinal sistem (GİS) deri ve akciğerler dahil birçok organı etkileyen sistemik bir hastalıktır ⁽³⁾. Temel olay mikrosirkülasyonda fibrin ve trombosit açısından zengin trombüs oluşumuna yol açan endotel hücrelerinin patolojik hasarıdır.

Çocuklarda akut böbrek hasarının en önemli nedenlerinden biri HÜS'tür ⁽⁴⁾. Hemolitik üremik sendromun klinik bulguları trombotik mikroanjiopati sonucu oluşur. Böbrek başta olmak üzere çok sayıda doku ve organ etkilenir ⁽⁵⁾. Zemininde mikroanjiopatinin olduğu ciddi anemi, periferik kan yaymasında parçalanmış eritrositler, retikülositoz, trombositopeni ve serum laktat dehidrogenaz (LDH) düzeyinde artış görülür ⁽⁶⁾.

Çocuklarda, HÜS vakalarının %90'ı Shigatoksijenik Escherichia coli (STEC) enfeksiyonlarının neden olduğu ishal ile ilişkilidir. Bu STEC HÜS olarak bilinir ⁽⁷⁾. Geri kalan HÜS vakaları çoğunlukla atipik HÜS (aHÜS) olarak adlandırılan ve farklı mekanizmalar ile HÜS geliştiği düşünülen bir gruptur. Atipik HÜS sporadik veya ailesel olabilir. Atipik HÜS vakalarının çoğu kompleman sisteminin genetik ya da kazanılmış düzensizliklerinden kaynaklanır ⁽⁸⁾.

Klinik uygulamada yakın geçmişe kadar yaygın olarak tipik ve atipik HÜS sınıflaması kullanılmaktaydı. Son yayınlanan uluslararası uzlaşma raporunda tipik HÜS yerine STEC HÜS kullanılması önerilmektedir ⁽⁹⁾. Sonuç olarak yayınlarda

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larda özellikle aHÜS hastalıklarının etyolojisinde çok önemli gelişmeler olmuştur ve birçok genetik ya da kazanılmış etyolojik neden artık saptanabilmektedir. Ecu'nun bu hastalarda etkili olduğu görülmektedir ve kompleman ilişkili aHÜS hastalarında ilk seçenek tedavi hâline gelmiştir.

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