

BÖLÜM 6

KONJENİTAL KİSTİK AKCİĞER HASTALIKLARI

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GİRİŞ

Konjenital kistik akciğer hastalıkları (KKAH) veya konjenital bronkopulmoner malformasyonları (KBPM), bronkopulmoner sistemin embriyolojik gelişimindeki bozukluğun sonucu olarak ortaya çıkar. Günümüze kadar bu hastalıkların patogenezi tam olarak belirlenememiştir. Ön bağırsaktan trakeal ayrılma ve akciğer tomurcuğundan bronşiyal dallanma sırasında embriyolojik defekt sonucu birkaç tip konjenital kistik lezyonun geliştiği düşünülmektedir. Histolojik ve moleküler tanı alanlarındaki ilerleme ile KKAH'nın ortak embriyolojik kaynaklı olduğu ama trakeobronşiyal ağacın gelişiminde zaman ve lokasyon farkından dolayı bu lezyonların farklı histolojisinin ortaya çıktığı düşünülmektedir.^{1,2} Prenatal tanı ve yaklaşımdaki gelişmeler sayesinde KKAH daha detaylı olarak değerlendirilmektedir. Yenidoğan yönetimi ve cerrahi alanda ilerlemeler ile lezyonların postnatal yönetiminde mortalite ve morbidite açısından daha iyi sonuçlar elde edilmektedir. Bu bölümde bronkopulmoner malformasyonların embriyolojisi, prenatal ve postnatal yaklaşımı, ileri dönem takip ve tedavilerinden bahsedilecektir.

BRONKOPULMONER AĞACIN VE AKCİĞERLERİN GELİŞİMİ

Akciğerin 6 gelişim aşaması mevcuttur:

1. **Embriyonik aşama** – Dördüncü haftada primitif ön bağırsak ön yüzeyinden larengotrakeal tomurcuklanma ile başlar, 7. haftaya kadar endodermal ve mezodermal yapılara sahip 2 adet primitif akciğer tomurcuğu oluşur. Yedinci haftadan itibaren lobar ve segmental hava yolu dallanması başlar. 4.-7. haftalar arasında organogenezle eş zamanlı vaskülogenez seyredir. 6. aortik arkten çıkan 2 adet pulmoner arter ileri dönemde hava yolu ve mezenkimini saran vasküler pleksus oluşturur. Bu pleksus önce ön bağırsak ve gelişen trakeanın sistemik venleri, daha sonra pulmoner venlerle birleşir. Yedinci haftanın sonunda santral vasküler, lobar ve segmental dallara sahip hava yolu yapıları gelişmiş olur.

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mevcuttur.^{169,170} Literatürde KPHM tanılı hastalarda plevropulmoner blastom, rabdomyosarkom, bronkoalveolar karsinom gelişme riski mevcuttur.¹⁷¹⁻¹⁷⁴ Çocuk ve erişkinlerin KPHM ilişkili akciğer tümör vakalarının incelemesinde malign progresyon için yaş sınırı, KPHM'in malign transformasyonu için belirli bir sınır görülmemiş, KPHM teşhisinde gecikme ile tümörün ortaya çıkışı arasında zaman kaybının ilişkisi tahmin edilememiştir.¹⁷⁵

3. **BÜYÜME - GELİŞME GERİLİĞİ:**Okul çağına kadar olan hastalarda motor fonksiyon bozuklukları görülebilir. İlk bir yılda mental gelişim ve okul çağında kognitif fonksiyonları normal olmasına rağmen, hastaların iş hızı, yetenek ve hafızalarında sorunlar yaşanabilir. Özellikle yenidoğan döneminde ECMO tedavisi hikâyesi olan hastalarda nörolojik sorunların yaşanma olasılığı yüksektir.^{176,177}

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