Journal of Korean Medical Science
Vol 5, No. 4, 237-242 December 1990

Retroperitoneal Teratoma with Predominance of Nephroblastic Elements

— A Case Report —

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The morphological features of a retroperitoneal teratoma in a 10-month-old girl are reported. Unlike the usual pattern of the teratoma, this tumor was composed predominantly of nephroblastomatous tissue. Histologically, glomeruloid and tubular structures were identified in nests of undifferentiated blastemal elements. Hyaline cartilage, adipose tissue, glial tissue and glands lined by mucin-secreting columnar epithelium were minor elements. A focal cystic structure lined by thin flattened epithelium was also noted.

Retroperitoneal teratoma with predominance of nephroblastic elements is of interest not only because of its rarity but also because it needs to be differentiated from extrarenal Wilms' tumor, since both of these tumors have different origins.

Key Words: *Retroperitoneal teratoma, Nephroblastic element, Extrarenal Wilms' tumor*

INTRODUCTION

Retroperitoneal teratoma is an extremely uncommon in term of primary site for extragonadal teratomas and, based upon incidence data reported in several series, these tumors account for about two to four per cent of all gonadal and extragonadal germ cell tumors. These tumors, which mainly appear in children, are approximately 90% benign (Gonzales-Crussi, 1982; Lack et al., 1987).

According to some detailed reports, renal tissue is an uncommon component of these teratomas in general. Moreover, nephroblastic elements of this retroperitoneal teratoma are extremely rare. Until recently, five cases showing teratoma with predominant nephroblastic tissue have been reported in world literature (Moyson et al., 1961; Malik et al., 1967; Ward and Dehner, 1974; Tebbi et al., 1974; Carney, 1975).

We had the opportunity to observe one case of retroperitoneal teratoma with predominance of nephroblastic elements in a 10-month-old girl. A detailed review of the literature and biologic behavior of this unique tumor are discussed.

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CASE REPORT

A 10-month-old girl was admitted for evaluation of a retroperitoneal mass. At birth she was noted to have a soft, child-fist sized, well-defined round mass in the sacrococcygeal region, along with meningomyelocele without skin defects. At that time, it was revealed through abdominal sonography and Magnetic Resonance Image (MRI) scan, that in addition to the meningomyelocele and sacral mass, there appeared a mixed echogenic mass in the left upper retroperitoneum, lateral to the spine and medial to the left kidney. The sacral mass was excised and the meningomyelocele was repaired. Histologically, the sacral mass was lipoma.

Three months later, the patient was readmitted for evaluation of a newlygrown retroperitoneal mass. On admission, an abdominal computed tomography revealed an irregularly marginated soft tissue mass at the medial aspect of the upper pole of the left kidney, measuring 3.5 × 2.5 × 3 cm. The left kidney was dis-

