BÖLÜM 110

# KARACİĞER KİSTİK HASTALIKLARI

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## GİRİŞ

Karaciğer kistleri, tek bir hücre tabakasıyla kaplı içi sıvı dolu lezyonlar olarak tanımlanır. Kistlerin patogenezinde anormal fetal duktal plak olgunlaşması yatar. Kistteki epitel hücreleri salgı fonksiyonunu korur ve sıvı üretimleri, kistin yapısına katkıda bulunan pozitif bir luminal basınç oluşturur. Kist sıvısı su ve elektrolitlerden oluşur ve bileşimi safraya benzer ancak safra asitleri veya bilirubinden yoksundur. Fibrokistik karaciğer hastalıklarını; soliter hepatik kistler, izole polikistik karaciğer hastalığın (PKH) veya otozomal dominant polikistik böbrek hastalığı (ODPBH), konjenital hepatik fibrozis (KHF), otozomal resesif polikistik böbrek hastalığı (ORPBH) ve Caroli hastalığı oluşturur (1,2).

Basit hepatik kistlerin prevalansı %2,5-18, PKH prevalansı 1/10.000-1/158.000'dir (3,4). PKH, safra kanalı sistemiyle bağlantısı olmayan 10'dan fazla hepatik parankimal kistin varlığı ile tanımlanır. Otozomal dominant bir şekilde kalıtsal olabilir ve en belirgin fenotipe bağlı olarak 2 farklı antite kategorize edilmiştir: otozomal dominant polikistik böbrek hastalığı (ODPBH) ile birlikte ve otozomal dominant polikistik karaciğer hastalığı (ODPKH) (5). Caroli hastalığı, intrahepatik safra kanallarının dilatasyonu ile karakterize edilir (6). Caroli hastalığı, daha büyük intrahepatik safra kanallarının dilatasyonu ile sınırlıyken, Caroli sendromu, küçük safra kanalı dilatasyonu ve konjenital hepatik fibrozisi birleştirir (7). Von Meyenburg kompleksleri olarak da adlandırılan biliyer hamartomların, duktal plak malformasyonları spektrumunun bir parçası olduğu kabul edilir (5). Safra kanalı hamartomları tipik olarak karaciğer boyunca dağılmış küçük (<1 cm) hipodens lezyonlar olarak tanınır ve ilk bakışta karaciğer metastazlarını taklit edebileceği için önemlidir.

## FİBROKİSTİK KARACİĞER HASTALIKLARININ BİYOLOJİSİ

### Duktal Plak Malformasyon Hipotezi

İnsanlarda safra ağacının oluşumu, fetal yaşamın ilk trimesterinde, portal yolların mezenkimal dokusu ile temas halindeki öncü hücrelerin glandüler morfolojiye farklılaştığı ve duktal plağı oluşturduğu zaman başlar (8,9). Fetal yaşamın ikinci trimester döneminin başlangıcında, duktal plağın

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ciğer nakli için diğer endikasyonlarla karşılaştırılabilir (160,178).

Kombine karaciğer ve böbrek nakli endikasyonu olan PKH'lı hastalarda, ardışık prosedürlere kıyasla kombine karaciğer-böbrek naklinin üstünlüğüne dair bazı görüşler vardır, ancak sağlam kanıtla dayanmamaktadır (179,180). Kombine karaciğer-böbrek naklinde, %20'ye varan kısa vadeli böbrek graft kaybı oranlarının yüksek bulunduğu için kombine nakil hakkında endişeler mevcuttur (181). Böbrek nakli fonksiyonu ve böbrek allotransplant yetmezliği, karaciğer nakliyle ilişkili majör perioperatif hemodinamik ve metabolik instabiliteye bağlanmıştır. Böbrek naklinin potansiyel yararsızlığını dengelemek için mevcut yaklaşımlar, aynı donörden bir böbreğin gecikmiş implantasyonu veya karaciğer naklinden bir süre sonra sonra metakron canlı donör böbrek naklini içerir (182).

## CAROLİ HASTLIĞI'NIN YÖNETİMİ

İntrahepatik kanalların multifokal segmental dilatasyonu hem Caroli hastalığında hem de Caroli sendromunda mevcuttur. Görüntülemede, bu safra kanalları çapı 5 cm'ye kadar büyüyebilen kesecikli veya fusiform kistik dilatasyonlar olarak görünür. Bu lezyonlar çoğunlukla intrahepatik büyük boyutlu safra kanallarında bulunur ve tip V koledok kistlerine (Todani sınıflandırmasının bir parçası) karşılık gelir. Ancak, ekstrahepatik dilatasyonlar genellikle Caroli hastalığında görülür ve tekrarlayan kolanjit ve taş geçişi ataklarından kaynaklandığı düşünülmektedir.

Hepatik fibroz, Caroli sendromunu Caroli hastalığından ayıran temel histolojik lezyondur. Caroli sendromu muhtemelen ORPBH fenotipik spektrumunun bir parçasıdır. Burada, klinik tablo konjenital hepatik fibrozun sonuçları tarafından domine edilir. Hastalıkları sırasında birçok hastada venöz portosistemik şant ve özofageal varisler ve splenomegali gelişimi ile portal hipertansiyon gelişir (183). Bu hastalarda karaciğer nakli düşünülmelidir. Caroli hastalığı ve sendromu olan hastalar, tanıdan sonra her 12 ayda bir MRCP ile kolanjiyokarsinom için takip edilmelidir (2).

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