BÖLÜM 35

MUKOZAL MALİGN MELANOM

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EPİDEMİYOLOJİ

Melanomlar, pigment hücreleri olan melanositlerden kaynaklanan malign tümörlerdir. Melanositler, nöral krest hücrelerinden elde edilir ve çeşitli bölgelere göç ederler (1). Bu nedenle en yaygın deride görülmesine rağmen, melanositler vücudun diğer organlarında da bulunabilir ve kutanöz olmayan melanomlara yol açabilir. Mukozal melanomlar nadir bir varyanttır. Tüm kanser tanılarının yaklaşık %0,03'ünü ve tüm melanomların %5'ini oluşturur. Yaklaşık %3-%5'i uveal, kalan %1-%2'si ise mukozaldır. Mukozal melanomlar, herhangi bir mukozal yüzeyden en tipik olarak melanositlerin bulunduğu solunum, gastrointestinal ve genitoüriner yolların mukozal epitelinden kaynaklanabilir (2, 3). Mukozal melanomların çoğu baş ve boyunda (%55; konjonktival, sinonazal ve orofaringeal) ve daha az sıklıkla anorektal (%24) ve vulvovajinal (%18) bölgelerde bulunur. Çok daha az sıklıkla idrar yollarında, solunum sisteminde, özefagusta, midede, barsaklarda, safra kesesinde ve servikste de bulunabilmektedir (4, 5). Artan kutanöz melanom insidansının aksine, mukozal melanom insidansının sabit kaldığına inanılmaktadır (6).

Mukozal melanomlu hastalar genel olarak kutanöz melanoma göre 10 ile 20 yıl daha geç yaşta ortaya çıkar. Vakaların çoğu 50-80 yaşları arasında olup tanıdaki medyan yaş 70'tir (7, 8). Kadınlarda erkeklere göre daha sık tanı konulmaktadır, birçok raporda vulvovajinal hastalık vakaları nedeniyle iki kata kadar daha sık teşhis edilmektedir buna karşılık baş ve boyun mukozal melanom alt tiplerinde hafif bir erkek baskınlığı öne sürülmüştür (3, 9, 10).

Mukozal melanom nadir bir melanom türü olmasının yanında oldukça da kötü prognozlu bir seyir gösterebilmektedir. Tahmini 5 yıllık genel sağkalım %25'dir (11). Mukozal melanomun etiyolojisi halen bilinmemektedir (12). Ayrıca, human papilloma virüsleri, herpes virüsleri ve poylomavirüs ile de bir ilişki saptanamamıştır. Sigaranın rolü bilinmemektedir (10, 13). Anatomik orijinleri nedeniyle,

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progrese olup yukarıda sayılan mutasyonların hiçbirini barındırmayan hastalarda ise kemoterapi halen geçerli bir seçenektir. Karboplatin+paklitaksel öncelikli kemoterapi seçeneği olup bu tedaviye bevacizumab eklenebilir. Bevacizumabın bu tedaviye eklenmesi FAZ-II çalışmada sağkalım yararı göstermiştir (44). Cerrahi veya radyoterapi palyatif amaçla hasta bazlı kullanılabilir.

SONUÇ

Mukozal melanom, etiyolojisi bilinmeyen, agresif davranış gösteren ve daha kötü prognozla seyreden melanom alt grubudur. Baş ve boyun mukozal melanomları en yaygın olanıdır. Mevcut tedavi seçenekleri kısıtlıdır. Kutanöz melanom alt tipinde olduğu gibi lokal hastalıkta geniş lokal eksizyonla cerrahi ilk tercih olup adjuvan tedavinin sağkalım üzerindeki etkisi tartışmalıdır. Radyoterapi daha iyi lokal kontrol sağlayabilmesine rağmen sağkalım üzerine etkisi bilinmemektedir. Metastatik hastalıkta başlangıç tedavisi olarak kombinasyon immunoterapi tedavisi veya kombinasyon immunoterapisine uygun değilse tek ajan immunoterapi önerilmektedir. Mukozal melanomda da diğer melanomlarda olduğu gibi immunoterapilere klinik olumlu yanıtlar sağlanmıştır. Son veriler, önümüzdeki yıllarda immünoterapi tedavileri ile sağkalımda iyileşmeler olabileceğini göstermektedir.

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