

CHAPTER 36

PEMPHIGUS VULGARIS (L10.0)

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REMEMBER

- ▶ Pemphigus vulgaris (PV) is an acquired autoimmune bullous disease. Skin involvement may or may not accompany involvement of mucous membranes.
- ▶ Epidemiological data of PV show variability between countries, the incidence is highest in Ashkenazi Jews. It is more common in women compared to men. PV usually occurs in individuals aged 40-60 years, and the disease can also be observed in the pediatric population.
- ▶ Key target antigens in PV are desmoglein (Dsg) 1 and 3. Desmogleins are important components of desmosomes that hold keratinocytes together in the epithelium, and in the defect of these proteins, keratinocytes separate from each other causing the formation of bullae.
- ▶ It is known that various genetic factors play a role in the pathogenesis of PV. It has been reported that HLA-DRB1 polymorphisms constitute an important risk factor for the development of PV.
- ▶ PV most commonly involves the oral mucosa. Less commonly, it may also affect the anogenital mucosa, and rarely the nasal, ocular, and/or esophageal mucosa. Ocular involvement may present as hyperemia, conjunctivitis, hyperemia, and/or erosion.
- ▶ Oral erosions in the areas of high friction risk, such as the retromolar trigone, may heal slowly due to ongoing trauma despite disease control.

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Dentistry

- ▶ The patients may be consulted in terms of involvement (stomatitis, gingivitis). It is also important to evaluate in terms of prostheses that are not compatible with the mouth.

Otorhinolaryngology

- ▶ Consultation should be requested in terms of nasopharynx and/or larynx involvement. However, it should not be forgotten that erosions may intensify and/or new lesions may occur during endoscopic examination, especially in the acute period.

Gynecology and Obstetrics

- ▶ Theoretically, it is known that the vagina, vulva and even cervix can be involved in women with PV although it is rare. It may result in synechia. In case of doubt, evaluation by a gynecologist is required in terms of involvement.

Gastroenterology

- ▶ Involvement may occur in all parts of the gastrointestinal tract, most commonly the esophagus. Therefore, symptoms such as dysphagia, reflux-like symptoms and/or rectal bleeding should be assessed by a gastroenterologist.

Nutrition and dietetics

- ▶ Some nutrients are known to trigger pemphigus (e.g., leek, cabbage, onion, cherry, peach, vanilla, cocoa and its products, some nuts such as walnuts, ginger, raspberry).
- ▶ For this reason, patients with pemphigus vulgaris should be given an appropriate diet program, and patient education should be provided.

Clinical pharmacology

- ▶ It is known that pemphigus can be triggered by various drugs. These include rifampin, nifedipine, some antiepileptics, aspirin, captopril, vancomycin, hydroxychloroquine, and penicillamine. In this respect, patients can be evaluated by a clinical pharmacology specialist.

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PEMFİGUS VULGARİS (L10.0)

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HATIRLA

- ▶ Pemfigus vulgaris (PV), edinsel bir otoimmün büllöz hastalıktır. Deri tutulumu müköz membranların tutulumuna eşlik edebilir veya etmeyebilir.
- ▶ PV epidemiyolojik verileri ülkeler arası değişkenlik gösterir, en sık insidans Aşkenazi yahudilerinde izlenir. Kadınlarda erkeklere göre daha sık rastlanır. Genellikle 40-60 yaş grubu bireylerde ortaya çıkar, pediyatrik popülasyonda da hastalık görülebilir.
- ▶ PV’de anahtar hedef antijenler, desmoglein (Dsg)1 ve Dsg3’tür. Desmogleinler, epitelde keratinositleri bir arada tutan dezmozomların önemli bileşenleridir ve bu proteinlerin bozukluklarında keratinositler birbirinden ayrılır ve büller ortaya çıkar.
- ▶ PV’de bazı genetik faktörlerin etkili olduğu bilinmektedir. HLA-DRB1 polimorfizmlerinin, PV gelişimi için önemli bir risk faktörü oluşturduğu bildirilmiştir.
- ▶ PV en sık oral mukozayı tutar. Daha az sıklıkla anogenital mukozayı ve nadiren nazal, oküler veya özofagus mukozasını da etkileyebilir. Oküler tutulum hiperemi, konjonktivit, hiperemi ve/veya erozyon şeklinde ortaya çıkabilir.
- ▶ Retromolar üçgen gibi sürtünme riski yüksek alanlarındaki oral erozyonlar, hastalık kontrolüne rağmen devam eden travma nedeniyle yavaş iyileşebilir.

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Kadın Hastalıkları ve Doğum

- ▶ Kuramsal olarak kadınlarda vajina, vulva ve hatta serviks tutulumu olabileceği bilinmektedir ancak nadirdir. Tutulum sineşi ile sonuçlanabilir. Şüphe durumunda tutulum yönünden konsültasyon istenir.

Gastroenteroloji

- ▶ En sık özofagus olmak üzere gastrointestinal sistemin tüm kısımlarında tutulum olabilir. Bu nedenle yutma güçlüğü, reflü benzeri semptomlar ve/veya rektal kanama gibi semptomlarda mutlaka değerlendirme yapılmalıdır.

Beslenme ve Diyetetik

- ▶ Bazı besinlerin pemfigusu tetiklediği bilinmektedir (ör: pırasa, lahanası, soğan, kiraz, şeftali, vanilya, kakao ve ürünleri, ceviz gibi bazı kuruyemişler, zencefil, ahududu).
- ▶ Bu nedenle hasta eğitimi ve uygun diyet program için diyetisyen kontrolünde olması önem arz etmektedir.

Klinik Farmakoloji

- ▶ Bazı ilaçlar tarafından da pemfigusun tetiklenebileceği bilinmektedir. Bu ilaçlar arasında rifampin, nifedipin, bazı antiepileptikler, aspirin, kaptopril, vankomisin, hidroklorokin ve penisilamin yer alır. Hastalar bu yönden klinik farmakoloji uzmanı ile birlikte değerlendirilebilir.

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