BÖLÜM 13
Mikroanjiyopatik Hemolitik Anemi

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Giriş

Trombositopeni, hemolitik anemi ile seyreden ve diğer sistemik bozukluklara yol açabilen erken teşhis ve tedavisi hayati öneme sahip hastalık grubunun genel adıdır mikroanjiopatik hemolitik anemi (MAHA). MAHA ve trombotik mikroanjiopati (TMA) sıklıkla birbirlerinin yerine kullanılsalar da tam olarak birbirlerini karşılayan ifadeler değillerdir.

Tanım

Mikroanjiopatik hemolitik anemi (MAHA): intravasküler alanda kırmızı küre yıkımına sebep olan ve şistosit oluşumu ile karakterize non-immün bir hemoliz olayıdır (şekil-1) (1). Küçük arterioller ve kılcal damarlar gibi mikrovasküler dolaşım patolojileri neden olabilmekle beraber protez kalp kapakları, intravasküler cihazlar gibi nedenlerde MAHA'ya neden olabilmektedir.

Trombotik mikroanjiyopati (TMA): sıklıkla MAHA ile aynı klinik durumu tanımlamak için kullanılsalar da tüm TMA'lar MAHA'ya neden olsa da tersi durum doğru değildir. TMA tanım olarak arteriyoler ve/veya kapiller damar duvarındaki anormalliklerin mikrovasküler tromboza yol açtığı özel bir patolojik lezyonu tanımlamaktadır (2).

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Renal Transplantasyon

Eculizumab tedavisine rağmen son dönem böbrek yetmeliği gelişen hastalarda renal transplantasyon düşünülebilir. Ancak canlı donör tercih edilmez çünkü alınan böbrekte C-TMA açısından risk altındadır.

CFH, CFI veya C3 gen mutasyonu olan hastalarda nakil sonuçları başarısızdır. %50 hastada rekürrens gelişirken bunların %90'ında graft kaybı gelişir. MCP muasyonunda veriler kısıtlı olmasına karşın başarı şansı daha yüksektir. CFH'a karşı otoantikor olan aHÜS vakalarında otoantikor eradikasyonu sonrasında nakil yapılmalıdır. DGKE mutasyonlu çocuklarda nakil sonrasında rekürrens gözlenmez. CFH, CFI veya C3 gen mutasyonu olan hastalarda karaciğer-böbrek kombine transplantasyonu ile kür sağlanabilir ancak veriler oldukça sınırlıdır.

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