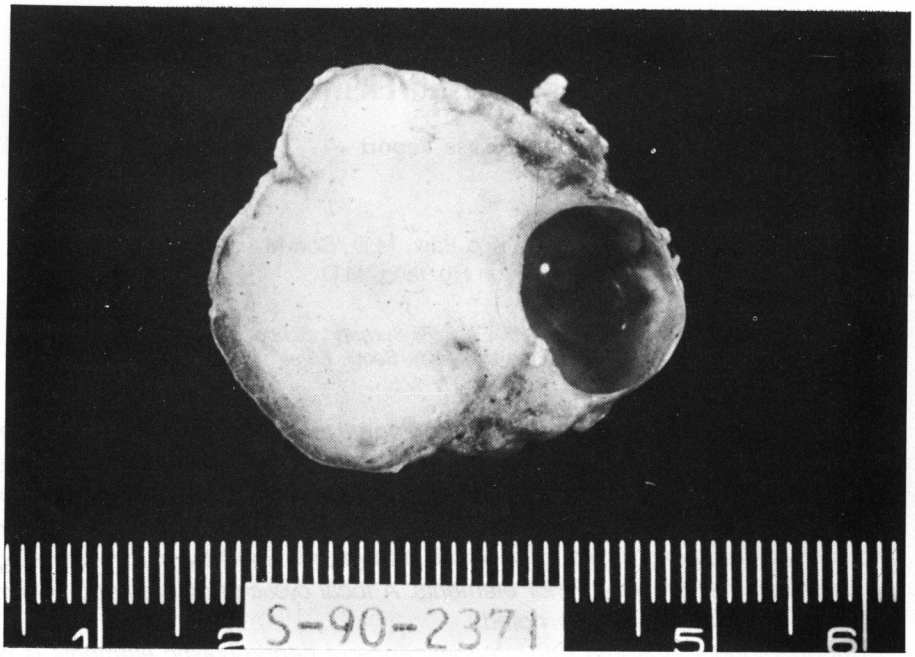

Fig 1. Cut surface of tumor shows gray-white to yellowish solid appearance with partial cystic change.

Fig 2. Light microscopic finding of solid area reveals diffuse proliferation of immature tumor cells separated by fibrous and glial tissues (H & E ×40).

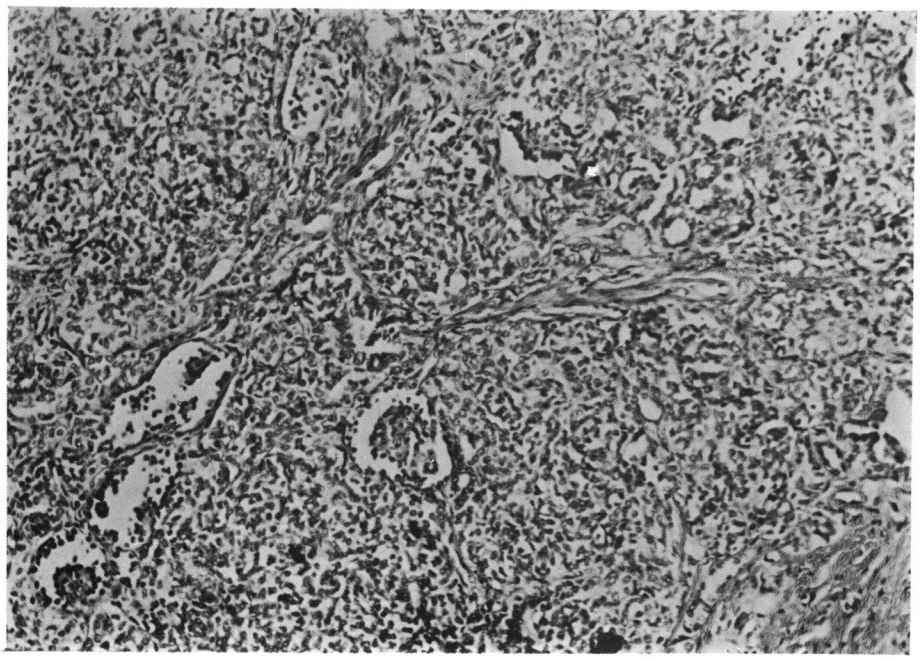

Fig 3. The tumor is composed of largely immature blastemal components with glomeruloid and tubular structures (H & E ×100).

