



5.1.3. RENAL HEMANJİOBLASTOM

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5.1.3.1. Tanım

- ▶ Renal hemanjioblastomlar (RHB), santral sinir sisteminin hemanjioblastomuna benzer şekilde kapiller ve stromal hücrelerden oluşan benign bir tümördür

5.1.3.2. Köken/Patogenezi

- ▶ Bilinen bir etyolojisi ya da yatkınlık oluşturan bir durum tanımlanmamıştır.
- ▶ VHL Sendromu ile ilişkili değil (1).
- ▶ Renal hemanjioblastomun mTOR yolağı mutasyonlarıyla yakın ilişkisi saptanmıştır ve fibromyomatoz stromalı RCC'ler içerisinde ayırıcı tanıda yer alması gerektiği düşünülmektedir (2-3).

5.1.3.3. Klinik

5.1.3.3.1. Epidemiyoloji

- ▶ Oldukça nadir (Literatürde ~30 vaka) (4).
- ▶ İlk RHB vakası Nonaka ve ark. tarafından rapor edilmiştir (5).
- ▶ K: E=1 (2).
- ▶ 16-71 yaş (ortanca 48) (4).

5.1.3.3.2. Semptom ve İşaretler

- ▶ Çoğu rastlantısal.
- ▶ Hematüri (%24), alt sırt ve karın ağrısı (%12).
- ▶ Hastaların sadece birinde ateş ve kilo kaybı gibi sistemik semptomlar bildirilmiştir. (4,6,7).

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