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The sonographic features of the retroperitoneal masses in infants and children.

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Sonograms of 23 infants and children with retroperitoneal mass were retrospectively reviewed, including 14 Wilms' tumors, 4 neuroblastomas, 3 teratomas and 2 lymphomas. The following sonographic criteria : size, location, internal echo pattern, anechoic areas, margination and the presence of hepatic metastasis were evaluated separately in each entity of disease. Most of the sonographic features of Wilms' tumor were large-sized, sharply marginated and echogenically heterogeneous with anechoic areas located at the kidney. In all cases of neuroblastomas, the mass was an inhomogeneously echoic solid mass of extra-renal origin; the presence of tumor calcification and hepatic metastases were presented in three of four cases. Three cases of teratomas showed the sonographic characteristics as a large, well-defined complex mass containing solid and cystic components with calcification. In two cases of non-Hodgkin's lymphoma; one with mass at the paraaortic area showed multiple, lobulated, low echoic masses representing lymph node enlargement. The other mass primarily arising from the cecum showed a target lesion. The spectrum of sonographic features of these retroperitoneal masses and the diagnostic value of sonography are discussed.

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ดรุณี บุญยืนเวทวัฒน์, จันทร์ทิพย์ ศรีรัตนกุลวงศ์, สุดี ชมเดช, เกษร วัชรพงศ์, ลัดดาวัลย์ วัชระคุปต์. ลักษณะภาพอุลตราซาวด์ของก้อนรีโทรเพอริโตเนียลในทารกและเด็ก. จุฬาลงกรณ์เวชสาร 2533 กันยายน; 34 (9) : 691-704

การวิเคราะห์ลักษณะภาพอุลตราซาวด์ของก้อน retroperitoneal ในผู้ป่วยทารกและเด็ก 23 ราย ประกอบด้วย Wilms' tumor 14 ราย, Neuroblastoma 4 ราย, Teratoma 3 ราย และ Lymphoma 2 ราย ได้ผลดังต่อไปนี้ Wilms' tumor ลักษณะภาพอุลตราซาวด์พบเป็น echoic mass ที่มี echo ของก้อนคล้ายเนื้อตับ บางครั้งจะมี anechoic areas และ calcification อยู่ภายในก้อน สำหรับ Neuroblastoma ลักษณะภาพอุลตราซาวด์พบเป็น Heterogenous echoic mass ส่วนใหญ่จะมี calcification ภายในก้อน และมีการกระจายของเนื้องอกมาที่ตับ ส่วน Teratoma ลักษณะภาพอุลตราซาวด์พบเป็น echoic mass ที่ประกอบด้วย cysts และ calcification อยู่ภายในก้อน สำหรับผู้ป่วย Lymphoma 2 ราย พบเป็น lobulated low echoic mass รอบเส้นเลือดใหญ่ aorta และ inferior vena cava อยู่หน้าต่อ lumbosacral spine 1 ราย ส่วนผู้ป่วยอีก 1 ราย เป็น lymphoma ของ cecum ให้ลักษณะภาพอุลตราซาวด์เป็น target lesion.

Retroperitoneal tumor is one of the most common tumors in infants and children frequently presenting with abdominal mass.[1-3] There are a multitude of investigations including plain radiographs, excretory urography, Barium studies, Ultrasonography, Computed tomography and angiography.[2] Among these, ultrasound should be the primary screening imaging method owing to it's noninvasiveness, lack of ionization, organ function independence, and highly informative technique.[1-3] This helps to locate the origin of the mass. Besides, many reports have been published on the ultrasound features of retroperitoneal tumors that can be distinguished in each entity.[3-6] The purpose of this study was to analyse the ultrasound features of 23 cases of proved retroperitoneal tumors of known pathology (Wilms' tumors, Neuroblastomas, teratomas and lymphomas.)

Material and Methods

Gray-scale sonograms of 23 infants and children with retroperitoneal mass detected before treatment, at Chulalongkorn Hospital from 1981-1989, were reviewed. The sonographic examinations were performed on a static B-scanner or a mechanical real-time sector scanner using 3.5 or 5.0 MHz transducer. Tissue diagnoses were established following biopsy or surgical removal of tumor.

The patients ranged from 2 months to 10 years in age. Nine patients were under 2 years of age at the time of diagnosis. There were 13 boys and 10 girls. The tumors included 14 Wilms' tumors, 4 neuroblastomas, 3 teratomas, and 2 nonHodgkin's lymphomas. Each tumor was analyzed for size, location, internal echo pattern, anechoic areas, margination, and the presence of hepatic metastasis. The intensity of the internal echoes was determined by comparing the tumor with the liver parenchyma and/or renal cortex. "Hyperechoic" is more echogenic than liver parenchyma, "isoechoic" is equal to the liver parenchyma, and "hypoechoic" is less echogenic than liver parenchyma.

