25. BÖLÜM

ÇOCUKLUK ÇAĞININ MEZENKİMAL BÖBREK TÜMÖRLERİ

Gökçe AŞKAN [1]

BERRAK HÜCRELİ SARKOM

İlk kez 1970 yılında Kidd ve arkadaşları tarafından, kemiğe metastaz yapma eğilimi sebebiyle Wilms tümörü (nefroblastom) nden ayrı bir antite olduğu öne sürülmüş ve 1978 yılından itibaren de böbreğin berrak hücreli sarkomu olarak tanımlanmıştır (1, 2). Wilms tümöründen sonra çocukluk çağının ikinci en sık görülen malign tümörüdür ve çocukluk çağı böbrek tümörlerinin yaklaşık %3-4' ünü oluşturur (3-6). Nefroblastom ilişkili sendromlarla ya da embryonik kalıntılarla herhangi bir ilişkisi bulunmamaktadır. Ortalama görülme yaşı 36 ay olup erkek çocuklarda kız çocuklara oranla iki kat daha fazla görülür (4, 7). Karın ağrısı, karında kitle ve hematüri en sık görülen semptomlardır (8).

Makroskopi

Böbreğin berrak hücreli sarkomu makroskopik olarak ortalama 11 cm çapta, tek taraflı ve renal medullada yerleşim gösteren, soliter, iyi sınırlı ancak kapsülsüz görünümdedir. Kesit yüzü kirli bej renkte, yumuşak kıvamda olup, yer yer kistik ve mukoid alanlar içerebilir (4, 5).

Histopatoloji

Mikroskopik olarak klasik, miksoid, sklerozan, sellüler, epiteloid (asiner ya da trabeküler), palizatlaşan ve iğsi hücreli patern olmak üzere çok farklı morfolojik paterne sahip olduğu için yanlış tanı konulabilmektedir (4, 9). Olguların yaklaşık %90' ında birbirinden fibrovasküler septalarla ayrılmış,

[1] Uzm. Dr., Rize Recep Tayyip Erdoğan Üniversitesi Eğitim ve Araştırma Hastanesi, Tıbbi Patoloji
gokceaskan@gmail.com

Mikroskopi

Mikroskopik olarak osteoid zemin ve etrafında dizilim gösteren osteoblastik ve benign görünümde iğsi hücrelerle karakterizedir. Yaş arttıkça osteoid zeminde de artış izlenir. İğsi hücreler invaziv büyüme paterni göstererek glomerül ve tubül yapılarını ortadan kaldırabilir. İğsi hücrelerde Vimentin ekpresyonu izlenirken, EMA ya da sitokeratin negatiftir. Bu hücrelerde aynı zamanda WT-1 ekspresyonu görülebildiği için Wilms tümörüyle karıştırılmamalıdır (7).

Ayırıcı Tanı

Ayırıcı tanıda Wilms tümörü ve konjenital mezoblastik nefrom yer alır. Wilms tümörünün 3 yaş civarında görülmesi ve en sık izlenen klinik bulgunun karında kitle olması tanıda yardımcıdır. Konjenital mezoblastik nefrom ise 3 ay ve altı çocuklarda görülmekle birlikte olgularda polihidroamniyoz eşlik eder (64).

Genetik

Trizomi 4 varlığı kemikleşen böbrek tümörü için şimdiye kadar bilinen tek moleküler özellik olmakla birlikte özellikle kalsifikasyon bulguları gösteren Wilms tümörünün ayırıcı tanısında yardımcıdır (7, 65).

Prognoz

Prognoz oldukça iyi olup tedavide genellikle parsiyel ya da total nefrektomi tercih edilmekte ve kemoterapi ya da radyoterapiye ihtiyaç duyulmamaktadır. Parsiyel nefrektomi sonrası bile hastalarda uzun dönemde nüks ya da metastaz bulgusu izlenmemektedir. Bu nedenle doğru tanı oldukça önemlidir (66).

KAYNAKLAR

1. Kidd J. Exclusion of certain renal neoplasms from the category of Wilms' tumor. Am J Pathol. 1970;59(16a).
2. Beckwith JB. Wilms' tumor and other renal tumors of childhood: a selective review from the National Wilms' Tumor Study Pathology Center. Hum Pathol. 1983;14(6):481-92.
3. Argani P, Beckwith JB. Renal Neoplasms of Childhood. In: Millis SE, editor. Sternberg`s Diagnostic Surgical Pathology. 2. 6th ed: LWW; 2015.
4. Argani P, Perlman EJ, Breslow NE, et al. Clear cell sarcoma of the kidney: a review of 351 cases from the National Wilms Tumor Study Group Pathology Center. Am J Surg Pathol. 2000;24(1):4-18.
5. Gooskens SL, Furtwangler R, Vujanic GM, et al. Clear cell sarcoma of the kidney: a review. Eur J Cancer. 2012;48(14):2219-26.
6. Huang CC, Cutcliffe C, Coffin C, et al. Classification of malignant pediatric renal tumors by gene expression. Pediatr Blood Cancer. 2006;46(7):728-38.
7. Argani P, Dehner L, Leuschner I. Mesenchymal tumors occuring mainly in children. In: Moch H, Humphrey PA, Ulbright TM, Reuter VE, editors. WHO Classifications of Tumors of the Urinary System and Male Genital Organs. 4th ed. Lyon: IARC; 2016. p. 54-8.

