Medical Quiz Answers

A1. CT scan with intravenous contrast of the abdomen.

A2. Large mass arising from the upper pole of the left kidney.

A3. Wilms tumor. Left nephrectomy.

DISCUSSION

Nephroblastoma or Wilms tumor is the second most common intraabdominal malignancy and the fifth most common cancer in children. Wilms tumor is the most frequent tumor of the kidney (more than 95% of all tumors of the kidney in children) and it accounts for 6% of all malignancies in children[1].

The differential diagnosis of nephroblastoma (Wilms tumor) includes other renal cancers in children, such as neuroblastoma, malignant rhabdoid tumor, clear cell sarcoma, and renal cell carcinoma, which have less favorable outcomes[1-3].

Children with congenital anomalies, such as Beckwith-Wiedemann syndrome, Denys-Drash syndrome, WAGR syndrome, or isolated hemihypertrophy are at higher risk of developing Wilms tumor[3].

The incidence of Wilms tumor peaks between ages 2 and 5 years; 95% of the cases are diagnosed before the age of 10 years. An abdominal mass is the most frequent initial sign, followed by hematuria. Some children may have arterial hypertension due to the pressure on the renal artery caused by the renal tumor. Other clinical presentations include anorexia, asthenia, weight loss and fever[4].

The management of pediatric renal tumors consists of advanced multimodal treatments, which include surgery and chemotherapy with or without radiotherapy depending on the stage of the disease. The 5-year survival rate has increased significantly in the past decade, from 25% to 90%[5].

Staging and management of Wilms tumor in children have been established by two collaborative groups: the International Society for Pediatric Oncology (SIOP) in Europe and the National Wilms Tumor Study Group (NWTSG) in the United States[6].

The prognostic factors in Wilms tumor are based on the histological characteristics and the staging. The histology of Wilms tumor is classified into three groups: low, intermediate and high risk. Anaplastic histology is the most unfavorable outcome. Clear cell sarcoma histology, blastemal histology, and renal rhabdoid histology are other high-risk groups. The stage of the tumor is also an important prognostic survival factor[7,8].

The survival of children with Wilms tumor is generally excellent. The survival rate is 95% for patients with stages I and II, 75–80% for patients with stage III, and 65–75% for patients with stage IV. Recurrent Wilms tumor in children depends on the histological types. An incidence of 50% in those with anaplastic histology had a recurrence of tumor, compared to 15% of patients with favorable histology. The most common sites of recurrence are the lungs, liver and tumor bed. Liver metastases in children with Wilms tumor have a poor prognosis compared to those with lung metastases[7,8].

CONCLUSION

Wilms tumor (nephroblastoma) is the most common solid renal tumor in children. It is diagnosed based on clinical and radiological features. The management of Wilms tumor in pediatric patients includes surgery, chemotherapy and radiotherapy, based on the staging of the disease.

Potential Conflicts of Interest: None.

Competing Interest: None.

Sponsorship: None.

Acceptance Date: 19 October 2019.

Ethical Approval: Approved by the Department of Pediatrics, Salmaniya Medical Complex, Bahrain.

REFERENCES

1. Visser YT, Uys R, van Zyl A, et al. Nephroblastoma—A 25-Year Review of a South African Unit. J Med Life 2014; 7(3):445-9.
2. Kembhavi SA, Qureshi S, Vora T, et al. Understanding the Principles in Management of Wilm's Tumor: Can Imaging Assist in Patient Selection? Clin Radiol 2013; 68(7):646-53.
3. Dome JS, Perlman EJ, Graf N. Risk Stratification for Wilm's Tumor: Current Approach and Future Directions. Am Soc Clin Oncol Educ Book 2014; 215-23.
4. Maureen D, Noorulhuda J, Kieran M. Neuroblastoma and Nephroblastoma: A Radiological Review. Cancer Imaging 2015; 15(1): 5.
5. Dome JS, Graf N, Geller JI, et al. Advances in Wilm's Tumor Treatment and Biology: Progress through International Collaboration. J Clin Oncol 2015; 33(27): 2999-3007.
6. Taran K, Sitkiewicz A, Kobos J. Histoclinical Study of Nephroblastoma in Relation to Current and Previous SIOP Classification of Renal Tumors in Childhood. Pol J Pathol 2010; 61(4):234-9.
7. Gill L. Renal Tumors: Long-Term Outcome. Pediatr Nephrol 2012; 27(6): 911–916.
8. Fawkner-Corbett DW, Howell L, Pizer BL, et al. Wilm's' Tumor—Lessons and Outcomes—A 25-Year Single Center UK Experience. Pediatr Hematol Oncol 2014; 31(5):400-8.