Wilms' Tumor Arising in a Horseshoe Kidney

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Wilms' tumor has been associated with multiple congenital anomalies and malformations. The most common anomalies are aniridia, hemihypertrophy and genitourinary anomalies(l,2). The association of Wilms' tumor with horseshoe kidney is, however, uncommon. The National Wilms' Tumor Study (NWTS) has reported 13 instances of this anomaly in a total of 2901 cases (0.4%)(3). The present communication describes a case of Wilms' tumor arising from the isthmus of a horseshoe kidney and aims to highlight the problems faced in the diagnosis and management.

Case Report

A 3.5 years old boy was brought with a progressively enlarging abdominal lump and abdominal pain for the preceding two months. Examination revealed a large (20x15 cm) abdominal mass lying transversely in the central and lower parts of abdomen, extending more to the right side. There was no liver enlargement and the blood pressure was normal. Urine examination did not show presence of red blood cells.

Ultrasonography showed the mass to be arising from the right kidney, while on computed tomography *(Figs. 1&2)* and intravenous pyelography it was visualized as an abdominal mass of uncertain origin causing bilateral hydronephrosis. Fine needle aspiration from the mass revealed the presence of blastema and epithelial cells suggesting a diagnosis of Wilms' tumor.

The patient was thus deemed to have Wilms' tumor of right kidney crossing -the mid line and causing bilateral hydronephrosis. After eight weeks of chemotherapy using vincristine, actinomyciu-D and doxorubicin, surgery was performed.

A large tumor was seen adhered to the isthmus of a horseshoe kidney (HSK) and pushing it upwards. The tumor was connected with the pelvic-calyceal system of the isthmus and was receiving blood supply from the internal iliac artery. The left ureter was coursing through the tumor. The right kidney had duplex ureter draining it. The tumor was dissected free from the ureters, isthmus and promontory, to which it was adherent. A small amount of residual tumor was left behind at the time of surgery.

The diagnosis of Wilms' tumor (WT) was confirmed on microscopic examination of the excised tumor, which showed it to be composed of diffuse

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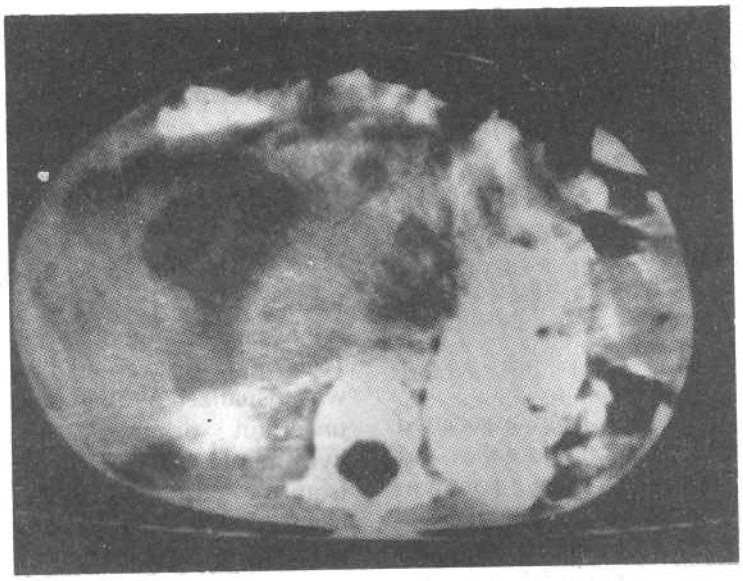

Fig. 1. CT abdomen showing the tumor apparently arising from the right kidney.

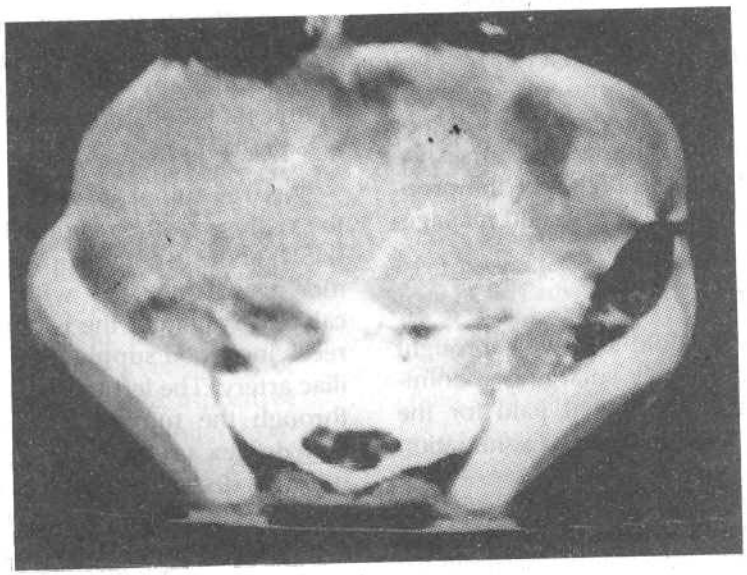

Fig. 2. CT abdomen showing the tumor occupying most of the lower half of the abdomen.

