The Turkish Journal of Pediatrics 29:45 - 49, 1987

BOTRYOID NEPHROBLASTOMA (WILMS' TUMOR) IN A HYPOPLASTIC KIDNEY*

*Nurdan Tunalı MD***, İlhan Tuncer MD***, Ünal Zorludemir MD****, Suzan Zorludemir MD*****, Candaş Tunalı MD******

Key words: Botryoid nephroblastoma, Wilms' tumor, hypoplastic kidney

Cases of nephroblastoma arising from the renal pelvis and/or calices have occasionally been reported[1-6]. In this case report we present a case of nephroblastoma (Wilms' tumor) which simulates sarcoma botryoides (embryonal rhabdomyosarcoma) both grossly and microscopically. The kidney in which the tumor was located appeared to be hypoplastic.

Case Report

A four-year-old boy was brought to the Çukurova University Hospital with the complaint of an abdominal mass which had been detected by the family four days prior to admission. On admission, physical examination revealed a circular solid mass about 15 x 15 cm in diameter in the left lumbar area. Laboratory examinations including a complete blood count, and kidney and liver function tests were all within normal limits. Urinalysis revealed (++) proteinuria and 30-40 leukocytes per high power field.

Intravenous pyelogram revealed a non-functioning left kidney and computerized tomography of the abdomen detected a large solid mass with focal cystic areas (Fig. 1). The mass measured 15 x 15 cm in diameter and was located in the left lumbar region. The left kidney was not visible. The patient was referred to surgery with a preoperative diagnosis of Wilms' Tumor. During an exploratory laparotomy, an intrarenal mass was discovered on the left kidney. A left nephroureterectomy with regional lymph node dissection was performed.

* From the Çukurova University Faculty of Medicine, Adana.
** Assistant Professor of Pathology, Çukurova University Faculty of Medicine
*** Associate Professor of Pathology, Çukurova University Faculty of Medicine
**** Assistant Professor of Pediatric Surgery, Çukurova University Faculty of Medicine
***** Research Assistant in Pathology, Çukurova University Faculty of Medicine.
****** Assistant Professor of Radiology, Çukurova University Faculty of Medicine

