CHAPTER 8

DERMATOMYOSITIS (M33)

Defne ÖZKOCA[1]

REMEMBER

- Dermatomyositis (DM) is a multisystem, autoimmune connective tissue disease.
- While the possibility of underlying malignancy is high in adults (approximately 40%), this situation is not expected in the pediatric age group.
- The female/male ratio is 2/1 in DM.
- Typical violaceous skin rash, heliotrope rash, Gottron papules (atrophic dermal papules of dermatomyositis), Gottron sign, Keining sign, photosensitivity, shawl sign, calcinosis cutis, poikiloderma, erythema, nail fold changes, alopecia, Raynaud's phenomenon can be observed during disease course.
- Erythematous scaly lesions resembling psoriasis or seborrheic dermatitis may be detected on the scalp.
- Painful fissures may develop in the fingers and impede movement: The mechanic's hand.
- Nail fold changes such as capillary enlargement and increased folding can be detected, thus capillaroscopy should be performed in every patient. In addition, periungual changes can give an idea about disease activity, treatment response, and/or internal organ involvement.
- Patients with DM may experience itching.
- Muscular involvement is usually occurs as symmetric extensor proximal myopathy, but it is not essential for diagnosis. The amyopathic form of the disease should not be forgotten.

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Gastroenterology

- The musculature of the gastrointestinal tract may be also involved in dermatomyositis. Weakness of the striated muscles esophagus is probable and this may cause difficulty in swallowing. In the presence of any symptoms, patients should be consulted with a gastroenterologist.

Neurology

- Evaluation of muscle weakness due to muscle involvement is important. Proximal, and symmetrical involvement is expected in DM. Electromyography (EMG) can be performed to aid the diagnosis.
- The only definitive way to demonstrate inflammation within the muscle is a muscle biopsy.
- Dermatomyositis can also lead to peripheral neuropathy. In case of doubt, the patient should be evaluated by a neurologist.

Physical Medicine and Rehabilitation (PM&R)

- Both the disease itself and systemic corticosteroids, which are frequently used in treatment, can cause proximal muscle weakness. Therefore, cooperation with PM&R specialists is essential for strengthening the proximal muscles.

Pulmonary Diseases

- If the pharynx muscles are involved, there may be voice disorders.
- In some cases, there may be respiratory tract disease with very serious involvement such as acute respiratory distress (ARDS), and diffuse interstitial fibrosis.
- Very rarely, pulmonary hypertension may develop. Patients should be followed for these aspects, and a pulmonologist should be consulted when necessary.

Cardiology

- There may also be involvement of the heart muscle (myocarditis). Rhythm problems such as bradycardia and tachycardia may occur.
- Serious involvements such as cardiomegaly, congestive heart failure, and bundle branch blocks may be observed and require evaluation by a cardiologist.

Clinical Pharmacology

- DM can sometimes be triggered by some drugs such as hydroxyurea, statin group lipid lowering agents, penicillamine, and cyclophosphamide. Patients should be questioned the culprit drugs.

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BÖLÜM 8

DERMATOMİYOZİT (M33)

Defne ÖZKOCA[1]

HATIRLA

- Dermatomiyozit (DM), multisistemik ve otoimmün bir bağ doku hastalığıdır.
- Erişkinlerde altta yatan malignite olasılığı yüksek iken (yaklaşık %40) pediatrik yaş grubunda bu durum beklenmez.
- Hastalık görülme sıklığında kadın/erkek oranı 2/1'dir.
- Hastalarda tipik viyolase deri döküntüsü, heliotrop raş, Gottron papülleri (dermatomiyozitin atrofik dermal papülleri), Gottron belirtisi, Keining işareti, fotosensitivite, şal belirtisi, kalsinosiz kutis, poikiloderma, eritem, tırnak yatağı değişiklikleri, alopesi, Raynaud fenomeni izlenebilir.
- DM'li bireylerin saçlı derisinde psöriazis veya seboreik dermatite benzeyen eritemli-skuamlı plaklar saptanabilir.
- El parmaklarında ağrılı fissürler gelişebilir ve hareketi engelleyebilir: makinist eli.
- Kapiller genişleme ve kıvrımlanma artışı gibi periungual değişiklikler görülebilir ve kapilleroskopik muayene mutlaka yapılmalıdır. Ayrıca periungual değişiklikler hastalık aktivitesi, tedavi cevabı ve/veya iç organ tutulumu gibi durumlar hakkında fikir verebilir.
- Dermatomiyozitli hastalarda kaşıntı görülebilir.
- Kas tutulumu simetrik ekstansör proksimal myopati şeklindedir ancak tanı için şart değildir. Hastalığın amyopatik formu olduğu da unutulmamalıdır.

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gili kaslarında da zayıflık görülebilir ve bu durum yutma güçlüğüne sebep olabilir. Semptomatik hastalar bu açıdan değerlendirilmelidir.

Nöroloji

- Kas tutulumuna bağlı kas güçsüzlüğünün değerlendirilmesi önemlidir. Dermatomiyozitte proksimal ve simetrik tutulum beklenir. Tanıya yardımcı olarak elektromiyografi ((EMG) yapılabilir.
- Kasta inflamasyonu göstermenin tek kesin yolu kas biyopsisidir.
- DM periferik nöropatiye de yol açabilir. Şüphe durumunda hastalar nöroloji uzmanı tarafından değerlendirilmelidir.

Fiziksel Tıp ve Rehabilitasyon (FTR)

- Hem hastalığın kendisi hem de tedavide sıklıkla kullanılan sistemik kortikosteroidler proksimal kas güçsüzlüğü ile sonuçlanabilir. Bu nedenle proksimal kasların güçlendirilmesi için FTR uzmanları ile işbirliği şarttır.

Göğüs Hastalıkları

- Farinks kaslarında tutulum olursa ses bozuklukları meydana gelebilir.
- Bazı vakalarda akut respiratuvar distress (ARDS) ve diffüz interstisyel fibrosis gibi çok ciddi tutulumların olabildiği solunum yolu hastalığı olabilir.
- Çok nadir olarak pulmoner hipertansiyon gelişebilir. Hastalar bu yönlerden takip edilmeli ve gerekli uzmanına danışılmalıdır.

Kardiyoloji

- Dermatomiyozitin seyrinde kalp kasında da tutulum olabilir (myokardit). Bradikardi ve taşikardi gibi ritm problemleri izlenebilir.
- Kardiyomegali, konjestif kalp yetmezliği ve dal blokları gibi ciddi tutulumlar görülebilir ve kardiyoloji uzmanı tarafından değerlendirmeyi gerektirir.

Klinik Farmakoloji

- Dermatomiyozit bazen hidroksiüre, statin grubu lipit düşürücüler, penisilamin ve siklofosfamid gibi bazı ilaçlar tarafından da tetiklenebilmektedir. Hastalar bu ilaçların kullanımı açısından sorgulanmalıdır.

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