sheets of spindle shaped mesenchymal cells as well as epithelial component in the form of glands, constituting a favorable histology. Lymph nodes were not involved. Post-operatively the tumor was assigned Stage III (incomplete resection) and radiotherapy in a dose of 2000 cGy in 10 fractions was delivered

to the tumor bed. Further chemotherapy using vincristine, actinomycin-D and doxorubicin was administered. The patient completed 10 months of post-operative therapy and was disease free, off treatment, till 17 months after initial diagnosis. A recurrence of a right sided abdominal mass was clinically detected during a routine follow-up visit. A FNAC from the mass confirmed that the Wilms' tumour had recurred. The family has opted out of further therapy because of financial restraints.

Discussion

The high incidence of congenital anomalies in children with WT has long been appreciated(l,2). The incidence of upper genitourinary anomalies has been reported to be 4-6%(l,2,4). The occurrence of WT in a HSK is, however, an uncommon event. The estimate varies from 0.4%(3) to 0.9%(5) of all Wilms' tumors. Although the overall incidence of malignancies arising in HSK is not increased^), the risk of developing WT' increases by 2-8 times when compared to the general population(3,6,7). The reason for the increased incidence of WT in these cases may be related to the embryogenesis of HSK. Children with Turner's syndrome, who have a 7% incidence of HSK, however, appear to be at no increased risk for developing WT(8). This discrepancy may be due to a different embryological origin of HSK in Turner's syndrome—thereby assuming that in the other cases the formation of HSK predisposes to a second event which results in WT(8,9). Other renal tumors which have been described in children with HSK include rhabdomyosarcoma(6) and teratoma(10,ll).

It is important to recognize when managing these children that HSK predisposes to the occurrence of several other congenital anomalies—especially involving the skeletal, cardiovascular and nervous systems(12,13). In addition there is great variation noted in the basic shape of HSK(14) with variable relationship between the great vessels, calyces and ureter. The blood supply to the isthmus may come from renal artery, directly from aorta, inferior mesenteric artery, or (as in the index case) from the iliac arteries.

Children with WT arising in HSK do not differ significantly from others where the tumor arises in a normal kidney with respect to age, sex, clinical stage at presentation and histologic pattern(3). Inspite of this, these children present greater problems in diagnosis and it is common for the diagnosis to be first established at the time of surgery. In the NVVT6 report(3) nearly half of the 13 cases remained undiagnosed prior to surgery. When the tumor arises from one of the sides of HSK, imaging procedures can delineate the isthmus and thus help clarify the diagnosis(15). More commonly, the tumor arises from or involves the isthmus(6), and obscures its visualization thus leading to difficulties in preoperative diagnosis(14,15).

Fine needle aspiration cytology (FNAC) is a useful diagnostic aid in such clinical situations(16). The accuracy of this technique in diagnosing WT has been demonstrated recently(17,18). The presence of blastemal cells in various combinations with epithelial cells and stroma allow a diagnosis of WT to be made without the aid of histology(15).

The presence of HSK does not alter

the prognosis of malignant tumors arising from such kidneys(10) and the same applies to WT arising in HSK as well(3). The prognosis depends on the histology and the stage of the tumor. Problems in management however, may arise during and after surgery. Because of frequent anatomic variations in such cases(14), preoperative arteriography can facilitate surgery(6). Excision of most of the tumor was obtained in the index case because it was growing out from the isthmus and not involving either of the sides.

Post-operative radiotherapy to the tumor bed in case of incomplete excision of the tumor (Stage III) has to be planned individually for each case depending upon the location of the tumor. Where isthmus is the site of origin the radiation field would involve the lumbar spine as well, with anticipated growth problems later on. A dose of 2000 cGy would still be advisable in our country because of relatively inferior long-term disease free survivals.