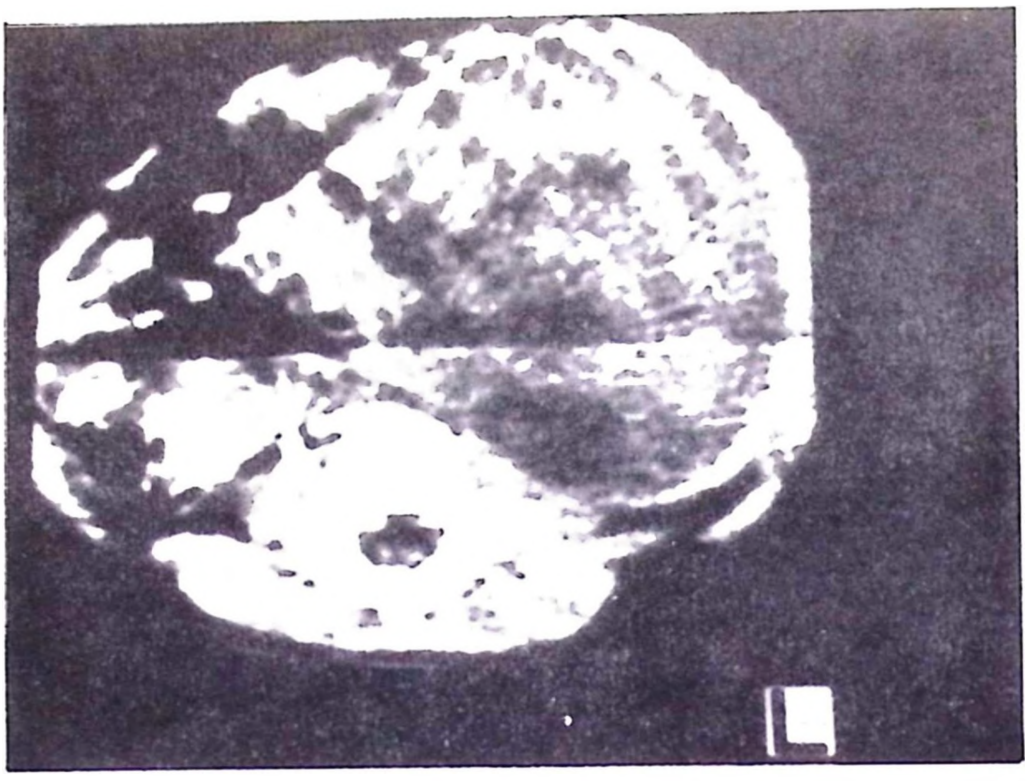

Fig. 1: CT scans of abdomen showed a large solid mass with focal cystic areas in the left kidney.

The specimen weighed 1050 g. and measured 17 x 15 x 8 cm. On sectioning, the pelviceal system was found to be dilated and was filled with a multifocal, soft, glistening botryoid-type tumor (Fig. 2).

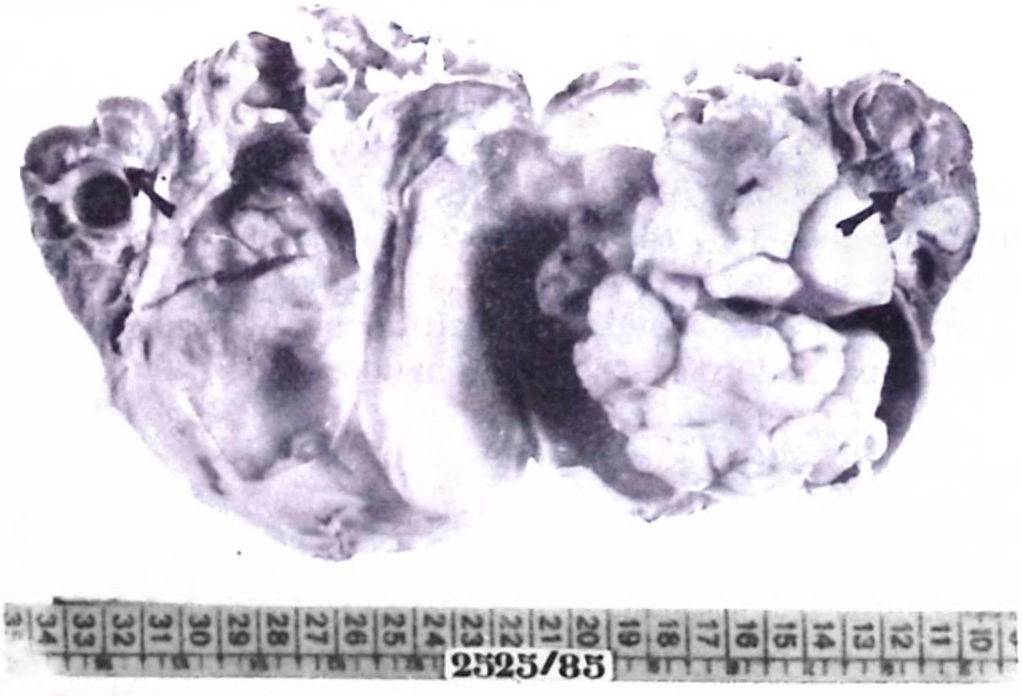

Fig. 2: Renal pelvis and dilated calices are filled with glistening neoplastic polyps and hypoplastic kidney contains four columns of Bertin (arrowhead).

