CHAPTER 38

POLYARTERITIS NODOSA (M30.0)

Elif ÇETİNKAYA[1]
Ayşe Nilhan ATSÜ[2]

REMEMBER

- Polyarteritis nodosa (PAN) is a segmental necrotizing vasculitis involving medium-sized muscular arteries.
- PAN may present as a skin-limited disease or systemic involvement may occur.
- Disease involvement is variable. The disease spectrum ranges from single organ involvement to multiple organ failure. Any organ can be affected, but PAN usually does not involve the lungs.
- The disease can be classified under three different types: Systemic idiopathic form (idiopathic generalized PAN), cutaneous PAN, and hepatitis B virus (HBV) associated PAN.
- In European countries, the incidence of PAN ranges from 0 to 1.6 cases per million, and its prevalence is approximately 31 cases per million.
- PAN commonly occurs in middle-aged adults, but it can also be observed in the pediatric age group, but it is rare. The disease is more frequent in compared to women.
- Adenosine deaminase 2 (ADA2) mutation has been detected in some cases.
- Occlusion or rupture of inflamed arteries can cause tissue ischemia or bleeding in various organs and systems. In conclusion, patients with PAN have a wide spectrum of clinical manifestations, including nonspecific manifestations such as fever, weight loss, malaise, myalgia, and/or arthralgia, as well as symptoms resulting from dysfunction or damage of target organs.

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BÖLÜM 38

POLİARTERİTİS NODOSA (M30.0)

Elif ÇETİNKAYA[1]
Ayşe Nilhan ATSÜ[2]

HATIRLA

- Poliarteritis nodosa (PAN) orta boy musküler arterleri tutan, segmental nekrotizan vaskülittir.
- PAN deriye sınırlı bir hastalık şeklinde prezente olabilir veya sistemik tutulum olabilir.
- Hastalık tutulumu değişkendir. Hastalık spektrumu, tek bir organ tutulumundan çoklu organ yetmezliğe kadar uzanır. Herhangi bir organ etkilenebilir, ancak PAN genellikle akciğerleri tutmaz.
- Hastalık üç farklı tip altında incelenebilir: Sistemik idiyopatik form (idiyopatik jeneralize PAN), kutanöz PAN, hepatit B virüsü (HBV) ile ilişkili PAN.
- Avrupa ülkelerinde, PAN insidansı milyonda 0 ile 1,6 vaka arasında değişmektedir ve prevalansı yaklaşık olarak milyonda 31 vakadır.
- PAN sıklıkla orta-yaşlı erişkinlerde görülür, pediyatrik yaş grubunda da görülebilir ancak nadirdir. Erkeklerde kadınlara göre daha sık izlenir.
- Bazı vakalarda adenozin deaminaz 2 (ADA2) mutasyonu saptanmıştır.
- İnflamasyonlu arterlerin tıkanması veya yırtılması, çeşitli organ ve sistemlerde doku iskemisi veya kanamaya neden olabilir. Sonuç olarak, PAN hastalarında ateş, kilo kaybı, kırgınlık, miyalji ve/veya artralji gibi spesifik olmayan belirtiler ile hedef organların işlev bozukluğu veya hasarından kaynaklanan semptomlar dahil olmak üzere geniş bir klinik belirti spektrumu görülür.

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