



5.2.2. KONJENİTAL MEZOBLASTİK NEFROM

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5.2.2.1. Tanım

- ▶ Çoğunlukla renal hilusta yerleşimli, *NTRK* füzyonu veya *EGFR* ITD gibi özgün moleküler özelliklere sahip, pre/perinatal dönemin (myo)fibroblastik farklılaşma gösteren renal mezenkimal neoplazisidir.

5.2.2.2. Köken/Patogenez

- ▶ Tümör oluşumunun tercihen mezenkimal progenitör hücrelerde meydana geldiği öne sürülmüştür (1).
- ▶ (Myo)Fibroblastik farklılaşma gösterir.
- ▶ Tirozin kinaz aktivasyonu (başta *EGFR* veya *NTRK*) söz konusudur.
- ▶ Mikst varyant, immünohistokimyasal özellikleri klasik varyant, moleküler özellikleri ise sellüler varyant ile uyumlu ara bir formdur.
- ▶ *MAPK/ERK* yolak aktivasyonu gösterilmiştir (2).

5.2.2.3. Klinik

5.2.2.3.1. Epidemiyoloji

- ▶ Tüm pediatrik renal tümörlerin %3-5'ini oluşturur.
- ▶ %90'ı <1 yaş, çoğu ilk 3 ay içinde tanı alır.
- ▶ İlk 2 yaş içindeki renal tümörlerin yaklaşık %90'ını oluşturur (3,4).
- ▶ Erkeklerde daha sıktır.

5.2.2.3.2. Semptom ve İşaretler

- ▶ %15 prenatal dönemde tespit edilebilir; polihidramniyoz, prematür doğum ve neonatal hipertansiyon görülür. (5)

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