

SWEET'S SYNDROME (ACUTE FEBRILE NEUTROPHILIC DERMATOSIS) (L98.2)

Nilgün SOLAK¹

REMEMBER

- ▶ Classic Sweet syndrome (SS) is a neutrophilic dermatosis of acute onset with constitutional symptoms such as fever, malaise, joint±muscle pain and erythematous papules and/or plaques.
- ▶ Sweet syndrome may be concomitant with other neutrophilic dermatoses (such as neutrophilic eccrine hidradenitis, erythema elevatum diutinum, subcorneal pustular dermatosis and pyoderma gangrenosum).
- ▶ Head-neck region, and upper extremities are frequently involved in SS.
- ▶ SS is more common in women compared to men (female/man: 4/1) and the disease usually occurs between the ages of 20-60.
- ▶ Pseudovesicular or pseudopustular appearance may be observed due to severe edema in papules, and plaques. True vesiculation, bullae and pustules may occur in some patients.
- ▶ Etiology of SS may include infections, pregnancy, drugs, malignancies (often hematological malignancies), inflammatory bowel diseases and autoimmune diseases. No etiology can be detected in half of the patients.
- ▶ Skin rashes resolve spontaneously in 1-3 months on average, but recurrence may be seen in some patients.

¹ MD., Memorial Ankara Hospital, Dermatology Clinic, nilgunstekin@gmail.com



headache, nausea, vomiting, and/or confusion.

Infectious Diseases

- ▶ Upper respiratory tract infections due to streptococcal species or rarely gastrointestinal *Yersinia* infection may be involved in the etiology of Sweet's syndrome. In case of doubt, the patients should be evaluated by an infectious diseases' specialist.
- ▶ In case of doubt, polymerase chain reaction (PCR) swab can be taken for SARS-Cov2 since an association was described between SS and the infection.

Hematology

- ▶ Hematological malignancies play a role in the etiology in 10-20% of patients. Examination for hematological malignancies, and further examination should be performed by a hematologist if there is any doubt.

Urology

- ▶ Among solid organ malignancies, genitourinary system carcinomas (e.g., renal cell carcinoma) may cause Sweet's syndrome.
- ▶ Screening for these carcinomas is recommended when necessary.

General Surgery

- ▶ Among solid organ malignancies, breast, and colon carcinomas may lead to Sweet's syndrome.
- ▶ For the purpose of screening for these carcinomas, mammography and fecal occult blood examination, and if necessary, further examination, are recommended.

Rheumatology

- ▶ Less frequently reported related diseases are rheumatoid arthritis, Behçet's Disease, autoimmune connective tissue diseases (e.g., Sjögren's syndrome, dermatomyositis, systemic lupus erythematosus, and recurrent polychondritis). Evaluations for autoimmune connective tissue diseases, and pathergy test may be requested from patients with clinical suspicion.

Clinical Pharmacology

- ▶ Sweet's syndrome may be triggered by certain drugs (e.g., trimethoprim-sulfamethoxazole, minocycline, furosemide, hydralazine, all-trans-retinoic acid). If mandatory use is required, it is recommended to replace these drugs with alternative options.

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SWEET SENDROMU (AKUT FEBRİL NÖTROFİLİK DERMATOZ) (L98.2)

Nilgün SOLAK¹

HATIRLA

- Klasik Sweet sendromu (SS), akut başlangıçlı olan ateş, halsizlik, eklem±kas ağrısı gibi konstitüsyonel semptomların ve eritematöz papül ve/veya plakların görüldüğü nötrofilik bir dermatozdur.
- Sweet sendromun diğer nötrofilik dermatozlara (nötrofilik ekrin hidradenit, eritema elevatum diutinum, subkorneal püstüler dermatoz ve pyoderma gangrenozum gibi) eşlik edebileceği akılda tutulmalıdır.
- Hastalıkta sıklıkla baş-boyun bölgesi ve üst ekstremiteler tutulur.
- Sweet sendromu kadınlarda daha sıktır (kadın/erkek: 4/1) ve genellikle 20-60 yaş arasında ortaya çıkar.
- Papül ve plaklardaki şiddetli ödem nedeniyle psödoveziküler veya psödopüstüler görünüm izlenebilir. Bazı hastalarda gerçek vezikülasyon, bül ve püstüller oluşabilir.
- Hastalık etiyojisinde enfeksiyonlar, gebelik, ilaçlar, maligniteler (sıklıkla hematolojik maligniteler), inflamatuvar bağırsak hastalıkları ve otoimmün hastalıklar yer alabilir. Hastaların yarısında herhangi bir etioloji saptanamamaktadır.
- Deri döküntüleri ortalama 1-3 ayda kendiliğinden düzelir, fakat bir kısım hastada nüks görülebilir.

¹ Dr., Memorial Ankara Hastanesi, Dermatoloji Kliniği, nilgunstekin@gmail.com



Nöroloji

- Sweet sendromunda ensefalit ve aseptik menenjit gelişebilir. Baş ağrısı, bulantı, kusma ve/veya bilinç bulanıklığı gibi semptomları olan hastalarda nörolojik değerlendirme yapılmalıdır.

Enfeksiyon Hastalıkları

- Streptokok türlerine bağlı üst solunum yolu enfeksiyonları veya nadiren gastrointestinal Yersinia enfeksiyonu Sweet sendromu etiolojisinde yer alabilir. Şüpheli durumda enfeksiyon hastalıkları uzmanının görüşü alınabilir.
- Şüpheli duruma SARS-Cov2 için polimeraz zincir reaksiyonu (PCR) sürüntüsü alınabilir.

Hematoloji

- Yaklaşık %10-20 hastada hematolojik maligniteler SS etiolojisinde rol oynamaktadır. Hastalarda hematolojik malignitelere yönelik tetkik, şüpheli varsa ileri inceleme yapılmalıdır. Hastalar hematoloji uzmanı tarafından değerlendirilmelidir.

Üroloji

- Solid organ malignitelerinden genitoüriner sistem karsinomları (örneğin renal hücreli karsinom) Sweet sendromuna neden olabilmektedir.
- Şüpheli durumda bu karsinomlara yönelik taramaların yapılması önerilir.

Genel Cerrahi

- Solid organ malignitelerinden meme ve kolon karsinomları Sweet sendromuna neden olabilmektedir.

- Bu karsinomlara yönelik tarama amacıyla mamografi ve gaitada gizli kan bakılması, gerekirse ileri inceleme yapılması önerilir.

Romatoloji

- SS ile daha nadir bildirilen ilişkili hastalıklar romatoid artrit, Behçet Hastalığı ve diğer otoimmün bağ doku hastalıklarıdır (örn: Sjögren sendromu, dermatomiyozit, sistemik lupus eritematozus ve tekrarlayan polikondrit). Klinik şüpheli olan hastalardan etiolojiye tetkikler ve paterji testi istenebilir. Hastalar romatoloji uzmanı tarafından değerlendirilmelidir.

Klinik Farmakoloji

- Sweet sendromu (örn; trimetoprim-sulfametaksazol, minosiklin, furosemid, hidralazin, all-trans-retinoik asit gibi) bazı ilaçlar tarafından tetiklenebilir. Zorunlu kullanım gerekiyorsa bu ilaçların alternatif tedavi seçenekleriyle değiştirilmesi önerilir.

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