

22. Bölüm

KONNEKTİF DOKU HASTALIKLARI VE İNTERSİTİSYEL AKCİĞER HASTALIĞI

Sinem SAĞ¹

GİRİŞ

Kollajen doku hastalıkları, inflamatuvar disfonksiyona bağlı olarak gelişen, birçok organ sistemini tutabilen , etyolojisi net olarak bilinmeyen heterojen bir hastalık grubudur. . En sık tutulan organlardan biri akciğerler olup en ciddi tutulumlarından biri interstisyel akciğer hastalığıdır. Akciğerin tüm komponentleri etkilenebilir. Akciğer tutulumu subklinik olabileceği gibi, hayatı tehdit edici boyutta da saptanabilir. Hastalığın prognozunun tayin edilmesinden tutulumun histopatolojisi önemlidir, romatoid artrit dışındaki bağ doku hastalıklarında en sık görülen histopatolojik patern nonspesifik interstisyel pnömonidir ve prognozu diğerlerine göre daha iyidir. Tanıda özellikle solunum fonksiyon testleri ve karbonmonoksit difüzyon kapasite ile görüntüleme önemli yer tutar. Tedavide randomize kontrollü çalışma sayısı sınırlıdır. Hafif olgulara takip önerilirken , ağır hastalık aktivitesi olan veya progresyon gösteren hastalarda immünosüpresif ajanlarla tedavi önerilmektedir. Bu derlemede, yaygın olarak görülen kollajen doku hastalıklarının akciğer tutulumu gözden geçirilmiştir.

Kollojen dokusu hastalıkları (KDH) etyolojisi bilinmeyen , kronik, sistemik, inflamatuvar, heterojen bir hastalık grubudur. KDH'ları içinde romatoid artrit

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in yer aldığı en az iki sistemik otoimmün hastalığa ait bulguların birlikte görüldüğü ve anti U1-RNP antikorlarının yüksek titrede oluşu ile karakterize farklı bir sistemik otoimmün hastalıktır. En sık klinik bulguları; Raynaud fenomeni, artralji, el veya parmaklarda şişlik, özafagus motilite bozukluğudur. Prognozu pulmoner, kardiyovasküler, gastrointestinal ve santral sinir sistemi tutulumuna bağlı olarak kötü olabilir ^{21,108} Kadınlarda erkeklere oranla 10 kat kadar fazla görülmektedir. Pulmoner tutulum hastaların %20-85 kadarında görülmektedir. En sık İAH görülmekle birlikte , plevral efüzyon ve pulmoner hipertansiyonda sık görülmektedir.^{2,77}

YRBT ‘ de buzlu cam görünümü hemen hemen tüm hastalarda mevcuttur. Buzlu cam görünümü dışında nodüler ve retiküler opasitelere sıkça rastlanır. Genellikle alt zonlarda ve periferal yerleşimli olarak görülmektedir. YRBT ‘deki bu görünüm NSIP ve UIP ile uyumludur. Miks bağ doku hastalığının YRBT tutulumu genelde SSK ‘ya benzemektedir¹⁰⁹ Ancak bu hastalıkta bal peteği çok nadir görülmektedir. Mozaik atenuasyon ve küçük havayolu hastalığı genelde görülmez. Pulmoner hipertansiyona bağlı pulmoner arterlerde genişleme görülebilmektedir. MKDH’da YRBT’de en sık görülen patern NSIP’dir ^{110,111}

Hastaların %67’sinde CO diffüzyon testinde bozulma ve %50’sinde restriktif tipte solunum fonksiyon bozukluğu rapor edilmiştir¹²⁰ CO diffüzyon testi MKDH’da pulmoner fonksiyon bozukluğunu değerlendirmede en sensitif parametredir¹¹²

Tedavide yüksek doz steroidler ve siklofosfamid , mikofenolat mofetil , azatioprin gibi ajanlar kullanılmaktadır. Hayatı tehdit eden durumlarda IVIG kullanımını önerilmektedir. Bununla ilgili özellikle MKDH ile ilişkili İAH vakalarında olumlu sonuçlar bildirilmiştir ⁸⁵

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