BÖLÜM 18

Akciğerin Depo Hastalıkları

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GİRİŞ

Metabolik hastalıklar ve depo hastalıkları, altta yatan biyokimyasal veya metabolik işlev bozuklukları ile karakterize bir grup hastalıktan oluşur. Bu bozuklukların çoğu ya spesifik olarak ya da sistemik bir sendromun parçası olarak akciğerleri etkiler. Örneğin, amiloidoz trakeobronşiyal ağaçla sınırlı olabileceği gibi; böbrekleri, akciğerleri ve kalbi etkileyen sistemik bir hastalık şeklinde de bulgu verebilir. Bu hastalık gruplarında bazen karşılaşılan hızlı progresyon ve nonspesifik klinik semptomlar, tanısal zorluklara neden olabilir.

Görüntüleme yöntemleri, özellikle yüksek çözünürlüklü bilgisayarlı tomografi (HRCT), bu gibi zorlu klinik durumların teşhisinde oldukça faydalıdır. Bu hastalık grubunun radyolojik ve histopatolojik bulgularının korelasyonu, bu bozuklukların anlaşılmasını sağlar. Bilgisayarlı tomografi (BT), doku örneklemesi gerektiğinde girişimsel radyoloğa kılavuzluk ederek, histopatolojik tanıya da yardımcı olabilir.

Akciğerin depo hastalıkları, oldukça nadir antitelerdir. Pulmoner hastalık en sık sistemik hastalığın akciğer tutulumunun yansımasıdır. Akciğerin depo hastalıkları denilince akla pulmoner alveolar mikrolitiazis, pulmoner amiloidoz, pulmoner alveolar proteinoz, Niemann-Pick hastalığı ve Gaucher hastalığı gelmektedir.

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Asit sfingomiyelinaz eksikliği için spesifik bir tedavi şu anda mevcut değildir. Bununla birlikte, ERT tip B Niemann-Pick hastalığı olan hastalar için yararlı olabilir. Akciğer tutulumunu tedavi etmek için hematopoietik kök hücre naklinin pulmoner infiltratları başarılı bir şekilde azalttığı bildirilmiştir. Lipoid pnömoni için klasik bir tedavi olan tüm akciğer lavajının, özellikle yetişkinlerde, tip B Niemann-Pick hastalığı için nispeten etkili bir tedavi olduğu da bildirilmiştir (97). Ayrıca literatürde akciğer nakli yapılmış hastalara ait vaka sunumları da bulunmaktadır (103).

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