The largest polypoid tumoral mass was 10 x 8 cm in diameter and it was attached to the pelvis by a stalk 3 cm in diameter. Others varied between 1-3 cm in diameter and they were located in the calices. The kidney appeared to be hypoplastic and had only four columns of Bertin. The renal parenchyma was only 0.5 cm in thickness and there was no grossly recognizable tumoral mass in the parenchyma nor could we see any remnant of metanephric blastemic foci. The ureter was obstructed by the tumoral mass but there was no invasion of the uretral wall. The sections from the tumor revealed histologic features of embryonal rhabdomyosarcoma. The embryonic tumor cells were characteristically grouped under the transitional epithelium that covered the polyps. But further sections from the polyps, especially the deeper sections, disclosed typical features of nephroblastoma with metanephric blastematous, epithelial and stromal components. In the deeper areas of the polyps, abortive tubular and/or glomerular structures were discovered surrounding the metanephric blastematous sheets within the stroma (Fig. 3). Scattered foci of rhabdomyoblasts with large eosinophilic cytoplasm and small areas of osteoid tissue were also found.

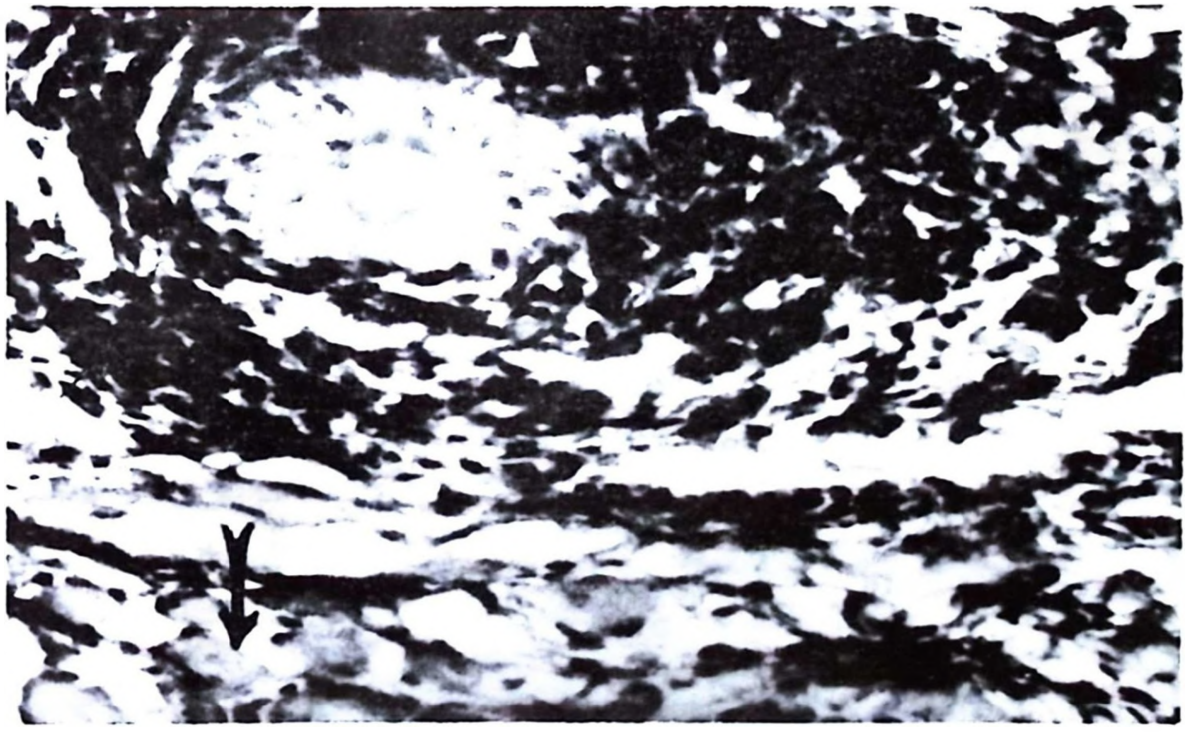

Fig. 3: Tubular structure surrounding blastematous sheet and rhabdomyoblasts (see arrow) (HE x 400)

The lymph nodes were negative for tumor and there was no invasion by the tumor of the renal capsule and renal vein.

