BÖLÜM 129

PRİMER VE SEKONDER SKLEROZAN KOLANJİT

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PRİMER SKLEROZAN KOLANJİT

Giriş

Primer sklerozan kolanjit (PSK) safra ağacının kronik fibroinflamatuvar hasar ile zarar görmesi sonucu oluşan ve sıklıkla inflamatuar bağırsak hastalığı (İBH) ile ilişkili olan kolanjiyopatidir (Resim 1). PSK'lı hastaların çoğunluğunda kolanjiyografide fibrotik safra darlıkları vardır, ancak az bir kısmında ise karaciğer biyopsisinde PSK'nın histolojik özelliklerini gösteren normal kolanjiyografili küçük kanal PSK izlenir, ek olarak küçük bir yüzde de otoimmün hepatit ile örtüşen özelliklere sahiptir (PSK-OİH). PSK otoimmün bir hastalık olarak kabul edilir. PSK'ye özgü Uluslararası Hastalık Sınıflandırma (ICD)-10 tanı kodu "K83.00"tür.

Tanımlar

1960'ların ortalarına kadar yayınlanan makalelerin çoğu vaka raporlarıydı.1970'ler boyunca endoskopik retrograd kolanjiyopankreatografi (ERCP)'nin yaygın olarak uygulanması hastalığın daha fazla tanınmasına yol açtı ve 1980'de ABD, İngiltere ve Norveç'ten üç önemli makale klinik bir tanım oluşturdu. Daha sonra PSK'nın kolanjiyokarsinom (CCA), kolon neoplazisi ve küçük kanal (small-duct) PSK'nin alt fenotipleri, yüksek immünoglobulin G4 (IgG4) seviyelerine sahip PSK ve otoimmün hepatit "overlap sendromu" tanımlamaları yapıldı (1-8). PSK, intrahepatik ve/veya ekstrahepatik safra yollarındaki inflamasyon, fibrozis ve orta ve büyük safra kanallarının daralması ile karakterize, etiyolojisi bilinmeyen, kronik ilerleyici bir hastalıktır ve sıklıkla İBH ile ilişkili olan bir kolanjiopatidir. PSK'li hastaların çoğunda kolanjiyogramda fibrotik biliyer darlıklar bulunurken, az bir kısmında normal bir kolanjiyogramla karakterize edilen ancak karaciğer biyopsisinde PSK'in histolojik özellikleri bulunan küçük kanal (small-duct) PSK'i vardır. Küçük bir yüzdede otoimmün hepatit ile (PSK-OİH) örtüşen özellikler içerir. PSK hem erkek hem de kadın bireyleri etkiler ve her yaşta ortaya çıkabilir. PSK bir otoimmün hastalık olarak kabul edilir; ancak patofizyolojisi tam olarak anlaşılamamıştır. PSK sıklıkla kolestatik karaciğer hasarı, siroz ve karaciğer yetmezliği ile sonuçlanır ve transplantasyondan sonra hastaların %20-30'unda tekrarlayabilir. PSK ayrıca CCA ve kolorektal kanser (KRK) riskini de önemli ölçüde artırır (8). Şu anda PSK için etkili bir tıbbi tedavi yoktur. Biliyer darlıkları tanımla-

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