

Bölüm 20

DEV HÜCRELİ MİYOKARDİT

Muzaffer KAHYAOĞLU¹

Dev hücreli miyokardit (DHM) ilk defa 1905 yılında Saltykow tarafından akut kalp yetmezliği kliniği ile başvuran genç bir erkek hastada tanımlanmış olup nadir görülen potansiyel olarak fatal seyreden bir hastalıktır (1). Sıklıkla genç-orta yaş yetişkinleri etkilemektedir (2). DHM tanısı; spesifik olmayan klinik görünümü, diğer kardiyovasküler hastalıklarla potansiyel örtüşmesi ve bu antite hakkında genel bir farkındalık eksikliği nedeniyle sıklıkla zor olabilmektedir. DHM'nin patogeneğinde özellikle miyokardın T lenfosit aracılı inflamasyonu suçlanmaktadır (2). Tedavide immünesupresif tedaviler başarılı olarak kullanılana kadar tüm olgular fatal seyretmekte ya da kalp nakli ihtiyacı olmakta iken günümüzde artan farkındalık ve erken tanı avantajı, immünesupresif tedaviler, gelişmiş hemodinamik destek ve gerektiğinde kalp nakli seçenekleri ile yönetilebilir bir hastalık olarak karşımıza çıkmaktadır.

Otopsi serilerine göre insidansı %0.007-0.051 olarak bildirilmektedir (3). Yapılan çalışmalarda etkilenen hastaların ortalama yaşı 42,6-60 yaş aralığında tespit edilmiştir ve cinsiyet predominansı gözlenmemektedir (2,4). Pediyatrik vakalar oldukça azdır. Çok merkezli DHM kayıtlarının sonucuna göre pediyatrik olgular tüm olguların yaklaşık % 6'sını oluşturmaktadır (5,6). Pediyatrik vakaların erişkinlerden önemli farkı hastalık seyrinin daha agresif olmasıdır (5,6).

¹ Uzm. Dr. İstanbul Beykoz Devlet Hastanesi, Kardiyoloji Kliniği, mkahyaoglu88@gmail.com

Kardiyak transplantasyon sonrası tekrarlayan DHM hastaların %20 ila %25'inde görülür (39). Hastalar kalp yetersizliği veya başka semptomlarla başvurabilse de, çoğunluğu asemptomatiktir ve tipik olarak EMB sonuçlarına göre saptanır. Şu anda, asemptomatik hastalarda kalp nakli sonrası rutin EMB taramasına ilişkin bir kılavuz bulunmamaktadır; ancak yeni başlayan kalp bloğu, ventriküler aritmiler veya SV sistolik disfonksiyonu gelişen olan hastalarda EMB'yi düşünmek mantıklıdır. Normal SV işlevine sahip asemptomatik tekrarlayan DHM'nin tedavisi, pulse steroid tedavisidir. Yüksek doz kortikosteroidler ve ATG, tekrarlayan DHM ve SV disfonksiyonu olan hastalarda sıklıkla birinci basamak tedavi olarak kullanılmaktadır. Sirolimus ve rituksimab başarıyla kullanılmış olsa da (40,41), alemtuzumab, T hücreleri üzerindeki CD52 aracılı etkisi nedeniyle refrakter vakalarda daha uygun olabilir (38).

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