TORAKS ANOMALİLERİNDE PRENATAL TANI

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Anahtar Noktalar

- Prenatal ultrasonografi ile konjenital diyafragma hernisi (KDH) vakalarının 2/3 ü yakalanır. Çoğu vakada tanı ikinci üçay obstetrik ultrasonografi incelemesi ile koyulur.
- KDH vakalarının kabaca yarısında kromozom anomalileri, ek anomaliler, sendromlar, tek gen hastalıkları birlikteliği vardır.
- Prognozun kötü olduğu KDH'li vakalarda prenatal tedavinin temelini fetoskopik endoluminal trakeal oklüzyon (FETO) oluşturur.
- Prenatal tanı koyulan konjenital pulmoner solunum yolu malformasyonu (KPSM) vakalarında prognoz genellikle olumludur. Genel sağkalım oranı %80 ler civarındadır. Hidrops gelişmesi durumunda prognoz çok olumsuzdur.
- Prenatal dönemde tanı koyulan pulmoner sekestrasyonların çoğu ekstralobar tiptedir.
- Özofagus atrezisi/trakeoözofageal fistülün kesin prenatal tanısı halen güçtür. Özellikle mide cebinin küçük izlendiği veya görülemediği ve polihidramniyosun eşlik ettiği olgularda kuşkulanılmalıdır.
- Daha nadir sıklıkla izlenen bronşiyal atrezi ve bronkojenik kistlerin prenatal tanısı önemlidir. Çünkü bu olgularda postnatal prognoz, prenatal bulguların ağırlığı ile doğrudan ilişkilidir.
- Konjenital üst hava yolu tıkanıklığı (KUHTS)'da prenatal tanı görece daha kolay olsa da, genetik sendromlar ile birlikteliği nedeniyle aileye danışmanlık verilmesi için ayrı önem taşımaktadır.

Anahtar kelimeler: Prenatal tanı, göğüs, cerrahi

Giriş

Fetal toraksın prenatal dönemde ultrasonografik muayenesi 25-26. gebelik haftasına kadar rahatlıkla yapılabilir. Ancak bu haftadan sonra kostaların mineralizasyon artışı nedeniyle akustik gölgelenme oluşur, bu durumda intratorasik organların özellikle koronal ve sagital planlarda değerlendirilmesi güçleşmektedir. Bununla beraber bazı intratorasik anomaliler üçüncü üçayda ortaya çıkabileceği gibi, doğumdan önce de kaybolabilirler. Bu sebepledir ki, fetal toraks ve kalp değerlendirmesi 12-14 gibi erken gebelik haftalarında yapılmalı, gerektiğinde takip muayeneleri üçüncü üçaya kadar aralıklar ile yapılmalıdır.

Toraks konotrunkal bir forma sahiptir. Üst sınırları klavikula ve boyun, alt sınırı diyafragma, ön ve yan sınırları ise sternum ve kostalar tarafından oluşturulmaktadır. Toraksta diyafragmadan klavikulalara kadar uzanım gösteren sağ ve sol hemitoraksı dolduran *akciğerler*, önde sternumun hemen arkasında mediyastenin önünde ve altında, diyafragmanın üzerinde, heriki akciğerin arasında *fetal kalp*, kalpten çıkan büyük damarların ve timusun bulunduğu *mediyasten* bulunmaktadır. Fetal hayatta timüs, yenidoğan ve erişkin hayata göre daha büyüktür.

Torakal organların kosta yapılarının içerisinde yer alması aslında değerlendirilirken standardizasyona olanak vermektedir. Fakat yine de toraksın en iyi değerlendirmesi klasik *dört oda aksiyal kesitinde* yapılmaktadır. Bu plan sadece intratorasik organların değil; kostalar, sternum ve cildin

ÖA/TÖF'e göre daha nadir gözlenen doğumsal hidrotoraks olgularının prenatal tanısı daha mümkündür, fakat geçici olabileceğinden gözden kolaylıkla kaçabilir. Prenatal hidrotoraksta en önemli unsur tanı koyulduğu andan itibaren fetal bulguların ciddiyetinin prognoza direkt katkısıdır. Doğumsal hidrotoraksta erken başlangıç, bilateral olma, tanı anında hidrops varlığı olumsuz prognoz lehinedir. Ancak torakoamniyotik şantın uygulanabildiği olgularda prognoz nispeten iyidir.