Results

The age and sex distributions, location, and the sonograghic features of each disease are presented in table 1, 2 and 3

Table 1. Sex and age distribution.

Diseases	Sex M : F	Total	<1yr.	1-2yr.	2-3yr.	>3yr.	Age range
			No. patient(s)				
Wilms' tumor	10:4	14	3	2	7	2	3mo. - 7 yr.
Neuroblastoma	1:3	4	2	-	1	1	8mo. - 4 yr.
Teratoma	1:2	3	2	-	-	1	2mo. - 10yr.
Lymphoma	1:1	2	-	-	-	2	3mo. - 12yr.

Table 2. Size and location of retroperitoneal tumors.

Diseases	Greatest Dimention (cm)	Location	No. patient(s)
Wilms' tumor	9.8	Intrarenal	= 14
Neuroblastoma	8.4	Adrenal	= 3
		Paravertebral	= 1
Teratoma	11	Retroperitoneum	= 3
Lymphoma	14	Paraaorta	= 1
		Cecum	= 1

Table 3. Ultrasonographic characteristics of retropertoneal tumors.

US findings	Wilms' tumor	Neuroblastoma	Teratoma	Lymphoma
Internal echo pattern				
# echogenicity				
* hyperechoic	7	2	3	-
* hypoechoic	3	1	1	2
* isoechoic	4	-	-	-
# homogeneity				
* homogeneous	5	-	-	-
* heterogeneous	9	3	4	2
# anechoic areas	5	3	2	-
Calcification	2	3	3	-
Margination				
* well-defined	12	3	2	2
* poorly-defined	2	-	2	-
Hepatic metastasis	-	-	3	-

WILMS' TUMOR

There were 4 females and 10 males ranging in age from 3 months to 7 years, with average age of 2 3/12 years. Twelth of 14 cases were under 3 years of age. The primary tumors were generally large, varying in size from 3×3×3 cm to 11×15×16 cm. The average greatest dimension of all tumors was 9.8 cm. All masses were intrarenal in location with entire or nearly entire renal involvement. Twelth tumors were sharply marginated, ten with hypoechoic rims, two with partly echogenic and partly sonolucent rims. Two tumors had illdefined margin.

The masses were hyperechoic in seven (Fig.1), isoechoic in four (Fig.2) and hypoechoic in the other three (Fig.3). Tumor calacification was detected in two cases. No liver metastasis was found in any patient. Distorted and dilated calyces were found in two cases (Fig.4).

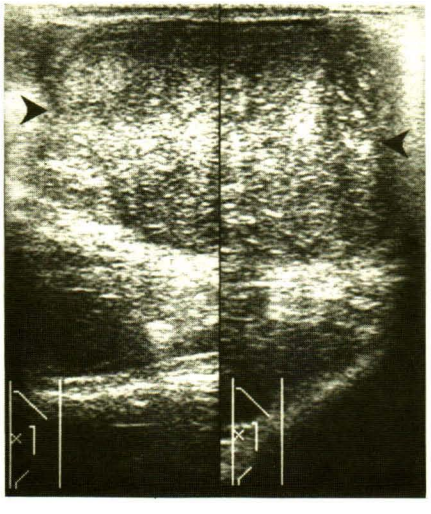


Figure 1. Wilm's tumor
Longitudinal scan showed a heterogenous hyperechoic mass with hypoechoic rim involving entired left kidney (arrow heads).

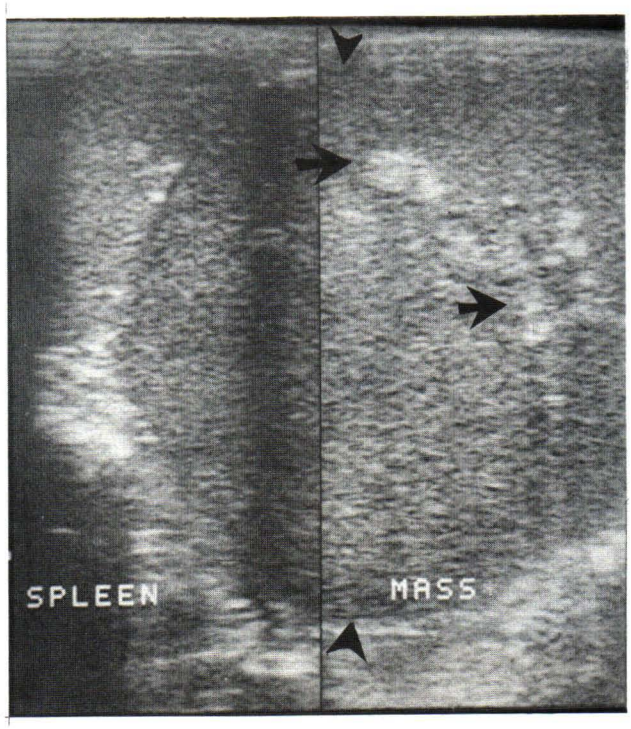


Figure 2. Wilm's tumor
Longitudinal scan showed a well defined isoechoic mass with anechoic rim (arrow heads), containing multiple calcifications (arrow heads) in the left kidney.

