Case Report

Coexistence of Cystic Nephroma and Neuroblastoma

A Rare Case of a Childhood Collision Tumor

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INTRODUCTION

A collision tumor is present when two distinct primary tumors are found together. Both collision tumors and cystic nephroma (CN) are very rare. Here, we report a case of collision tumor composed of an ipsilateral CN and a neuroblastoma.

CASE REPORT

A 14-month-old girl was admitted to Dr. Sami Ulus Children's Hospital in September 2002 because of a lump on the right side of the abdomen with no other symptoms. On her physical examination, a mass at the right upper lateral abdomen nearly 7 × 6 cm in diameter was palpated. Her past and family histories were unremarkable.

The abdominal ultrasonography and tomography scan revealed a 93 × 75 × 81 mm mass in the right renal localization, which had a smooth surface and multiple septae with solid and cystic areas. The 24-hour urine sample examination revealed increased amounts of vanilylmandelic acid and homovanilic acid (2.8 mg/day and 4.7 mg/day, respectively). Other laboratory findings and echocardiography were within normal limits.

Right radical nephrectomy was performed and a mass at the inferior region of the right adrenal gland was removed. The renal tumor was well circumscribed with a thick, fibrous capsule in macroscopic examination and the adjacent renal pa-

renchyma was compressed by a mass of 100 × 80 mm. The cut surface of the tumor was multicystic with non-communicating cysts that varied in size from a few millimeters to 4 cm. The other was soft, gray-white and gray-purple in color and 20 × 25 mm in size.

On microscopic examination, the cysts were lined by flattened to cuboidal or hobnail epithelium. Septae were composed of fibrous tissue and contained differentiated tubules (Figure 1). The surrounding kidney tissue showed minimal mesangial proliferation and tubule epithelium with no evidence of any other dysplastic lesions. No blastemal rest or other precursor lesions of Wilms tumor were encountered. Microscopic examination of the other tumor in the inferior region of the adrenal gland revealed all stages of neuronal differentiation throughout the tumor. The tumor had a lobular appearance owing to the presence of thin fibrovascular septae between the groups of tumor cells, and was composed of sheets of small cells with hyperchromatic nuclei and scanty cytoplasm. Between the tumor cells were immature, multinucleated, or completely abnormal ganglion cells that were placed either solitary or in groups. A neurofibrillary stroma was seen between the masses of cells.

Immunohistochemically, tumor cells were shown to express synaptophysin and chromogranin (Figure 2). N-myc amplification test was not performed because of technical problem. This pathologic diagnosis was well differentiated neuroblastoma and CN. The patient is alive and has been disease-free for nine years.

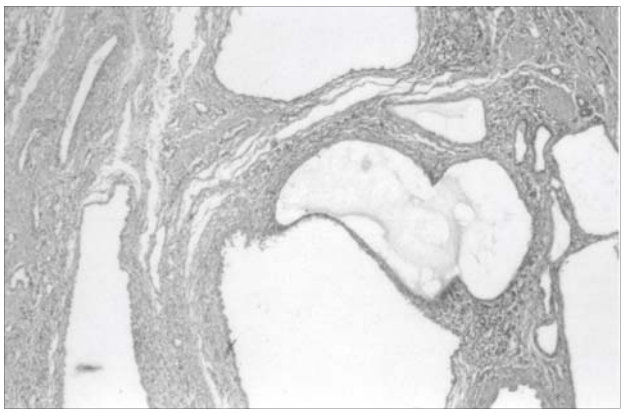

Figure 1. Cystic nephroma which consists of varied size cysts and septae (hematoxylin and eosin, × 80)

DISCUSSION

Neuroblastic tumors are mainly originated from primordial neural crest cells that generate adrenal medulla and sympathetic ganglia.

Cystic renal neoplasms in childhood represent a spectrum of clinicopathologic entities with CN and cystic, partially-differentiated nephroblastoma on the benign end and polycystic Wilms tumor on the malignant end.[1,2] There are two peaks in the incidence of the tumor, one in the childhood and one in the middle age. Most childhood cases occur between the ages of 3 months and 2 years and 73% of the subjects are male. The second peak consists of women in the age range of 40 to 60 years.[3]

In a clinicopathologic study on renal neoplasms of childhood, only 3 cases with CN out of 111 renal neoplasms were reported.[4] The etiology of CN is unknown.[5] The most frequent symptom in CN is painless abdominal mass[6] as in our case. Cystic nephroma may also cause infection, hypertension, and pain.[7]

Pre-operative diagnosis with radiological methods may be imprecise or even inadequate.[8] Some authors suggest that ultrasonography is better than computed tomography in terms of showing internal structure of CN.[9]

Collision tumor is a rare entity where two distinct primary tumors exist together. Collision tumor in childhood has previously been reported as coexistent mesoblastic nephroma and neuroblastoma,[10,11] pulmonary blastoma and CN,[12] CN and Wilms tumor,[13] and neuroblastoma and nephroblastoma.[14] Nisen and colleagues have reported a 2.5-year-old boy with CN on the right kidney and intrarenal neuroblastoma on the left kidney.[15] But in our patient, both tumors were on the same side. Vegunta and associates have discussed the etiology of collision

