BÖLÜM 17

KARACİĞER VASKÜLER HASTALIKLARI

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GİRİŞ

Sirozlu hastalarda görülen portal ven trombozu (PVT) hariç karaciğerin vasküler bozukluklarının çoğu nadirdir. Bu hastalıklar nadir görülmelerine karşın önemli morbidite ve mortaliteye sahiptir. Karaciğer vasküler bozukluklarının nadirliği göz önüne alındığında sadece portal ven trombozu, Budd-Chiari sendromu, hepatik sinüzoidal obstrüksiyon sendromu, konjenital vasküler malformasyonlar ve idiopatik nonsirotik portal hipertansiyon gözden geçirilecektir. Tablo-1 de karaciğer vasküler hastalıkları özetlenmiştir.

Tablo-1:Karaciğer Vasküler Hastalıkları

Portal ven trombozu
Hepatik sinüzoidal obstrüksiyon sendromu
Budd-Chiari sendromu
Konjenital vasküler malfarmasyonlar
İdiopatik nonsirotik portal hipertansiyon
Peliosis hepatitis ve sinüzoidal dilatasyon
Hepatik arter tromboz ve anevrizması
Radyasyona sekonder karaciğer hastalıkları

PORTAL VEN TROMBOZU

Tanım

Portal ven, splenik ven ve süperior mezenterik venin pankreas boynu arkasında birleşmesiyle oluşan, kapakçıkları olmayan yaklaşık 8 cm uzunluğunda venöz bir damardır. Tüm sindirim sisteminin kanını karaciğere taşır ve karaciğere gelen kanın %75 ini sağlar. Portal ven porta hepatiste sağ ve sol dallarına ayrıldıktan sonra hepatik loblar içerisinde dalcıklar şeklinde ilerler ve hepatik sinüzoidlerle son bulur (1).

Portal ven trombozu(PVT) portal ven ve / veya dallarının ekstrahepatik kısmının trombozu şeklinde tanımlanır. Portal ven trombozu nadir bir hastalık olup, genel popülasyonda prevalansı 0,7 ila 1/100.000 arasında değişmekle birlikte bazı durumlarda prevalansı daha fazladır. Örneğin karaciğer sirozu hastalarında %2,1-%16,2, karaciğer nakli adaylarında ise %5,5-%26 arasında bildirilmiştir (2,3).

Glikojenin yıkımındaki bozukluk sonucunda organlarda metabolitlerin birikmesi ile karakterize etyolojisine göre sirotik portal ven trombozu ve nonsirotik portal ven trombozu şeklinde iki gruba ayrılabilir. Ayrıca PVT klinik açıdan, aynı hastalığın ardışık aşamalarını temsil eden ve benzer etyolojileri paylaşan, ancak tedavileri farklılık

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daki hastaların belirlenmesi, koruyucu önlemlerin alınması ve destek tedavisi şeklindedir.

Herediter hemorajik telenjiektazi otozomal dominant kalıtımla geçen genetik bir hastalıktır. Klinik prezantasyon hastadan hastaya büyük ölçüde değişir. Tanı çoğu zaman doppler ultrasonografi ile konur. Tedavi semptomatik olup, büyük venöz malfarmasyonların tedavisi için embolizasyon ya da karaciğer nakli düşünülebilir

İdiopatik nonsirotik portal hipertansiyon(İNSPH), portal hipertansiyon yapan diğer tüm nedenler ekarte edildikten sonra tanı konulur. Etyolojisi tam olarak bilinmemektedir. Tanı için karaciğer biyopsisi şarttır. Tedavi portal hipertansiyon komplikasyonlarına yöneliktir.

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