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Indian Journal of Pathology and Oncology

Journal homepage: www.innovativepublication.com

Case Report

Wilms' tumor: A rare presentation

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ARTICLE INFO

Article history:
Received 26-09-2019
Accepted 15-11-2019
Available online 25-05-2020

Keywords:
Wilms' tumor
Metastasis
Lung

ABSTRACT

Wilms' tumor is the most common malignant renal tumor of childhood with a majority of them diagnosed before 5 years of age. The incidence of metastases of Wilms' tumor is approximately 10%. The primary distant site for metastasis is the lung; bone metastasis being extremely rare.Relapses after 5 years of diagnosis is uncommon in Wilms' tumor and there are very few cases of relapse after 20 years of initial diagnosis. We present here a rare case of a patient who was diagnosed with Wilms' tumor 17 years ago and now presented with a lung mass.

1. Introduction

Wilms' tumor is the most commonly diagnosed malignant kidney tumor in children with a majority of them diagnosed before 5 years of age. In Asia, the peak incidence of the disease is in the second year of life with a male predilection.[1] Usually the patients present with an asymptomatic abdominal mass. Like other malignancies, Wilms' tumors also have tendency to metastasize. The incidence of metastases of Wilms' tumor is approximately 10%. The primary distant site for metastasis is the lung; bone metastasis being extremely rare.[2] Relapses in Wilms' tumor after 5 years of diagnosis is uncommon. There are very few cases of relapse after 20 years of initial diagnosis.[3]

We present here a rare case of a patient who was diagnosed with Wilms' tumor 17 years ago at the age of three years and now had a lung mass.

2. Case Report

A 20 year old man presented with vague respiratory symptoms in 2017. The CT scan showed consolidation collapse involving predominantly lingular segment of left lung. Imaging did not reveal any mass in the abdomen. Subsequently, he underwent a trucut biopsy from the lung. The H and E section showed a tumor composed of round or ovoid cells with hyperchromatic nuclei, inconspicuous nucleoli and scanty cytoplasm. The cells were arranged in cords, trabeculae, acini, and papillae. At places, they appeared to form glomeruloid structures (Figure 1). Mitotic figures were scant. No necrosis was found.

We performed the following immunohistochemistry markers on Ventana Benchmark XT platform (TTF1, CK7, CD56, Napsin A, Synaptophysin, chromogranin, CK, WT1, CD99, S100, Desmin and calretinin) to rule out primary lung tumors. TTF1 (DAKO: 8G7G3/1), CK7 (DAKO; OV-TL12/30), Napsin A (LEICA:IP64), synaptophysin (DAKO-DK-SYNAP), and chromogranin (VENTANA; LK2H10), were all negative. Further immunohistochemistry panel showed the cells were positive for CK (DAKO:AE1/AE3), WT1(DAKO; 6F-H2), and negative for CD99 (VENTANA;013), S100 (DAKO; Pa RABBIT), Desmin (LEICA; DE-R-11) and calretinin (DAKO;-CALRET-1) (Figure 2).

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https://doi.org/10.18231/j.ijpo.2020.066

On further interrogation, the patient revealed past history of Wilms' tumor of left kidney. He had undergone nephrectomy 17 years ago followed by adjuvant therapy. In conjunction with the immunohistochemical findings, past history and the histologic findings, the tumor was diagnosed as a late metastasis of Wilms' tumor. We could not review the original slides of the Wilms' tumor, resected 17 years ago, as they were not available.

