Bölüm 26

POLİANJİTİS İLE BİRLİKTE EOZİNOFİLİK GRANÜLOMATOZİS (EGPA) VE KARDİYAK TUTULUM

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GİRİŞ

Polianjitis ile birlikte Eozinofilik Granülamatozis (EGPA), eski adıyla Churg-Strauss sendromu, başta solunum sistemi olmak üzere birden fazla sistemi tutan ve küçük ve orta çaplı damarların nekrotizan granülomatöz vasküliti ile karakterize bir hastalıktır(1). Kronik rinosinüzit ve/veya nazal polipozis, astım ve belirgin periferik kan eozinofilisi birlikteliği görülür. Kanda ve dokuda eozinofiller ön plandadır. EGPA'nın majör histopatolojik bulguları; belirgin eozinofilik infiltrasyon ve yaygın nekrozdur.(1). Hastaların hem serum hem de dokularında Th2 tipi yanıtın göstergesi olan sitokinler (IL-4, IL-5 ve IL-13 gibi) yüksek saptanır(2, 3).

Patogenez

EGPA'nın kesin patogenezi bilinmemektedir. Anormal immün fonksiyon ve genetiğin rol oynadığı bildirilmiştir. Hastaların yaklaşık yarısında Anti-nötrofil sitoplazmik antikorlar (ANCA) tespit edilir. Bu nedenle EGPA, ANCA ilişkili vaskülitler içinde sınıflanır. Renal tutulumu olmayan EGPA'lı hastaların sadece yaklaşık %25'i ANCA pozitif iken, bir derece renal tutulumu olanların %75'i ve kanıtlanmış nekrotizan glomerülonefriti olanların %100'ü ANCA pozitifliği gösterir(4).

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