BÖLÜM 24

Memede Dermatofibrosarkomaprotuberans

Kazım GEMİCİ[1]

Dermatofibrosarkomaprotuberans (DFSP) ilk olarak 1924'de Darier ve Ferrand tarafından tanımlanmış ve daha sonra 1925'te Hoffmann tarafından adlandırılmıştır(1). DFSP, nadir, lokal olarak agresif bir deri tümörüdür. Yavaş, infiltratif büyümesi ve cerrahi rezeksiyon sonrası belirgin lokal rekürrens eğilimi ile karakterizedir(2). Bu tümör bir fibrosarkomdur, daha doğrusu kutanöz bir yumuşak doku sarkomudur. Pek çok açıdan DFSP, iyi huylu bir tümör gibi davranmasına rağmen vakaların %2-5'inde metastaz yapabildiğinden malign potansiyele sahip olduğu düşünülmelidir(3). DFSP insidansı yılda bir milyonda 0,8-5 arasında olup, tüm malignitelerin% 0,1'ni oluşturduğu tahmin edilmektedir(4). Moleküler çalışmalar, kromozo-mal translokasyonun vakaların %90'ından fazlasında olduğunu ortaya koyar, bunlar 17q22 ve 22q13'ü içerir.Translokasyon, COL1A1 genlerini trombosit kaynaklı büyüme faktörü beta (PDGFβ) ile, genellikle bir halka kromozomunun oluşumuyla birleştirir. Bu gen, yapısal olarak ifade edilen PDGF reseptörüne bağlanan ve DFSP hücrelerinin büyümesini uyarmak için bir otokrin faktör görevi gören bir füzyon proteinini kodlar(5). DFSP, CD34'ün güçlü ve yaygın expresyonunu gösterir; ayrıca vimentin, nestin ve apolipoprotein D pozitiftiliği gösterirken ve sitokeratinler, düz kas aktin (SMA), S100, CD56 Faktör XIIIa, Stromelysin 3, and Cathepsin K negatiftir (13-4). Hastalığın %10-20'sinden lokal travmanın sorumlu olduğu bildirilmektedir(6). Çoğu gövdede (% 42), ardından üst ekstremitelerde (% 23), alt ekstremitelerde (% 18) ve baş ve boyunda (% 16) görülür (7). En sık 30 yaşın üzerindeki yetişkinlerde görülür ve tüm yumuşak doku sarkomlarının% 2-6'sını oluşturur. Erkek:Kadın oranı 1:1'dir. Tedavinin temel dayanağı geniş lokal eksizyondur, ancak Mohs mikrografik cerrahi (MMS) alternatif bir yaklaşım olarak ortaya çıkmaktadır(8).

DPFS genellikle kemoterapi ve radyoterapiye dirençlidir, bu nedenle tam cerrahi rezeksiyon veya geniş rezeksiyon tedavinin altın standartı olarak kabul edilmektedir(9). Son yıllarda (MMS) ile nüks oranlarının% 0,6 ile % 6 arasında olduğu söylenmektedir(10). Yetersiz rezeksiyon olgularında lokal nüks oranı %20

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nişliği sınırlanmaktadır. Bu cerrahi dezavantaj, doğal olarak erken nükslere yol açan güvenli cerrahi sınırı daraltır. Cerrahi tedavi öncesi ve sonrası daha etkili bir yöntem olmadıkça, biz cerrahların gerekirse kaburga rezeksiyonu yapmak için agresif cerrahiyi tercih etmek zorunda kalacağı bir gerçektir.

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