Long Term Survival in Pediatric Renal Cell Carcinoma Despite Multiple Relapses: A Case Study

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INTRODUCTION

- Although common in adults, Renal cell carcinoma (RCC) is extremely rare in pediatric populations, comprising less than 5% of malignant renal tumors
- RCC presents with a triad of symptoms, however this is not a common case presentation
  - Abdominal Pain/palpable mass
  - Hematuria
  - Weight loss
- There are several types of RCC, with the clear cell type being the most common
  - The second common type is the papillary type, of which there is a type I and type II based on cellular morphology
- Papillary RCC is associated with Xp11.2 translocation/TFE-3 gene fusion

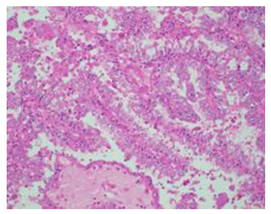

Figure 1
H&E stain: Highlights the papillary architecture, the thin fibrovascular septa between cells, and an acidophilic cytoplasm

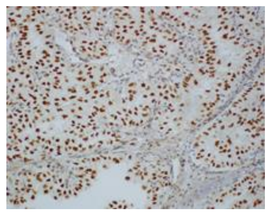

Figure 2
Immunohistochemistry: Shows that the tumor has nuclear reactivity to TFE3 protein antibodies

CASE PRESENTATION

- 2006
  - 6-year-old male presented with a chief complaint of 1 week of painless hematuria
  - Abdominal Ultrasound
    - Right intrarenal mass - 5.1cm sagittal x 4.6cm transverse x 3.4cm AP
    - No regional lymphadenopathy was noticed.
  - Right radical nephroureterectomy, and regional lymph node excision (periaortic and caval)
  - Diagnosis of type II papillary RCC (stage II)
- 2007
  - CT scan showed enlarging right hilar lymphadenopathy
  - Patient underwent a right pneumonectomy
  - Post surgery, patient developed iatrogenic retrodextrocardia, and showed evidence of left lung hypertrophy into the right chest cavity.
- 2008
  - MRI showed evidence of a right retroperitoneal mass, extending into the neural foramen of L3 and L4
  - Patient underwent stereotactic radiosurgery in February, and additional treatment with Sunitinib was prescribed in September

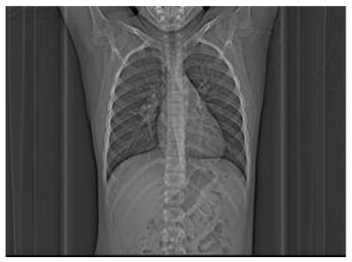

Figure 3
CT Scan: shows right hilar lymphadenopathy

CASE PRESENTATION (cont)

- 2013
  - CT scan showed evidence of metastasis to periaortic lymph node, which was then resected.
- 2018
  - Patient has not shown any further evidence of metastasis.

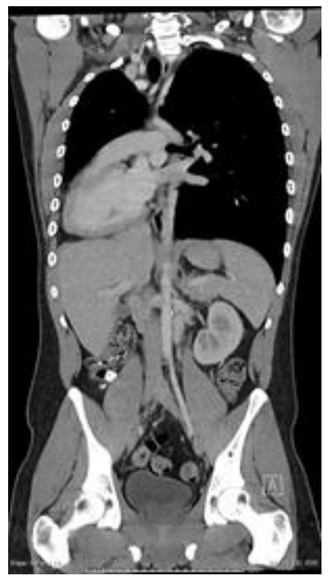

Figure 4
CT Scan: right retrodextrocardia, and hypertrophy of the left lung into the right thoracic cavity

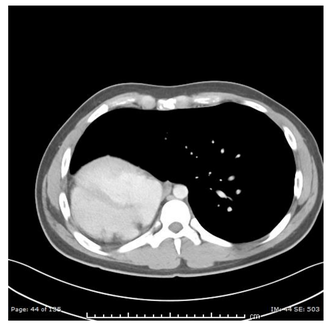

Figure 5
CT Scan: right retrodextrocardia

CONCLUSION

- Positive nodal status at the time of diagnosis, and recurring metastasis are indicative of a poor prognosis in patients.
- The lack of a treatment protocol provided challenges in the treatment.
- Despite these factors, an individual based approach to the treatment of this patient's disease proved to be successful, with the patient being in remission for more than 5 years.
- Importance of collaboration between healthcare providers to allow for a good quality of life for the patient

REFERENCES

- Rialon KL, Gulack BC, Englum BR, Routh JC, Rice HE. Factors impacting survival in children with renal cell carcinoma. J Pediatric Surgery. 2015;50:1014-8.
- Abdellah A, Selma K, et al. Renal cell carcinoma in children: case report and literature review. Pan Afr Med J. 2015;20:84.
- Young EE, Brown CT, Merguerian PA, Akhavan A. Pediatric and adolescent renal cell carcinoma. Urol Oncol. 2016;34:42-9.
- Chowdhury T, Prichard-Jones K, Sebire NJ, Bier N, Cherian A, Sullivan MO, O'Meara A, Anderson J. Persistent complete response after single-agent sunitinib treatment in a case of TFE translocation positive relapsed metastatic pediatric renal cell carcinoma. J Pediatric Hematology Oncology. 2013;35(1):e1-3.
- Altinok G, Kattar MM, Mohamed A, Poulik J, Grignon D, Rabah R. Pediatric renal carcinoma associated with Xp11.2 translocations/TFE3 gene fusions and clinicopathologic associations. Pediatric Developmental Pathology. 2005;8(2):168-80.
- Perlman EJ. Pediatric renal cell carcinoma. Surg Pathol Clin. 2010;3(3):641-651.

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