8. Sebire NJ, Vujanic GM. Paediatric renal tumours: recent developments, new entities and pathological features. Histopathology. 2009;54(5):516-28.
9. Tournade MF, Com-Nougue C, de Kraker J, et al. Optimal duration of preoperative therapy in unilateral and nonmetastatic Wilms' tumor in children older than 6 months: results of the Ninth International Society of Pediatric Oncology Wilms' Tumor Trial and Study. J Clin Oncol. 2001;19(2):488-500.
10. Argani P, Pawel B, Szabo S, et al. Diffuse Strong BCOR Immunoreactivity Is a Sensitive and Specific Marker for Clear Cell Sarcoma of the Kidney (CCSK) in Pediatric Renal Neoplasia. Am J Surg Pathol. 2018;42(8):1128-31.
11. Argani P, Kao YC, Zhang L, et al. Primary Renal Sarcomas With BCOR-CCNB3 Gene Fusion: A Report of 2 Cases Showing Histologic Overlap With Clear Cell Sarcoma of Kidney, Suggesting Further Link Between BCOR-related Sarcomas of the Kidney and Soft Tissues. Am J Surg Pathol. 2017;41(12):1702-12.
12. Mirkovic J, Calicchio M, Fletcher CD, et al. Diffuse and strong cyclin D1 immunoreactivity in clear cell sarcoma of the kidney. Histopathology. 2015;67(3):306-12.
13. Jet Aw S, Hong Kuick C, Hwee Yong M, et al. Novel Karyotypes and Cyclin D1 Immunoreactivity in Clear Cell Sarcoma of the Kidney. Pediatr Dev Pathol. 2015;18(4):297-304.
14. Kao YC, Sung YS, Zhang L, et al. BCOR Overexpression Is a Highly Sensitive Marker in Round Cell Sarcomas With BCOR Genetic Abnormalities. Am J Surg Pathol. 2016;40(12):1670-8.
15. Aldera AP, Pillay K. Clear Cell Sarcoma of the Kidney. Arch Pathol Lab Med. 2020;144(1):119-23.
16. Wong MK, Ng CCY, Kuick CH, et al. Clear cell sarcomas of the kidney are characterised by BCOR gene abnormalities, including exon 15 internal tandem duplications and BCOR-CCNB3 gene fusion. Histopathology. 2018;72(2):320-9.
17. Uddin N, Minhas K, Abdul-Ghafar J, et al. Expression of cyclin D1 in clear cell sarcoma of kidney. Is it useful in differentiating it from its histological mimics? Diagn Pathol. 2019;14(1):13.
18. Dhamne S, Brown RE, Covinsky M, et al. Clear cell sarcoma of kidney: morphoproteomic analysis reveals genomic correlates and therapeutic options. Pediatr Dev Pathol. 2013;16(1):20-7.
19. O'Meara E, Stack D, Lee CH, et al. Characterization of the chromosomal translocation t(10;17)(q22;p13) in clear cell sarcoma of kidney. J Pathol. 2012;227(1):72-80.
20. Lee JE, Spiegelman D, Hunter DJ, et al. Fat, protein, and meat consumption and renal cell cancer risk: a pooled analysis of 13 prospective studies. J Natl Cancer Inst. 2008;100(23):1695-706.
21. Seibel NL, Chi YY, Perlman EJ, et al. Impact of cyclophosphamide and etoposide on outcome of clear cell sarcoma of the kidney treated on the National Wilms Tumor Study-5 (NWTS-5). Pediatr Blood Cancer. 2019;66(1):e27450.
22. Furtwangler R, Gooskens SL, van Tinteren H, et al. Clear cell sarcomas of the kidney registered on International Society of Pediatric Oncology (SIOP) 93-01 and SIOP 2001 protocols: a report of the SIOP Renal Tumour Study Group. Eur J Cancer. 2013;49(16):3497-506.
23. Gooskens SL, Furtwangler R, Spreafico F, et al. Treatment and outcome of patients with relapsed clear cell sarcoma of the kidney: a combined SIOP and AIEOP study. Br J Cancer. 2014;111(2):227-33.
24. Weaver J, Ho T, Lang A, et al. Bladder Recurrence of Clear Cell Sarcoma of the Kidney Seven Years After Initial Presentation. Urology. 2017;106:193-5.
25. Gooskens SL, Graf N, Furtwangler R, et al. Position paper: Rationale for the treatment of children with CCSK in the UMBRELLA SIOP-RTSG 2016 protocol. Nat Rev Urol. 2018;15(5):309-19.
26. Vujanic GM, Sandstedt B, Harms D, et al. Rhabdoid tumour of the kidney: a clinicopathological study of 22 patients from the International Society of Paediatric Oncology (SIOP) nephroblastoma file. Histopathology. 1996;28(4):333-40.