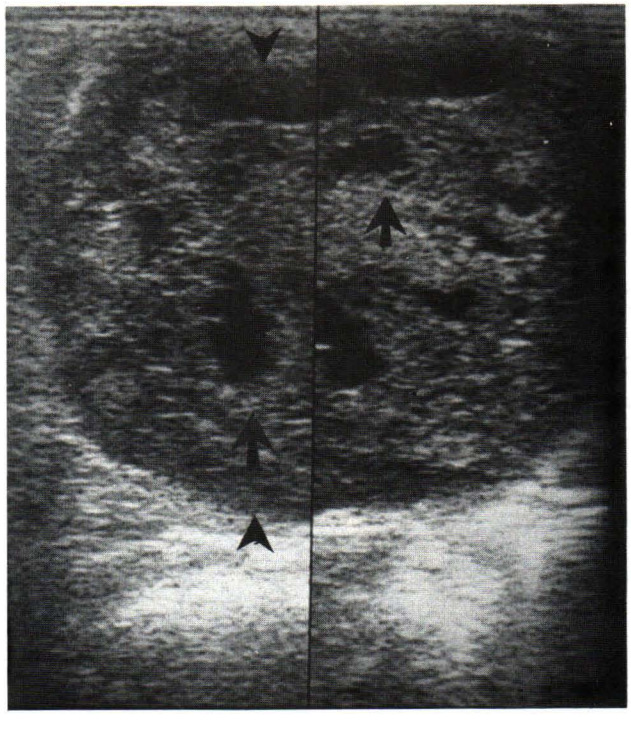


Figure 3. Wilms' tumor
Longitudinal scan showed a hypoechoic mass (arrow heads) with multiple anechoic area (arrows).

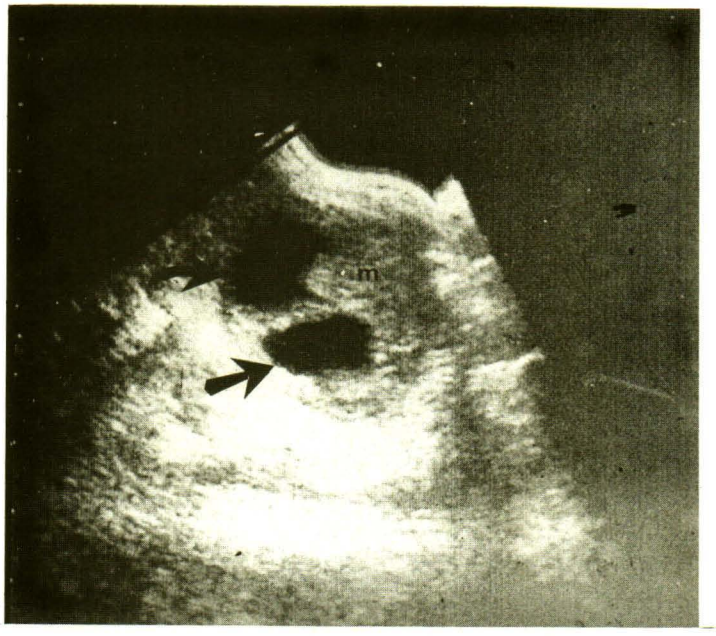


Figure 4. Wilms' tumor
Longitudinal scan through right kidney showed dilatation of pelvicalyceal system (arrows), compressed by the mass (m).

Five tumors had a homogeneous echo pattern. Five of the 9 in the heterogeneous group contained anechoic areas, varying from 1 to 4 cm in diameter, throughout the mass.

NEUROBLATIOMA

There were four children, 3 girls and 1 boy. The ages at the time of diagnosis ranged from 8 months to 4 years (mean, 2 2/12 years); of these, two were less than 1 year-old. The tumor originated from the adrenal gland in 3 patients causing downward displacement of the ipsilateral kindey (Fig.5). In one case, the mass causing upward displacement of the kidney was thought to be arising from the lumber paravertebral region (Fig.6). The primary tumor masses varied in size from 4.5×5×5.5 cm to 7.5×8×11 cm. The average greatest dimension of primary tumor was 8.4 cm.