The overall survival of 85% in WT arising in HSK(3) indicates that management according to existing guidelines is satisfactory. The use. of preoperative chemotherapy, may offer the advantage of more conservative surgery. In such atypical presentations of WT, FNAC is a valuable adjunct to radiological investigations to make a more confident preoperative diagnosis. The propositus relapsed about 5 months after completing therapy (chemotherapy, surgery and radiotherapy). The unfavorable outcome is probably a reflection of the huge size of the tumor at initial presentation and the fact that there was tumor spillage and incomplete surgical resection.

REFERENCES

1. Miller RW, Fraumeni JF, Manning MD. Excessive concurrence of Wilms' tumor with aniridia, hemihypertrophy and other congenital defects. J Pediatr 1964, 65:1088-1089.
2. Pendergrass TW. Congenital anomalies in children with Wilms' tumor. Cancer 1976, 37: 403-408.
3. Mesrobian HJ, Kelalis PP, Hrabovsky E, Othersen HB Jr, DeLorimier A, Nesmith B. Wilms' tumor in horseshoe kidney: A report from the National Wilms' Tumor Study. J Urol 1985,133:1002-1003.
4. Jagasia KH, Thurman WG. Congenital anomalies of the kidney in association with Wilms' tumor. Pediatrics 1965, 35: 338-340.
5. Miller RW, Fraumeni JF Jr, Manning MD. Association of Wilms' tumor with aniridia, hemihypertrophy and other congenital malformations. N Eng J Med 1964, 270: 992-994.
6. Buntley D. Malignancy associated with horseshoe kidney. Urology 1976, 8: 146-148.
8. Lippe B, Geffner ME, Dietrich RB, Boeschat MI, Kangarloo H. Renal malformations in patients with Turner syndrome: Imaging in 141 patients. Pediatrics 1988, 82: 852-875.
9. Knudson AG, Jr, Strong LC. Mutation and cancer: A model for Wilms' tumor of the kidney. J Natl Cancer Inst 1972, 48:313-316.
10. Murphy DM, Zincke H. Transitional cell carcinoma in horseshoe kidney. Report of 3 cases and review of literature. Br J Urol 1982, 54: 484-485.
11. Dische MR, Johnston R. Teratoma in horseshoe kidney. Urology 1979, 13: 435-436.

12. Zondek LH, Zondek T. Horseshoe kid- and associated congenital anoma-Urol Int 1964,18: 347-349.

13. Whitaker RH, Hunt GM. Incidence and distribution of renal anomalies in patients with neural tube defects. Eur Urol 1987,13: 322-325.

14. Bauer SB, Perlmutter AD, Retik AB. Anomalies of upper urinary tract. *In:* Campbell's Urology, 6th edn. Eds. Walsh PC, Retik AB, Stamey TA, Vaughan ED. Philadelphia, W.B. Saunders Company, 1992, pp 1376-1381.

15. Gay BB Jr, Dawes RK, Atkinson GO Jr, Ball TI Jr. Wilms' tumor in horseshoe kidneys: Radiologic diagnosis. Radiology 1983,146: 693-679.

16. Pilotti S, Rilke F, Alasio L, Garbagnati F. The role of fine needle aspiration in the assessment of renal masses. Acta Cytol 1988, 32:1-10.

17. Quijano G, Drut R. Cytologic characteristics of Wilms' tumor in fine needle aspirates. A study of ten cases. Acta Cytol 1989, 33: 263-266.

18. Dey P, Radhika S, Rajwanshi A, *et al.* Aspiration cytology of Wilms' tumor. Acta Cytol 1993, 37: 477-482.

Fungal Peritonitis Complicating Peritoneal Dialysis

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Peritonitis is the most frequent serious complication in patients requiring multiple peritoneal dialysis (PD). The causative organisms are usually bacteria(l). Fungi are uncommonly implicated, .being responsible for only 2-10% of all peritonitis episodes associated with PD(2). The majority of the published experience deals with fungal peritonitis in patients undergoing continuous ambulatory peritoneal dialysis, the incidence being 0.2 to 1.7 episodes per 12 patient months of dialysis(3). Fungal peritonitis, if not detected early and treated appropriately, is associated with increased morbidity (development of peritoneal adhesions and inability to resume PD) and mortality. We describe the case of a 21 month old girl with hemolytic uremic syndrome who needed multiple peritoneal dialysis and developed fungal peritonitis, which was not suspected antemortem.

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Case Report

A 21-months-old girl with dysentery