Renal Anjiomyolipoma Sekonder Retroperitoneal Kanama: Wunderlich Sendromu

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GİRİŞ

Renal anjiomyolipom, dismorfik damarsal yapılar, düz kas ve yağ hücrelerinden oluşan iyi huylu böbrek tümörlerindendir. Yoğun şekilde mezenkimal elemanlar ihtiva etmesinden dolayı renal hamartom olarak da isimlendirilir. Batın görüntülemelerinin günümüzde yaygınlaşması ile birlikte tanı koyma oranları son on yılda ivmeli bir şekilde artış göstermiştir. Anjiomyolipom tüm böbrek tümörlerinin yaklaşık %10'unu oluşturmaktadır. Otopsi serilerinde ise tüm toplumda %0.3-%0.13 arasında görüldüğü bildirilmiştir.[1] Orta yaş kadınlarda daha sık görülen anjiomyolipom, genelde sporadik ve asemptomatik paterndedir. Sporadik renal anjiomyolipomlar genelde insidental olarak tespit edilir ve yavaş büyüme paternine sahiptir. Renal anjiomyolipom tespit edilen hastaların yaklaşık %20 'sinde ise otozomal dominant geçişli tubero-skleroz kompleks hastalığı veya pulmoner lenfanjiyoleiomyomatoz mevcuttur. Tubero-skleroz kompleks olan hastaların %70'inde ise renal anjiomyolipom mevcuttur.[2] Böbrek tümörleri arasında spontan perirenal kanama potansiyeline en fazla anjiomyolipom sahiptir. Renal anjiomyolipomun büyüklüğü ile doğru orantılı olarak spontan rüptür ve kanama gelişebilir. Ayrıca büyüklüğü artan anjiomyolipomlarda kanama potansiyeli olan mikro ve makro anevrizmalar oluşabilir. Bu kanamalar nadiren ani bir şekilde gelişip hayatı tehdit edecek potansiyele sahip olur. Anjiomyolipoma sekonder kanamaların %10'unda Wunderlich sendromu olarak isimlendirilen masif spontan ret-

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tedavisinde kitle boyutu, semptomların varlığı, hastanın doğurganlık çağında olup olmaması ve kanama potansiyeli son derece önemlidir. Anjio-embolizasyon ilk önerilen tedavi yaklaşımıdır ancak diğer böbrek koruyucu tedavi seçenekleri ve instabil hastalarda radikal nefrektomi tercih edilebilir.

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