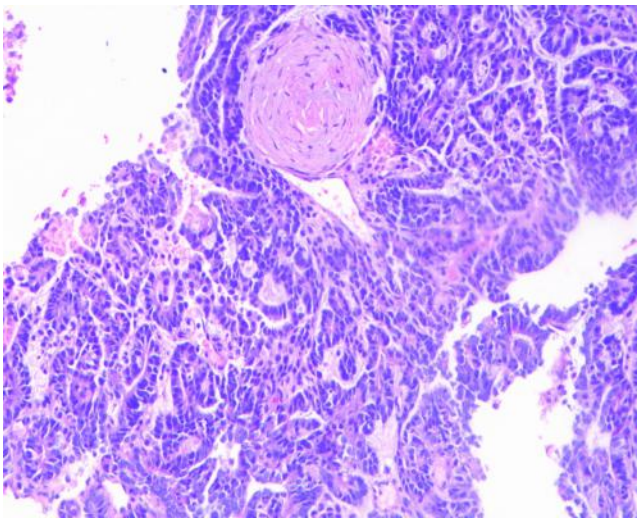

Fig. 1: Photomicrograph showing the tumor cells arranged in tubules and anastomosing cords(H & EX10)

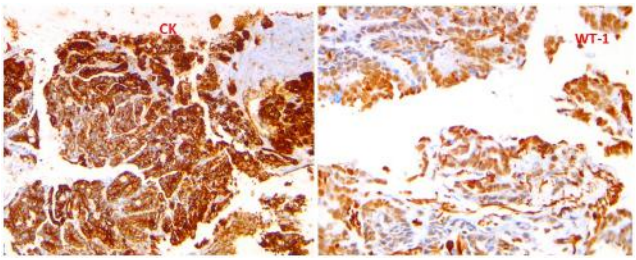

Fig. 2: (a & b): Photomicrographs showing the tumor cells are positive for CK, and WT1

3. Discussion

Wilms' tumor is an embryonic malignancy that is thought to arise from the persisting nephrogenic rests. Wilms tumor is mostly diagnosed in patients between 1–4 years in about 70–80% cases with peak of incidence being 2–5 years of age.[4] Wilms' tumour mimics the development of the normal kidney and consists of three components: blastema, epithelium and stroma. The blastemal component comprises sheets and anastomosing cords of undifferentiated cells having round to oval hyperchromatic nuclei and small nucleoli. When the tumor contains all the three components namely blastema, epithelium and stromal cells it is called triphasic. Biphasic as well as monophasic forms also exist. Metastasis in Wilms' tumor is not uncommon and occurs in about 15% cases with favourable histology.[5]

Most recurrent Wilms' tumors are diagnosed within 2 years of diagnosis. The primary distant site for metastasis is the lung. From a study of 208 children with Wilms' tumor, 31 children (14.9%) had metastases at diagnosis and the lung was affected in 29 children (93.5%).[6] Long-delayed late recurrences is defined as reappearance of the disease 5 years after the initial diagnosis. It is an extremely rare event.

Our case diagnosed in 2000, at the age of 3 years underwent nephrectomy and adjuvant therapy. There was a disease-free interval of 17 years. While in most of the cases the site of recurrence is in the retroperitoneum, our case presented with lung mass. The histology showed epithelial predominant Wilms' tumor.

The pathogenesis of late relapse in Wilms' tumor and in other neoplasms is not well understood, but several hypotheses have been suggested. One of the possibilities is that long-delayed recurrence may be due to eradication of the high-grade immature component and the induction of a high degree of differentiation, thus enhancing the disease-free interval time.[7]

Other mechanisms that have been suggested, are escape from immunevigilance, scarce vascularization in some portion of the tumor, and cell dependency on growth factors.[8]

4. Conclusion

This case therefore highlights the need for keeping the diagnosis of Wilms' tumors in the differentials for the histopathologist and the benefit of continued follow up for patients with Wilms' tumor. Therefore in young adults even if the morphology is of a round cell tumor, the possibility of a blastemal phase of Wilms' tumor should be considered.

5. Acknowledgement

The authors will like to acknowledge the technical team who helped in slide preparation, staining and immunohistochemistry.

6. Statement of Ethics

Since this case study did not have any serious implication on patient outcome, approval of ethical committee was not considered deemed necessary.

7. Conflict of Interest

None.

8. Sources of Funding

None.

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Cite this article: Dey D, Mitra B, Sengupta S. Wilms' tumor: A rare presentation. *Indian J Pathol Oncol* 2020;7(2):335-337.