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Intrarenal Teratoma*

—Report of a Case Occurring in an Infant—

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=국문초록=

영아의 신장 기형종

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영유아기에 발생한 신장 기형종은 Wilms씨 종양과의 임상적감별이 대단히 어려울 뿐 아니라 발생빈도가 매우 드물기 때문에 영유아기 신장종양의 대부분을 차지하는 Wilms씨 종양으로 간주하기 쉽다.

본 증례는 3개월된 여아에서 방사선과적소견과 수술시의 임상적 진단이 Wilms씨 종양으로 조직학적 확진 이전에 actinomycin D화학요법이 시작되었던 신장기형종의 1례이다. 발생빈도는 드물지만 영유아기에 신장의 종괴가 발견되면 반드시 동결절편을 시행하여 신장기형종을 비롯한 양성종양의 가능성을 배제하여야 할 것 같다.

After a review of literature, it becomes obvious that germ cell tumors arise in a variety of extragonadal locations during infancy, and that the kidney is the least frequent site of occurrence. There have been only seven well documented cases of intrarenal teratoma in English literature; four in infants[1~4], one in an adolescent[5] and two in adults[6,7]. There are, of course some, additional reports, but the literatures have not been available to us.

Since intrarenal teratoma is extremely rare and simulates Wilms' tumor in the roentogenographic and even the operative findings, postoperative chemotherapy used to be initiated without histological diagnosis. Here we report such a case.

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Case Report

The patient, a 3-month-old female, was admitted because of marked abdominal distention with huge left abdominal mass which was discovered by the mother 4 days prior to visiting this hospital. She was born after normal delivery. There was no heredofamilial disease or significant past history.

On admission, the body weight was 7.4 kg, temperature 38.4 C, pulse rate 140/min and the respiratory rate 35/min. The blood pressure was elevated markedly to 260/200. Physical examination revealed a voluminous intra-abdominal mass extended to fill most of the left abdomen. No other findings were significant. Laboratory findings were not remarkable except for the urinalysis that showed one positive protein, 10~25 WBC and 25

or more RBC per high power field. Other routine tests including electrolytes, LFT and EKG were within normal limits. Flat abdomen film confirmed the left abdominal mass which was presented as a solid one by ultrasonogram. Intravenous pyelogram disclosed the left calyceo-pelvic system distorted and medially displaced, the kidney outline and the ureter being nonvisualized.

An operation was performed with the preoperative diagnosis of nephroblastoma and a large round encapsulated mass was demonstrated at the left kidney area, which displaced the colon upward and stretched the left adrenal gland. A portion of grossly uninvolved renal cortical tissue was found at the lower pole of the mass (Fig. 1). Aspiration of the tumor yielded about 100 cc of yellow turbid but not viscid fluid. The liver was slightly enlarged and pale. The spleen appeared normal. The postoperative clinical diagnosis was also a nephroblastoma and actinomycin D was started immediately after the operation before histological diagnosis was made.

Pathological Description

The excised mass, including the compressed renal tissue located at the inferior aspect of the specimen, was globular and measured 15 cm in diameter and weighed 840 gm. Both the mass and renal tissue were enclosed within a common fibrous capsule which seemed grossly uninvaded by the tumor. The cut surface disclosed a well circumscribed tumor which was sharply demarcated but intimately apposed to the compressed renal cortex. Hemorrhage and necrosis were not noted. There were a few small cysts in the background of solid gray-white tissue.

Microscopically a wide variety of tissue types representing the three germ layers were readily identified. Differentiated neuroglia and nonkeratinizing squamous epithelium lining small cystic spaces were the predominant ectodermal tissues. Numerous rudimentary hair follicles were seen just beneath the squamous epithelium (Fig. 2). Entodermal components such as salivary gland tissue and glands lined by tall columnar epithelial cells reminiscent of mucin secreting cells were demonstrated. Pseudostratified columnar epithelium-lined tubular structures with underlying plates of immature hyaline cartilage were also noted (Fig. 3). Calcified bone, glomeruloid structure with irregular tubules resembling primitive renal tissue, and immature skeletal muscle bundles were found as mesodermal elements (Fig. 4).

There was no histological evidence of malignancy, although there were small patches of hemorrhage and necrosis in several tissue sections.

Discussion

Teratomas arising in the retroperitoneum and sacrococcygeal areas represent the most common type of extragonadal germ cell tumors during the 1st 12 months of life[8]. If gonads and soft tissues of the retroperitoneum and sacrococcygeal region are excluded from consideration, the intracranial region including the pineal gland, liver, stomach and kidney have been identified as the only organs in which teratomas have been described, the kidney being the least frequent site[9].

