

# CHAPTER 32

## MYCOSIS FUNGOIDES (C84.0)

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### REMEMBER

- ▶ Cutaneous T-cell lymphomas (CTCLs) are a group of diseases that originate from skin-tropic CD41memory T lymphocytes (CD8-, CD4-, and CD81 subtypes can also be seen). CTCLs are a group of diseases belong to non-Hodgkin lymphomas.
- ▶ More than half of these CTCL cases are mycosis fungoides (MF), and the incidence of the disease has been reported as 5.6 per million.
- ▶ Mycosis fungoides and its variants (granulomatous slack skin, folliculotropic mycosis fungoides, and pagetoid reticulosis), Sézary syndrome, lymphomatoid papulosis, and cutaneous anaplastic large cell lymphoma account for 90% of all CTCL cases.
- ▶ Both MF and SS occur more frequently in males. The mean age of onset of MF and SS is the sixth decade. Although the disease is usually seen in adults, it may also occur in the pediatric population.
- ▶ The exact etiology of MF is unclear. Occupational exposure, environmental pollutants (e.g. 2,3,7,8-tetrachlorodibenzo-p-dioxin) and ultraviolet (UV) radiation are discussed as possible triggers. Chronic antigen or superantigen stimulation is thought to result in T-cell clonal proliferation and malignant transformation, however, data on this topic are limited. Although it is discussed that some infectious agents such as human t-cell lymphotropic virus (HTLV-1) may trigger MF, clear results have not been reported.

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- ▶ T-cell receptor (TCR) polymerase chain reaction (PCR) (used to identify a clonal T cell population studies from peripheral blood/skin/lymph node can be performed and/or flow cytometry may be utilized.

### General Surgery

- ▶ The planning of excisional biopsy of the lymph node should be performed by a general surgeon.

### Pathology

- ▶ Immunostaining of the pathology is very important in the diagnosis of mycosis fungoides. It is important to always work with a skin pathologist.

### Infectious Diseases

- ▶ Patients with MF and SS are at increased risk of bacterial infection, particularly in the advanced stages, due to the disruption of the skin barrier by ulcerated tumors as well as suppressed systemic, and local immune response to pathogens. Therefore, it is appropriate for patients to be evaluated periodically by a specialist.

### Psychiatry

- ▶ The prevalence of anxiety, and depression are increased in patients with MF compared to the normal population and, are associated with disease severity. It may be appropriate for patients to be followed up by a psychiatrist.

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## MİKOZİS FUNGOİDES (C84.0)

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### HATIRLA

- Kutanöz T hücreli lenfomalar (KTHL'ler), deri-tropik CD41hafıza T lenfositlerinden kaynaklanan (CD8<sup>+</sup>, CD4<sup>-</sup> ve CD81 alt tipleri de görülebilir) ve Hodgkin olmayan lenfomalar içinde incelenen bir grup hastalıktır.
- KTHL'lerin genel insidansı milyonda 10.2'dir. Bu vakaların yarısından fazlası mikozis fungoides (MF) ve hastalık insidansı milyonda 5.6 olarak bildirilmiştir.
- Mikozis fungoides ve varyantları (granümatöz gevşek deri, folikülotropik mikozis fungoides, pagetoid retiküloz), Sézary sendromu (SS), lenfomatoid papüloz ve kutanöz anaplastik büyük hücreli lenfoma, tüm KTHL vakalarının %90'ını oluşturur.
- Hem MF hem de SS erkeklerde daha sık ortaya çıkar. MF ve SS'in ortalama ortaya çıkış yaşları altıncı dekattır. Hastalık genellikle erişkinlerde görülmesine rağmen pediatrik popülasyonda da ortaya çıkabilir.
- MF'in kesin etiyoloji belirsizdir. Mesleki maruziyet, çevre kirlenmeleri (ör: 2,3,7,8- tetraklorodibenzo-p-dioksin) ve ultraviyole (UV) radyasyonu olası tetikleyiciler olarak tartışılmaktadır. Kronik antijen veya süperantijen stimülasyonunun, T-hücresinin klonal proliferasyonuna ve malign transformasyona yol açtığı düşünülmektedir ancak bu konu hakkındaki veriler sınırlıdır. İnsan T-lenfotropik virüsü (HTLV-1) gibi bazı enfeksiyöz ajanların MF'i tetikleyebileceği tartışılrsa da net sonuçlar elde edilememiştir.

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## Genel Cerrahi

- › Lenf nodlarına yönelik eksizyonel biyopsinin planlaması genel cerrahi uzmanı tarafından yapılmalıdır.

## Patoloji

- › Mikozis fungoides tanısında patolojinin immün boyamaları oldukça önemlidir. Her zaman bir deri patoloğu ile birlikte çalışılmalıdır.

## Enfeksiyon Hastalıkları

- › MF/SS'li hastalar, özellikle ileri evrelerde, ülsera tümörler tarafından deri bariyerinin bozulmasının yanı sıra patojenlere karşı bastırılmış sistemik ve lokal bağışıklık tepkisi nedeniyle artmış bakteriyel enfeksiyon riski altındadır. Hastaların bu nedenle bir enfeksiyon uzmanı tarafından belirli periyotlarla değerlendirilmesi uygundur.

## Psikiyatri

- › MF'li hastalarda normal popülasyonda göre anksiyete ve depresyon artmıştır ve hastalık şiddeti ile ilişkilidir. Hastaların psikiyatri uzmanınca takip altına alınması uygun olabilir.

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