

CHAPTER 43

SARCOIDOSIS (D86.9)

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REMEMBER

- Sarcoidosis is a systemic granulomatous disease of unknown cause, most commonly involving the lungs.
- Sarcoidosis is one of the diseases known as “great imitators” in dermatology.
- The etiopathogenesis of sarcoidosis, and the triggering factors (some infectious agents, and professions have been accused) have not been fully elucidated. It is known that the risk of disease increases with a family history (e.g., HLA-DRB1*04:01 gene).
- Sarcoidosis is slightly more likely to occur in women compared to men.
- Skin manifestations of sarcoidosis may be the first clinical manifestation of the disease.
- There are disease-specific and non-specific cutaneous lesions in sarcoidosis.
- Red-purple papules, and plaques are most commonly located on the face (especially on the nose) neck, upper back, and extremities. These are non-specific skin lesions, and heal without scarring.
- It is known that the most common non-specific skin lesion is erythema nodosum in sarcoidosis.
- Lupus pernio is a specific lesion of sarcoidosis, and occurs in areas exposed to cold (mostly cheeks, nose, ears). It is characterized by violaceous papules, and plaques, and may heal with scar tissue.
- Cutaneous lesions of sarcoidosis may place on tattoos, and scarred areas.

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Hematology

- ▶ Lymphopenia, leukopenia, eosinophilia, hypergammaglobulinemia, and anemia may occur in bone marrow involvement.
- ▶ In the presence of abnormalities in complete blood count, and serum protein electrophoresis, hematology specialist should be consulted in terms of bone marrow involvement.

Endocrine and Metabolism Disorders

- ▶ Patients may experience hypercalcemia, pituitary and thyroid dysfunction secondary to calcitriol released from sarcoidal histiocytes. Extended hormone testing and endocrinology consultation is required when clinically indicated.

Nephrology

- ▶ The amount of calcium excreted in the urine increases in patients due to hypercalcemia secondary to calcitriol released from sarcoidal histiocytes.
- ▶ As a result, nephrolithiasis, and deterioration in kidney function tests may occur. In the presence of increased creatinine, BUN, serum, and spot urinary calcium, patients should be referred to nephrology specialist.

Obstetrics & Gynecology and Urology

- ▶ In patients with sarcoidosis, granulomatous masses that are usually asymptomatic may occur in the ovaries and testicles. Patients should be consulted with related branches in terms of differential diagnosis that may cause ovarian and testicular masses.

Clinical Pharmacology

- ▶ The possibility of drug-induced cutaneous sarcoidosis should be excluded in patients. For example, IFN- α which is for hepatitis C viral infection, and TNF- α inhibitors

may lead to cutaneous sarcoidosis. If the patient's existing lesions are associated with the triggering drug, a clinical pharmacologist's evaluation is required.

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SARKOİDOZ (D 86.9)

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HATIRLA

- Sarkoidoz, en sık akciğerleri tutan ve nedeni bilinmeyen sistemik granülatöz bir hastalıktır.
- Dermatolojide “büyük taklitçi” olarak anılan hastalıklardandır.
- Hastalığın etyopatogenezi ve tetikleyici faktörler (bazı enfeksiyöz ajanlar ve meslekler suçlanmış- tır) tam olarak aydınlatılamamıştır. Aile hikayesi varsa hastalık riskinin arttığı bilinmektedir (ör: *HLA-DRB1*04:01 geni*).
- Sarkoidozun kadınlarda görülme olasılığı erkeklere göre hafif olarak daha yüksektir.
- Sarkoidozun deri bulguları hastalığın ilk klinik belirtisi olabilir.
- Sarkoidozda hastalığa spesifik ve spesifik olmayan deri lezyonları vardır.
- Kırmızı-morumsu papüller ve/veya plaklar özellikle burun olmak üzere en sık yüzde, boyun, üst sırt ve ekstremitelerde yerleşir. Bunlar spesifik olmayan deri lezyonlarıdır ve skarsız iyileşirler.
- En sık spesifik olmayan deri lezyonunun ise eritema nodozum olduğu bilinmektedir.
- Lupus pernio sarkoidozun spesifik bir deri lezyonudur ve soğuğa maruz kalan bölgelerde (daha çok yanaklar, burun, kulaklar gibi) görülür. Viyolase papül ve plaklar ile karakterize olup skar dokusu ile iyileşebilir.
- Sarkoidozun deri lezyonları deride dövme ve skar alanları gibi yerleri seçebilir.

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Endokrin ve Metabolizma Hastalıkları

- ▶ Hastalarda sarkoidal histiyositlerden salınan kalsitriole sekonder hiperkalsemi, hipofiz ve tiroid fonksiyon bozukluğu meydana gelebilir. Klinik endike olduğunda genişletilmiş hormon testi ve endokrinoloji konsültasyonu istenebilir.

Nefroloji

- ▶ Hastalarda sarkoidal histiyositlerden salınan kalsitriole sekonder gelişen hiperkalsemiye bağlı idrarla atılan kalsiyum miktarı artar.
- ▶ Buna bağlı olarak nefrolitiazis ve böbrek fonksiyon testlerinde bozulma meydana gelebilir. Kreatinin, BUN, serum ve spot idrar kalsiyum miktarında artış varlığında hastalar nefroloji uzmanı tarafından değerlendirilmelidir.

Kadın Hastalıkları ve Doğum & Üroloji

- ▶ Sarkoidozlu hastalarda over ve testislerde genellikle asemptomatik seyreden granüloamatöz kitleler oluşabilir. Overler ve testislerde kitleye neden olabilecek başka hastalıkların dışlanması ve gerekli görülürse biyopsi açısından hastalar ilgili branşlara danışılmalıdır.

Klinik Farmakoloji

- ▶ Hastalarda ilaca bağlı kutanöz sarkoidoz olasılığını dışlanmalıdır. Örneğin hepatit C viral enfeksiyonu için IFN- α kullanımı veya TNF- α inhibitörleri kutanöz sarkoidoza neden olabilir. Hastanın mevcut lezyonları tetikleyici ilaçla ilişkilendiriliyorsa klinik farmakoloji uzmanından görüş alınmalıdır.

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