In the retroperitoneum, the teratomas occur most often at the region of the upper pole of the kidney and particularly on the left side[10,11], and not infrequently, they are firmly attached to the kidney[12~14]. In these cases,

Table 1. Review of the English literature on intrarenal teratoma

Authors		Age & Sex		Location	Gross	Glomeruloid structure	Remark
Baldwin JF,	1915	16 y,	F	Right lower half	Cystic	—	Dermoid and bony plate
McCurdy GA,	1934	7 w,	M	Left mid & lower	Solid & Cystic	+	Ass with meningocele, hypospadia and absence of abdominal muscle, Immature teratoma
Dehner LP,	1973	5 m,	F	Left lower pole	Cystic	+	Immature teratoma
Kojiro M et al,	1976	40 y,	M	Left	Cystic	—	Mature teratoma with carcinoid tumor
Aubert J et al,	1978	2.5 m,	?	Left upper pole	Cystic	—	Immature teratoma
Dische MR & Johnston R,	1979	stillborn	F	Both kidneys	Cystic	+	In horseshoe kidney, Ass with multiple anomalies
Glazier WB et al,	1980	59 y,	F	Left lower pole	Cystic	—	In horseshoe kidney, Mature teratoma
Park et al,	1982	3 m,	F	Left mid & upper	Solid & Cystic	+	Immature teratoma

the kidney may be flattened and mold over the tumor[15] or be displaced usually downward without direct involvement. In the present case, although a small portion of the kidney was identified as being displaced at the lower portion of the tumor, it had undoubtedly a common capsule which it shared with the tumor.

If the pararenal teratomas are excluded, this is the 7th example of an intrarenal teratoma reported to date in English literature. There are two additional reports we could cite, but the original papers have not been available to us. Bilger et al[16] described an intrarenal teratoma in a 7-year-old girl and Ipiens Aznar and Penúela Verseda[17] in a 6-day-old baby.

As cases accumulate, it becomes clear that the intrarenal teratoma tends to occur predominantly in females and in the left kidney, and is frequently associated with other congenital anomalies, especially the horseshoe anomaly of the kidney (Table 1). Two of the seven previous cases were found with horseshoe kidney: one in a stillborn female with low set ears, micrognathia, webbed arms and multiple skeletal anomalies, and one in a 59-year-old woman[4,7]. A case of intrarenal teratoma in association with multiple anomalies consisting of meningocele, hypospadia and absence of abdominal muscle was reported in a 7 week-old male[1].

The tumors tend to be cystic and those already reported were largely cystic except for one that was described as partly solid and partly cystic. In three cases which arose in adolescents or adults, the tumor parenchyme was exclusively mature and contained no area reminiscent of glomeruloid or other nephroblastematous tissue[5~7]. In contrast, all cases arising in infants contained immature tissue elements. It is worthy to note that glomeruloid and tubular tissues suggestive of differentiating nephroblastema were described in three out of four cases[1,2,4].

With few exceptions, the occurrence of malignant germ cell tumor in infancy is restricted to the sacrococcygeal teratoma[2]. It is

difficult, however, to assess the natural history of teratoma arising in adults with certainty, because metastasis can occur in well differentiated teratomas of other organs and could presumably occur with those arising in the kidney[6,7]. Misugi and Reiner[18] reported a malignant teratoma of the liver in a 2-year-old boy. None of the already published teratomas of the kidney including those of the adults pursued a malignant clinical course.

Considering that nephroblastoma is relatively uncommon as the cause of an abdominal mass during the initial 12 months of life, one should contemplate the possibility of benign tumors such as mesoblastic nephroma, leiomyomatous hamartoma and teratoma, when an infant presents with an intrarenal neoplasm. The hazards of relying upon a clinical diagnosis of nephroblastoma without histologic confirmation have been emphasized in infants having such a benign neoplasm.

Summary

Intrarenal teratoma is extremely rare and tends to be overtreated as a malignancy before histologic diagnosis is made. This was such a case. A 3-month-old female infant presented intra-abdominal mass which was diagnosed as a Wilms' tumor clinically and at operation, and actinomycin D was initiated immediately after operation before a histologic diagnosis was obtained. The case has been reported for not only being its rarity, but also to remind to include benign lesions in the differential diagnosis of intra-renal mass during the first year of life, and the necessity of frozen section diagnosis during operation.

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