Discussion

Several cases of nephroblastoma protruding into the renal pelvis, like botryoid sarcoma, have been reported in the literature. In these rare presentations a tumoral mass in the renal parenchyma was either limited or absent[3-8]. Primary embryonal rhabdomyosarcoma has also occasionally been reported originating from the kidney or its pelvis. Macroscopic differential diagnosis between embryonal rhabdomyosarcoma and nephroblastoma with botryoid development is difficult, if not impossible.

When a nephroblastoma contains a predominantly rhabdomyoblastic component the same problem in differential diagnosis may exist in microscopic examination too. In these cases, examination of multiple tissue samples with deep and/or serial sections are necessary to detect metanephric blastematous epithelial tissue which is essential in the diagnosis of nephroblastoma[1-2].

Most nephroblastomas are located in the kidney parenchyma. Some of them may originate from renal subcapsular areas and sometimes there is multifocal distribution[1-2]. Persistent metanephric blastematous nodules have occasionally been detected in the renal parenchyma as in the subcapsular region, the mid portion of the columns of Bertin and the peripelvic areas. It has been suggested that these persistent blastematous areas are related to the histogenesis of nephroblastoma[1]. Occurrence of polypoid nephroblastoma located in the renal pelvis may support this theoretical relationship.

Marsden and Lawler[9] use the term "renal dysplasia" for nodular metanephric blastematous residue. They report an 18% incidence of renal dysplasia in nephroblastoma in their own series and refer to a 33.3% incidence in the Bove and McAdams series. They suggest that either the tumor may originate directly from dysplastic foci or that the two are independent but are linked etiologically.

In the case presented the involved kidney appeared to be hypoplastic. It is known that there is a significant correlation between nephroblastoma and genitourinary developmental anomaly[10].

Summary

A case of nephroblastoma (Wilms' tumor) which grossly and microscopically simulated sarcoma botryoides embryonal rhabdomyosarcoma is presented. Macroscopic examination of the nephrectomy material revealed a grape-like tumor occupying the left renal pelvis and colyceal system and which had the macroscopic appearance of botryoid sarcoma. Although preliminary microscopic examination simulated botryoid sarcoma, further examination of the multiple sections taken from different sites of the tumoral mass revealed metanephritic blastemic epithelial and mesenchymal components confirming the diagnosis of

Wilms' tumor. No tumoral mass was demonstrable in the renal parenchyma. The involved kidney appeared to be hypoplastic, four columns of Bertin being recognized.

REFERENCES

1. Benington JL, Beckwith JB. Tumors of the kidney, renal pelvis and ureter. *Atlas of Tumor Pathology*. Washington D.C: Armed Forces Institute of Pathology, 1975, 2nd series, fascicle 12, pp 31-91.
2. Rosai J. *Ackerman's Surgical Pathology* (6th ed) Vol 1. St Louis: CV Mosby Co, 1981, pp 795-800.
3. Chiba T, Ohashi E. Wilms' tumor extending into the dilated renal pelvis as a mold. *J Urol* 124:130, 1980.
4. Mahoney JP, Saffos RO. Fetal rhabdomyomatous nephroblastoma with a renal pelvic mass simulating sarcoma botryoides. *Am J Surg Pathol* 5:297, 1981.
5. Weinberg AG, Currarino G, Hurt GE. Botryoid Wilms' tumor of the renal pelvis. *Arch Pathol Lab Med* 108:147, 1984.
6. Wicklund RA, Tank ES. Polypoid renal pelvic lesions in children. *J Urol* 123:943, 1980.
7. Harbaugh JT. Botryoid sarcoma of the renal pelvis: a case report. *J Urol* 100:424, 1968.
8. Seaburry JC Jr. Renal rhabdomyosarcoma. *JAMA* 201:1043, 1967.
9. Marsden HB, Lawler W. Wilms' tumour and renal dysplasia: an hypothesis. *J Clin Pathol* 35:1069, 1982.
10. Pendergrass TW. Congenital anomalies in children with Wilms' tumor: a new survey. *Cancer* 37:403, 1976.