CHAPTER 23

KAWASAKI DISEASE (M30.3)

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REMEMBER

- Kawasaki disease (KD) is defined as mucocutaneous lymph node syndrome.
- The incidence of the disease differs according to geographical regions and is most frequently observed in Japan.
- Some HLA loci such as HLA B5, and HLA-DRB1 were associated with KD. It has also been implicated in some genetic pathways such as CASP3 (Caspase3).
- It has been suggested that the disease may be triggered by some infectious agents (e.g., Staphylococcal toxin, Ebstein-Barr virus).
- KD is a multisystemic medium-sized vessel vasculitis that frequently occurs in childhood.
- KD is most commonly observed in children younger than 6 years old, and more prevalent in males (about 1.5 times).
- The disease may be observed very rarely in adulthood.
- KD is one of the leading causes of acquired heart disease in the pediatric population.
- The disease is usually self-limited, but coronary artery aneurysms may occur in 15-25% of patients.
- Diagnostic criteria for KD have been established. Includes high fever (>38.5) unresponsive to antipyretics lasting at least five days and the presence of **four of the five criteria** listed below:

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- Close follow-up is required because Kawasaki disease may rarely cause hemolytic uremic syndrome. Suspicious cases should be consulted with a pediatric nephrologist.

Pediatric Neurology

- Central nervous system involvement is very rare. Cerebral vasculitis can be observed. Peripheral nerve involvement may occur.
- Cases of ataxia, extreme irritability, cranial nerve palsies, aseptic meningitis, and sensorineural hearing loss have been reported in literature. Patients should be followed in these aspects and the specialist should be consulted if necessary.

Gastroenterology

- Appendicitis, paralytic ileus, pancreatitis, cholestasis, acute gallbladder hydrops, hemorrhagic duodenitis may occur in KD patients. In case of doubt in clinical follow-up, the gastroenterologist should be consulted.

Pediatric Surgery

- Patients with gastrointestinal obstruction, pseudoobstruction, and intussusception have been reported in some children diagnosed with Kawasaki disease. In these cases, acute surgical evaluation is required.

Hematology

- Macrophage activation syndrome (MAS) is a rare complication of KD and is characterized by cytopenia, hepatosplenomegaly, hypofibrinogenemia, and markedly elevated ferritin. Patients in doubt should be consulted with a pediatric hematologist.

Cardiac Surgery

- KD may also involve peripheral arteries. Ischemia, and gangrene may develop in these patients, and should be evaluated together with a cardiovascular surgeon when necessary.
- In cases where coronary artery disease is developed, an indication for catheterization or cardiac bypass may arise.

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BÖLÜM 23

KAWASAKİ HASTALIĞI (M30.3)

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HATIRLA

- Kawasaki hastalığı (KH) mukokutanöz lenf nodu sendromudur.
- Hastalık insidansı coğrafi bölgelere göre farklılık gösterir ve en sık Japonya'da izlenir.
- HLA B5 ve HLA-DRB1 gibi bazı HLA lokusları ile hastalık arasında ilişki saptanmıştır. CASP3 (Caspase3) gibi bazı genetik yolaklarla da KH ilişkilendirilmiştir.
- Hastalığın bazı enfeksiyöz ajanlar tarafından tetiklenebildiği öne sürülmüştür (ör. Stafilokoksik toksin, Ebstein-Barr virüs).
- Sıklıkla çocukluk çağında görülen hastalıkta ana patoloji multisistemik orta damar vaskülitidir.
- En sık 6 yaşından küçük çocuklarda izlenmektedir. Erkek çocuklarda kızlara göre daha sıktır (yaklaşık 1.5 kat).
- KH, çok nadir olarak erişkin çağda da izlenebilir.
- Pediatrik popülasyonda edinilmiş kalp hastalığının önde gelen nedenlerinden birisidir.
- Hastalık genelde kendini sınırlar ancak hastalarının %15-25'inde koroner arter anevrizmaları gelebilir.
- KH için tanı kriterleri oluşturulmuştur. En az beş gün süren antipiretiklere yanıtsız yüksek ateş (>38.5) ve aşağıdaki **beş kriterden dördünün varlığını** içerir:

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yakın takip gerekir. Şüpheli vakalar pediatrik nefroloji uzmanına danışılmalıdır.

Pediatrik Nöroloji

- Santral sinir sistemi tutulumu oldukça nadirdir. Serebral vaskülit izlenebilir. Periferik sinir tutulumu görülebilir.
- Ataksi, aşırı sinirlilik, kraniyal sinir felçleri, aseptik menenjit ve sensörinöral işitme kaybı gelişen vakalar bildirilmiştir. Hastalar bu açılardan takip edilmeli ve uzmana gerekirse danışılmalıdır.

Gastroenteroloji

- Hastalık seyri sırasında apandisit, paralitik ileus, pankreatit, kolestaz, akut safra kesesi hidropsu, hemorajik duodenit gelişen hastalar bildirilmiştir. Klinik takipte şüphe duyulduğunda gastroenteroloji uzmanına danışılmalıdır.

Çocuk Cerrahisi

- Kawasaki hastalığı tanısı alan bazı çocuklarda gastrointestinal obstrüksiyon, psödoobstrüksiyon ve invajinasyon gelişen hastalar bildirilmiştir. Bu durumlarda akut cerrahi görüş alınmalıdır.

Hematoloji

- Makrofaj aktivasyon sendromu (MAS) KH'nin nadir bir komplikasyonudur ve sitopeni, hepatosplenomegali, hipofibrinojenemi ile belirgin ferritin yüksekliği ile karakterizedir. Şüphe olan hastalar pediatrik hematoloji uzmanına konsulte edilmelidir.

Kalp ve Damar Cerrahisi

- KH periferik arterleri de tutabilmektedir. Bu hastalarda iskemi ve gangren gelişebilmektedir ve gerekli durumlarda kalp damar cerrahisi uzmanı ile birlikte değerlendirilmelidir.
- Koroner arter hastalığı gelişen vakalarda kateterizasyon veya kardiyak by-pass endikasyonu doğabilir.

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