

# 30. Bölüm

## PULMONER ALVEOLAR PROTEİNOZİS

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### TANIM VE GİRİŞ

Pulmoner alveolar proteinozis (PAP), sürfaktanın alveollerde ve terminal hava yollarında biriktiği nadir görülen diffüz bir akciğer hastalığıdır<sup>1</sup>. İlk olarak 1958'de Rosen ve arkadaşları<sup>2</sup> tarafından alveollerde lipid bakımından zengin periyodik asit-Schiff (PAS) pozitif materyal birikimiyle seyreden bir bozukluk olarak bildirilmiştir. PAP'ta esas sorun sürfaktan üretiminde ve/veya ortamdan uzaklaştırılmasında meydana gelen bir dizi bozukluğun oluşmasıdır. Bunun sonucu alveolar sahada sürfaktan fosfolipid ve apoproteinlerinden oluşan lipoproteinöz materyal birikir. PAP'lı olgular asemptomatik olabileceği gibi ilerleyici dispne kliniği, artan oksijen ihtiyacı ve solunum yetmezliğine kadar değişik şekillerde prezente olabilirler<sup>3</sup>.

### SINIFLAMA

PAP yerine literatürde, alveolar lipoproteinososis, alveolar fosfolipidozis ve alveolar fosfolipoproteinozis gibi diğer isimlendirmeler de kullanılır. PAP patogenezinin açıklanmasına yönelik olarak, 1958'deki ilk vaka sunumundan sonra yaklaşık 40 yıl boyunca anlamlı bir ilerleme gösterilememiştir. Granülosit makro-

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olmakla beraber özellikle otoimmün PAP ta GM-CSF (subkutan, inhalasyon) tedavileri söz konusudur çalışmalar umut vericidir. Pulmoner fibrosis gelişimi kötü prognoz göstergesi olup, olgudan olguya klinik ve prognoz değişkenlik gösterir.

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