Mediyastenin nadir saptanan doğumsal lezyonlarının prenatal tanısı oldukça güç olup, nihai tanı çoğu olguda doğum sonrasında kesinleşmektedir. Gerek intratorasik gerekse mediyastene ait yer kaplayıcı lezyonlarda, prenatal tanı sürecinde kitlenin özelliklerini (kistik, septalı, solid, ekojenite, diffüz veya lokal), lokalizasyonunu (bronşlara yakın, arka/ön mediyasten) ve çevre organlar ile ilişkisini (mediyasten kayma, vertebra ile ilişki, servikal veya mediyasten yerleşimi) tanımlamak, var ise vasküler desteğini gösterebilmek çoğu olguda tanı da yol göstericidir.

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Sorular

1. FETO girişiminin prognozu etkileyen en önemli komplikasyonu nedir?
2. KPSM'de prognoz ve tedavi ile ilgili güncel yaklaşım nedir?
3. Pulmoner sekestrasyonların prenatal ultrasonografide tanısında önemli noktalar nelerdir?
4. ÖA/TÖF'ün prenatal tanısında indirekt/direkt ultrasonografik/MRG bulgular nelerdir?
5. Prenatal tanı almış bir larinks atrezisi olgusu için hangi genetik sendrom hakkında aileye danışmanlık verilmelidir?

Yanıtlar

1. FETO girişiminin prognozu olumsuz etkileyen en önemli komplikasyonu erken membran rüptürü ve erken doğumdur.
2. Mikrokistik tipteki konjenital pulmoner solunum yolu malformasyonunda (KPSM) prognoz daha olumsuzdur. Konjenital pulmoner solunum yolu malformasyonunda (KPSM) en olumsuz prognostik gösterge non-immun hidrops varlığıdır. Hidrops gelişen mikrokistik tipteki konjenital pulmoner solunum yolu malformasyonunda antenatal steroid kullanımı seçili olgularda yararlı olabilir. Makrokistik tipte şant uygulanabilir, mikrokistik KPSM'de radyofrekans ablasyon başarılı değildir.
3. Ekstralobar pulmoner sekestrasyonlar prenatal ultrason incelemesinde sınırları belirgin, solid, üçgen şekilli, diffüz hiperekojen kitle olarak görülürler. Ekstralobar pulmoner sekestrasyonlarda fetal dönemde hidrops gelişme şansı %5-10 civarındadır. Ekstralobar pulmoner sekestrasyonların besleyici damarının sitemik yataktan gelmesi belirleyici özelliğidir. 30. haftadan önce ciddi hidrotoraks ve hidrops gelişen pulmoner sekestrasyon vakaları fetal girişim için uygundur.
4. Fetal arka mediyastende/servikal bölgede lümen benzeri kistik dilatasyon, mide cebinin görüntülenememesi, polihidramniyos ve MRG'de hipofarinksin distandü görünüm prenatal tanıda ÖA/TÖF açısından kuşku oluşturmalıdır.
5. Fraser sendromu, kriptooftalmi, sindaktili, doğumsal respiratuar ve ürogenital sistem anomalileri ile birliktelik gösteren otozomal resesif bir hastalıktır. Laringeal/trakea atrezisi, kimi otörlere göre renal anomaliler ile beraber majör klinik kriterler arasında yer almaktadır. Bu sebeple prenatal dönemde larinks atrezisi kuşkusu olması durumunda, özellikle de böbrek anomalileri eşlik ediyorsa aile öyküsü sorgulanarak ayırıcı tanıda Fraser sendromu yer almalıdır.