BÖLÜM 34

Skleroderma ve Kalp

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GİRİŞ

Skleroderma olarak da adlandırılan sistemik skleroz (SSc), doğal ve edinsel immün sistem bozukluğu, mikrovasküler hasar ve birden fazla organda jeneralize fibroz ile karakterize sistemik heterojen bağ dokusu hastalığıdır (1). Raynaud fenomeni çoğunlukla hastalığın ilk bulgusu olmak ile beraber akciğer fibrozu, skleroderma renal krizi ve kardiyak komplikasyonlar gibi birçok organın tutulumu hastalık süresinin erken dönemlerinde ortaya çıkabilir (2). SSc'de kalp tutulumu ilk olarak 1926'da SSc'li bir hastanın otopsisinde koroner arterler, perikard ve miyokardda patolojik değişiklikler ile Heine tarafından tanımlandı (3). Kalp, çeşitli semptomlar ile SSc'nin erken evrelerinde yer alan organlardan biridir. Kardiyak belirtiler SSc'de yaygındır ve tahmini klinik prevalans %15-35'tir. Bu popülasyon romatologlar ve kardiyologlar gibi farklı tıp uzmanlıkları arasında farklı kabul görmektedir, branşlar arasında bu hastalıkların farkındalığı arttırılmalıdır (4). Ancak SSc hastalarının çoğunda kardiyak belirtiler subklinik kalabilir ve bazen sadece otopside tanı konulabilirken, klinik olarak aşikar kalp tutulumu artmış mortalite oranı ile ilişkili ciddi bir negatif prognostik belirteçtir (5). Primer kardiyak tutulum SSc'de yaygındır ve perikard, miyokardiyum, endokardiyum, kardiyak kapakçıklar ve iletim sistemi dahil olmak üzere kalbin tüm yapısal bileşenlerini etkiler (Şekil-1). Anti-topoizomeraz pozitif SSc hastaları daha sık ve daha şiddetli etkilenir (6). Perikardiyal efüzyon, aritmiler, iletim sistemi defektleri, kapak bozuklukları, miyozit, miyokardiyal iskemi, miyokardiyal hipertrofi ve kalp yetmezliği ile sonuçlanabilir. Miyokardiyal belirtiler muhtemelen SSc'nin

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SSc ile ilişkili vaskülopati için yaygın olarak uygulanan vazodilatatör ilaçlar arasında, bosentanın çarpıntı, hipotansiyon ve sıvı retansiyonuna neden olduğu bildirilmiştir ve eşlik eden sol ventrikül yetmezliği olan hastalara verildiğinde dikkatli olunması gerekir. Aynı şekilde iloprost, ciddi koroner kalp hastalığı, yakın zamanda kardiyovasküler veya serebrovasküler tromboembolik olayları, konjestif kalp yetmezliği veya şiddetli aritmileri olan hastalarda bu durumları şiddetlendirebileceği için kontrendikedir. Son olarak, kalsiyum kanal blokerleri kararsız anjina ve yakın zamanda geçirilmiş miyokard enfarktüsünde kontrendikedir. Bu ilaçların bildirilen kardiyak yan etkileri taşikardi, çarpıntı, hipotansiyon ve senkoptur (59).

SONUÇ

Kardiyak komplikasyonlar, SSc'de önemli bir rol oynar ve özellikle yaygın kutanöz SSc'de ani kardiyak ölüm riski taşır. Bu nedenle, miyokardiyal ve vasküler tutulumun erken saptanması ve izlenmesi, SSc hastalarının yönetiminde çok önemlidir. Romatologlar ve kardiyologlar , SSc'de primer kardiyak tutulumun farklı formlarının farkında olmalıdır ve erken tanı ve tedavi hasta prognozunda önem taşımaktadır.

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