27. Zhao G, Na R, Yang Y, et al. Pure malignant rhabdoid tumor of the left kidney in an adult: A case report and review of the literature. Oncol Lett. 2013;5(5):1481-4.
28. Shukla D, Pradhan A, Bhardwaj M, et al. Malignant rhabdoid tumor of kidney and brain in an infant. Indian Pediatr. 2015;52(1):65-6.
29. Tomlinson GE, Breslow NE, Dome J, et al. Rhabdoid tumor of the kidney in the National Wilms' Tumor Study: age at diagnosis as a prognostic factor. J Clin Oncol. 2005;23(30):7641-5.
30. Przybycin CG, McKenney JK, Reynolds JP, et al. Rhabdoid differentiation is associated with aggressive behavior in renal cell carcinoma: a clinicopathologic analysis of 76 cases with clinical follow-up. Am J Surg Pathol. 2014;38(9):1260-5.
31. Rao Q, Xia QY, Shen Q, et al. Coexistent loss of INI1 and BRG1 expression in a rhabdoid renal cell carcinoma (RCC): implications for a possible role of SWI/SNF complex in the pathogenesis of RCC. Int J Clin Exp Pathol. 2014;7(4):1782-7.
32. Horazdovsky R, Manivel JC, Cheng EY. Surgery and actinomycin improve survival in malignant rhabdoid tumor. Sarcoma. 2013;2013:315170.
33. van den Heuvel-Eibrink MM, van Tinteren H, Rehorst H, et al. Malignant rhabdoid tumours of the kidney (MRTKs), registered on recent SIOP protocols from 1993 to 2005: a report of the SIOP renal tumour study group. Pediatr Blood Cancer. 2011;56(5):733-7.
34. Peng HQ, Stanek AE, Teichberg S, et al. Malignant rhabdoid tumor of the kidney in an adult: a case report and review of the literature. Arch Pathol Lab Med. 2003;127(9):e371-3.
35. Bolande RP, Brough AJ, Izant RJ, Jr. Congenital mesoblastic nephroma of infancy. A report of eight cases and the relationship to Wilms' tumor. Pediatrics. 1967;40(2):272-8.
36. Gooskens SL, Houwing ME, Vujanic GM, et al. Congenital mesoblastic nephroma 50 years after its recognition: A narrative review. Pediatr Blood Cancer. 2017;64(7).
37. Anakievski D, Ivanova P, Kitanova M. Laparoscopic management of congenital mesoblastic nephroma- case report. Urol Case Rep. 2019;26:100979.
38. Wang ZP, Li K, Dong KR, et al. Congenital mesoblastic nephroma: Clinical analysis of eight cases and a review of the literature. Oncol Lett. 2014;8(5):2007-11.
39. Santos LG, Carvalho Jde S, Reis MA, et al. Cellular congenital mesoblastic nephroma: case report. J Bras Nefrol. 2011;33(1):109-12.
40. Malkan AD, Loh A, Bahrami A, et al. An approach to renal masses in pediatrics. Pediatrics. 2015;135(1):142-58.
41. Takahashi H, Ohkuchi A, Kuwata T, et al. Congenital mesoblastic nephroma: Its diverse clinical features - A literature review with a case report. J Obstet Gynaecol. 2016;36(3):340-4.
42. Leclair MD, El-Ghoneimi A, Audry G, et al. The outcome of prenatally diagnosed renal tumors. J Urol. 2005;173(1):186-9.
43. Ko SM, Kim MJ, Im YJ, et al. Cellular mesoblastic nephroma with liver metastasis in a neonate: prenatal and postnatal diffusion-weighted MR imaging. Korean J Radiol. 2013;14(2):361-5.
44. Fuchs IB, Henrich W, Kalache KD, et al. Prenatal sonographic features of a rhabdoid tumor of the kidney. Ultrasound Obstet Gynecol. 2004;23(4):407-10.
45. Fuchs IB, Henrich W, Brauer M, et al. Prenatal diagnosis of congenital mesoblastic nephroma in 2 siblings. J Ultrasound Med. 2003;22(8):823-7; quiz 8-9.
46. Bayindir P, Guillerman RP, Hicks MJ, et al. Cellular mesoblastic nephroma (infantile renal fibrosarcoma): institutional review of the clinical, diagnostic imaging, and pathologic features of a distinctive neoplasm of infancy. Pediatr Radiol. 2009;39(10):1066-74.
47. Campos A, Chamyan G, Gunasekaran S, et al. Pathology teach and tell: cellular mesoblastic nephroma. Pediatr Pathol Mol Med. 2001;20(5):439-44.
48. Chaudry G, Perez-Atayde AR, Ngan BY, et al. Imaging of congenital mesoblastic nephroma with pathological correlation. Pediatr Radiol. 2009;39(10):1080-6.
49. El Demellawy D, Cundiff CA, Nasr A, et al. Congenital mesoblastic nephroma: a study of 19 cases using immunohistochemistry and ETV6-NTRK3 fusion gene rearrangement. Pathology. 2016;48(1):47-50.