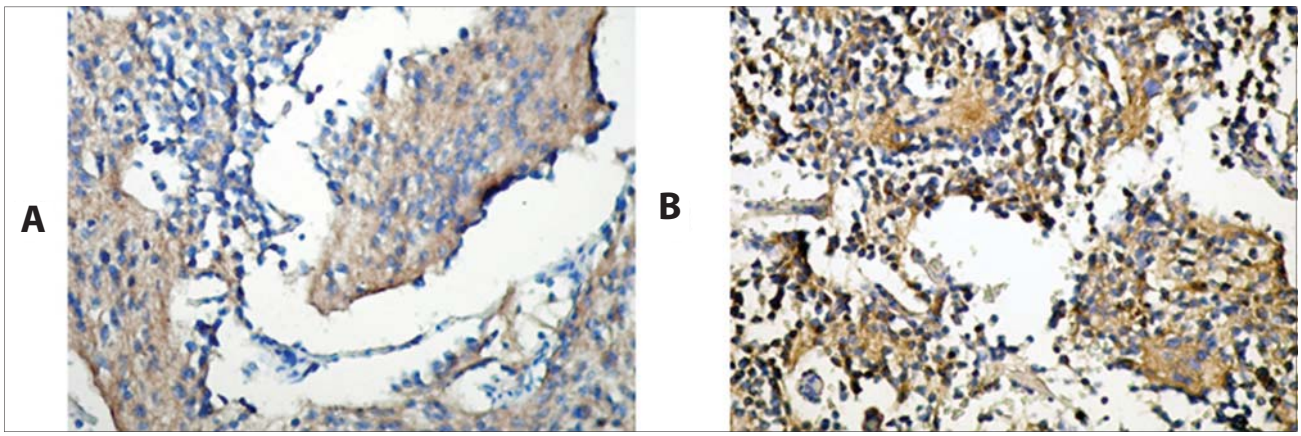

Figure 2. (A) Diffuse synaptophysin staining in neoplasm, antisynaptophysin (× 200).
(B) Chromogranin positive areas of the neoplasm, anti-chromogranin (× 200)

tumors and proposed that a common tumorigenic stimulus triggers the neoplastic transformation of different cell types and their tumorigenic development.[10] We propose that therapy for collision tumor must be designed for both tumors if they have a malignant component.

CONFLICT OF INTERESTS

None declared.

REFERENCES

1. Joshi VV, Beckwith JB. Multilocular cyst of the kidney (cystic nephroma) and cystic, partially differentiated nephroblastoma. Terminology and criteria for diagnosis. Cancer. 1989;64:466-79.
2. Sacher P, Willi UV, Niggli F, Stallmach T. Cystic nephroma: a rare benign renal tumor. Pediatr Surg Int. 1998;13:197-9.
3. Madewell JE, Goldman SM, Davis CJ, Jr., Hartman DS, Feigin DS, Lichtenstein JE. Multilocular cystic nephroma: a radiographic-pathologic correlation of 58 patients. Radiology. 1983;146:309-21.
4. Jenkner A, Camassei FD, Boldrini R, et al. 111 renal neoplasms of childhood: a clinicopathologic study. J Pediatr Surg. 2001;36:1522-7.
5. Eble JN, Bonsib SM. Extensively cystic renal neoplasms: cystic nephroma, cystic partially differentiated nephroblastoma, multilocular cystic renal cell carcinoma, and cystic hamartoma of renal pelvis. Semin Diagn Pathol. 1998;15:2-20.
6. Boulanger SC, Brisseau GF. Cystic nephroma: a benign renal tumor of children and adults. Surgery. 2003;133:596-7.
7. Castillo OA, Boyle ET, Jr., Kramer SA. Multilocular cysts of kidney. A study of 29 patients and review of literature. Urology. 1991;37:156-62.
8. Babut JM, Bawab F, Jouan H, Coeurdacier P, Treguier C, Fremond B. Renal cystic tumours in children--a diagnostic challenge. Eur J Pediatr Surg. 1993;3:157-60.
9. Hopkins JK, Giles HW, Jr., Wyatt-Ashmead J, Bigler SA. Best cases from the AFIP: cystic nephroma. Radiographics. 2004;24:589-93.
10. Vegunta RK, Morotti RA, Shiels WE, 2nd, Rauck A, Besner GE. Collision tumors in children: a review of the literature and presentation of a rare case of mesoblastic nephroma and neuroblastoma in an infant. J Pediatr Surg. 2000;35:1359-61.
11. Hero B, Kremens B, Sudermann T, Haas RJ. Collision tumor in children: a review of the literature and presentation of a rare case of mesoblastic nephroma and neuroblastoma in an infant. J Pediatr Surg. 2001;36:1607-8.
12. Delahunt B, Thomson KJ, Ferguson AF, Neale TJ, Meffan PJ, Nacey JN. Familial cystic nephroma and pleuropulmonary blastoma. Cancer. 1993;71:1338-42.
13. Vujanic GM, Jenney ME, Adams H, Meyrick SM. Juxtaposed cystic nephroma and Wilms' tumor. Pediatr Dev Pathol. 2000;3:91-4.
14. Bissig H, Staehelin F, Tolnay M, et al. Co-occurrence of neuroblastoma and nephroblastoma in an infant with Fanconi's anemia. Hum Pathol. 2002;33:1047-51.
15. Nisen PD, Rich MA, Gloster E, et al. N-myc oncogene expression in histopathologically unrelated bilateral pediatric renal tumors. Cancer. 1988;61:1821-6.