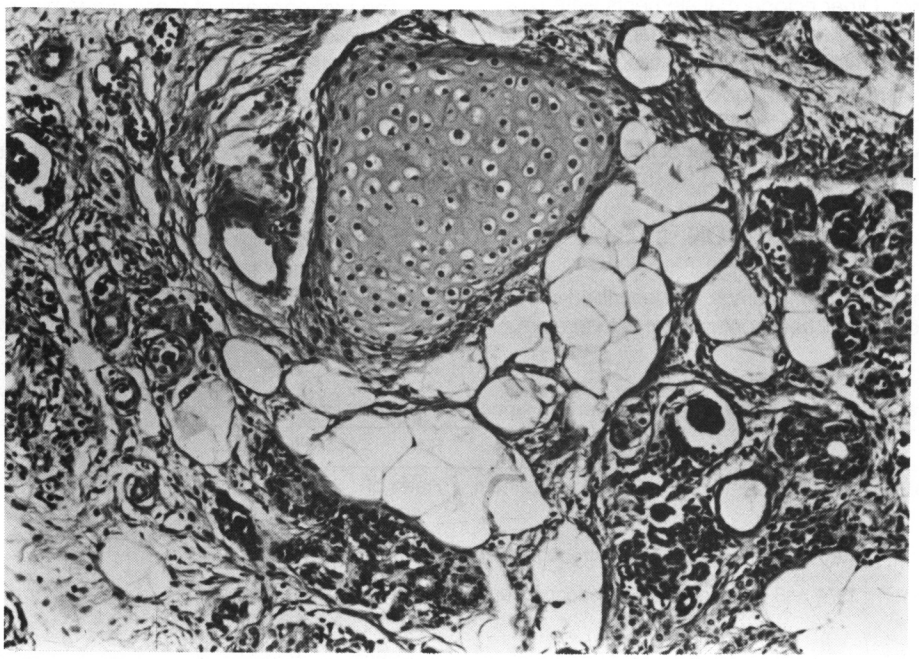

Fig 4. Hyaline cartilage and fat tissue is also seen (H & E ×100).

placed laterally and downward. With the impression of neuroblastoma, exploratory laparotomy was done. On operation, a left retroperitoneal mass, without any attachment to other surrounding organs, was found and easily removed. The left kidney and left adrenal gland were intact.

The resected specimen was a well-circumscribed mass measuring 3 × 3 × 2.5 cm. The outer surface was covered by a thin fibrous capsule. The cut surface had a gray-white to yellowish solid appearance with partial cystic change. The cyst measured 0.8 × 0.5 cm and was filled with serous fluid (Figure 1).

A light microscopic finding of the solid area revealed a diffuse proliferation of immature tumor cells, which resembled renal blastemal components, separated by mature fatty, fibrous and glial tissues (Figure 2). Approximately 70% of the tumor was composed of immature blastemal components with glomeruloid and tubular structures. The blastemal cells had scanty cytoplasm with ill-defined cellular margins and round-to-oval hyperchromatic nuclei (Figure 3).

The microscopic findings of this lesion were similar to those of Wilms' tumor of the kidney, The immature-to-mature glomeruloid structures and tubules with foci of calcifications were scattered between the teratomatous elements. Other ectodermal elements were glial tissue, ganglion cells, neuroectodermal tubules; the mesodermal elements were adipose tissue, and hyaline cartilage; and the endodermal elements were glands lined by mucin-secreting columnar epithelium. The cyst was lined by a single layer of flattened cuboidal epithelium (Figure 4).

DISCUSSION

The main histological findings we described in this report are features of nephroblastoma, with teratoma showing incomplete organogenesis. In general, renal tissue, either mature or immature, is an uncommon component of teratomas.

Willis (1935) first described a 9-week-old female with retroperitoneal teratoma that contained a single area of immature renal tissue. The appearance of the tissue as a whole recalled that of Wilms' tumor found in children. Willis' report (1935) is one of very few available reports which describe immature renal tissue in the teratoma (Ward and Dehner, 1974). Six cases of teratoma with predominant nephroblastic tissue, including our case, are reported in world literature, and are summarized in Table 1.

The locations of these six teratomas with predominant nephroblastic elements were in the retroperitoneum (three), in the sacrococcyx (two), and in the posterior mediastinum (one). Except for Carney's report (Carney, 1975), all the patients were young girls. In every case, microscopic study showed that the tumors were largely composed of immature blastema, glomeruloid, and tubular structures. The focal areas of teratomatous components were also included. In the Carney case's, combined foci of renal cell carcinoma and Wilms' tumor in the reteroperitoneal teratoma found.

The pathogenesis of these predominant nephroblastic tissues in teratoma is still obscure. This has led to a debate as to whether the origin is embryonic or neoplastic, but most authors believe the origin of this tumor to be embryonic (Ward and Dehner, 1974; Carney, 1975; Akhtar et al., 1977; Koretz et al., 1987).