50. Vujanic GM, Kelsey A, Mitchell C, et al. The role of biopsy in the diagnosis of renal tumors of childhood: Results of the UKCCSG Wilms tumor study 3. Med Pediatr Oncol. 2003;40(1):18-22.
51. Lowe LH, Isuani BH, Heller RM, et al. Pediatric renal masses: Wilms tumor and beyond. Radiographics. 2000;20(6):1585-603.
52. Kurian JJ, Nongpiur KRL, Jehangir S. Use of Pretherapy Core Biopsy in the Diagnosis of Pediatric Renal Tumors. J Indian Assoc Pediatr Surg. 2018;23(2):66-9.
53. Sandberg AA, Bridge JA. Updates on the cytogenetics and molecular genetics of bone and soft tissue tumors. desmoplastic small round-cell tumors. Cancer Genet Cytogenet. 2002;138(1):1-10.
54. Jehangir S, Kurian JJ, Selvarajah D, et al. Recurrent and metastatic congenital mesoblastic nephroma: where does the evidence stand? Pediatr Surg Int. 2017;33(11):1183-8.
55. Loeb DM, Hill DA, Dome JS. Complete response of recurrent cellular congenital mesoblastic nephroma to chemotherapy. J Pediatr Hematol Oncol. 2002;24(6):478-81.
56. England RJ, Haider N, Vujanic GM, et al. Mesoblastic nephroma: a report of the United Kingdom Children's Cancer and Leukaemia Group (CCLG). Pediatr Blood Cancer. 2011;56(5):744-8.
57. Chatten J, Cromie WJ, Duckett JW. Ossifying tumor of infantile kidney: report of two cases. Cancer. 1980;45(3):609-12.
58. Vaillancourt B, Oligny L, Dery J, et al. Ossifying Renal Tumor of Infancy: Report of a Case With Positive WT1 Immunohistochemistry and High Mitotic Index and Review of the Literature. Pediatr Dev Pathol. 2017;20(6):511-6.
59. Guan W, Yan Y, He W, et al. Ossifying renal tumor of infancy (ORIT): The clinicopathological and cytogenetic feature of two cases and literature review. Pathol Res Pract. 2016;212(11):1004-9.
60. Schelling J, Schroder A, Stein R, et al. Ossifying renal tumor of infancy. J Pediatr Urol. 2007;3(3):258-61.
61. Lee SH, Choi YH, Kim WS, et al. Ossifying renal tumor of infancy: findings at ultrasound, CT and MRI. Pediatr Radiol. 2014;44(5):625-8.
62. Seixas-Mikelus SA, Khan A, Williot PE, et al. Three-month-old boy with juvenile granulosa cell tumor of testis and ossifying renal tumor of infancy. Urology. 2009;74(2):311-3.
63. Hoglund HH, Kellner MW, Korber F, et al. Ossifying renal tumor of infancy (ORTI) - a rare diagnosis. Klin Padiatr. 2011;223(3):178-9.
64. Ritchey M, Daley S, Shamberger RC, et al. Ureteral extension in Wilms' tumor: a report from the National Wilms' Tumor Study Group (NWTSG). J Pediatr Surg. 2008;43(9):1625-9.
65. Liu J, Guzman MA, Pawel BR, et al. Clonal trisomy 4 cells detected in the ossifying renal tumor of infancy: study of 3 cases. Mod Pathol. 2013;26(2):275-81.
66. Hu J, Wu Y, Qi J, et al. Ossifying renal tumor of infancy (ORTI): a case report and review of the literature. J Pediatr Surg. 2013;48(2):e37-40.