In 1977, Akhtar et al. described that extrarenal Wilms' tumors can be subdivided into two groups based on their probable histogenesis. The first group is comprised of Wilms' tumors arising within teratomas. The second group includes those cases of extrarenal Wilms' tumor in which no evidence of teratomatous

Table. 1. Reported Cases of Teratoma with Predominance of Nephroblastic Elements

Author	Year	Age	Sex	Location	Size	Treatment	Follow-up
Moyson et al.	1961	3 yr	F	Post. mediastinum	Head of newborn		Died several wk later
Malik et al.	1967	6 yr	F	Lt. retroperitoneum	12×8×12cm		
Ward & Dehner	1974	3 yr	F	Sacrococcygeum	8×6×6cm	Radiation & chemotherapy	Free of tumor 12 mo. later
Tebbi et al.	1974	3 yr	F	Sacrococcygeum	6×8×6cm	Radiation & chemotherapy	Free of tumor 46 mo. later
Carney	1975	41 yr	M	Lt. retroperitoneum	Huge	Radiation	Died 4 mo. later
Present case	1990	10 mo	F	Lt. retroperitoneum	3×3×2.5cm	Chemotherapy	Free of tumor 4 mo. later

origin has been demonstrated. It is reasonable to presume that these tumors arose within embryonic rests of renal tissue that may be derived from renal anlage, such as the pronephros, mesonephros or metanephros.

Koretz et al. (1987) concluded that teratoma containing nephroblastic tissue should be classified separately, since the embryonic origin of this tumor may be different from that of the true Wilms' tumor. Therefore, careful examination of extrarenal Wilms' tumors is mandatory in order to exclude teratomatous elements. Most authors believe that the true extrarenal Wilms' tumor probably originates from embryonic remnants of mesonephric tissue (Akhtar et al., 1977, McCauley et al., 1979). Development of Wilms' tumor outside the renal parenchyme is an uncommon event. Pure extrarenal Wilms' tumors, which are not associated with a teratoma, have been situated in the retroperitoneum (Bhajekar et al., 1964; Edelstein et al., 1965; McCauley et al., 1979; Wakely et al., 1989), the uterus (Bittencourt et al., 1981), the inguinal region (Thompson et al., 1973; Ahktar et al., 1977; Luchtrach et al., 1984), the ovaries (Sabin et al., 1988), and the chest wall (Madanat et al., 1978).

Metanephric dysplastic hamartoma or renal dysplasia of the sacral region is an unusual congenital sacral tumor-like lesion characterized histologically by the presence of dysplastic and immature renal-like tissue with a predominantly glomerular and tubular structure. This lesion is regarded as a dysplastic growth (Cozzutto et al., 1983). The absence of undifferentiated cellular mesenchyme and the prevalence of well differentiated and dysplastic renal structure are the basic distinguishing features in comparison to our case. And teratomatous components, such as hyaline cartilage, or glial tissue etc., are not seen in metanephric dysplastic hamartoma of the sacral region.

In cases of extrarenal Wilms' tumor, therapeutic consideration can be based on those generally applied to renal tumors. Thus, treatment should utilize multidrug chemotherapy similar to the guidelines set by the National Wilms' Tumor Study (Madanat et al., 1978). However, the treatment of teratoma with predominant nephroblastic tissue is not settled, because of the small number of cases reported and the different types of therapy used. Malik et al. (1967) described that the patient did not required further treatment. Ward & Dehner (1974) and Tebbi et al. (1974) described that the patients were treated with radiation to the tumor bed and received chemotherapic agents.

In our case, teratoma components are mature, but approximately 70% of the area of the tumor shows histological features of nephroblastoma. We thought that our case was a malignant tumor and required further treatment. Our patient received vincristin, actinomycin and cyclophosphamide after removal of the mass. Since the operation four months after the patient has been in good health without any evidence of recurrence of metastasis.

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