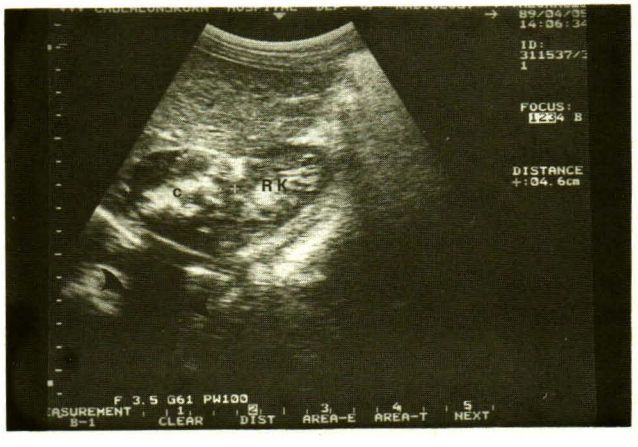


Figure 5. Neuroblastoma
Longitudinal scan showed a heterogenous echoic mass (++) located at superior aspect of right kidney (R.K.). The mass contained multiple bright echoes (c) with posterior shadowing (arrow heads) representing calcification.

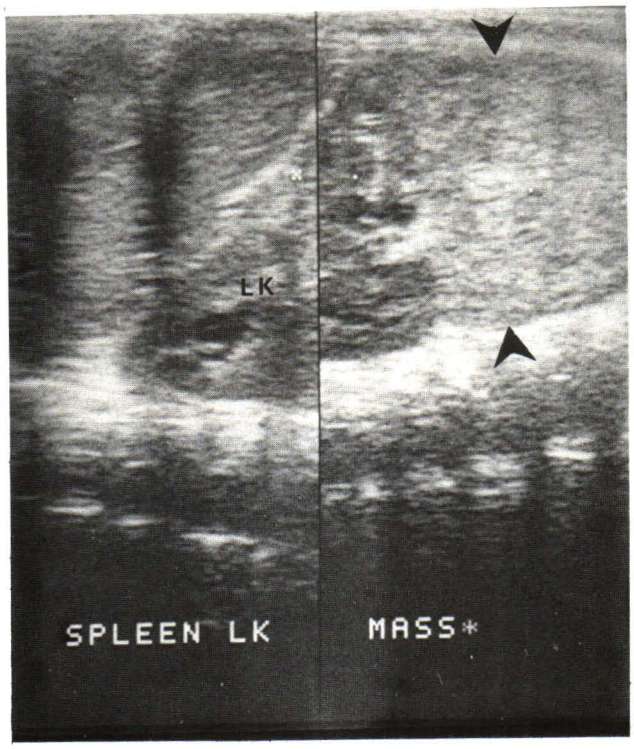


Figure 6. Neuroblastoma
Longitudinal scan showed a heterogenous hyperechoic mass (arrow heads) located inferiorly to the left kidney (L.K.).

All masses were solid and heterogeneously echogenic. Among 4 primary tumors, three were predominantly hyperechoic. Of these, two tumors contained discrete small anechoic areas. In one case, the tumor was hypoechoic. The calcification were presented in 3 of 4 cases.

The tumor margins were poorly defined in two cases. One tumor was sharply defined with hyperechoic rim. There was abrupt change in tissue texture in another case. No hypoechoic halo was seen.

Hepatic metastases were detected in 3 of 4 cases (Fig.7).

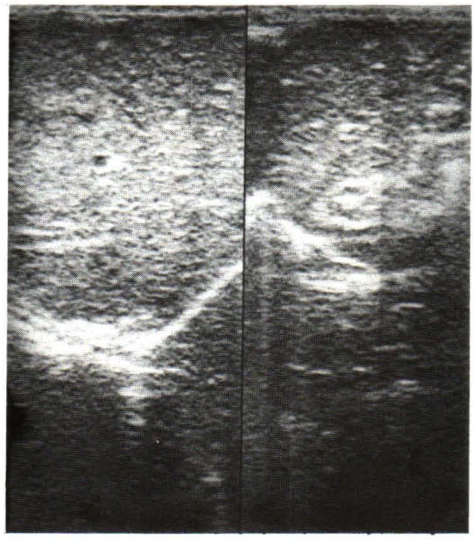


Figure 7. Neuroblastoma with liver metastases
Transverse scan of liver showed irregular hyperecho throughout.

TERATOMA

There were 2 girls and 1 boy, aged 2 months, and 10 years (mean age, 3 8/12 years). The tumors ranged from 5×8×10 cm to 7×9×12 cm, with the average greatest dimension of 10 cm. Two tumors were found superoposteriorly to the kidney extending downward and causing slight anteroinferior displacement of the ipsilateral kidney. Another tumor was located inferior to the kidney. All tumors were well demarcated with hypoechoic rim and completely separated from othe adjacent structures. The components of the masses wer predominantly solid, with multiple anechoic areas repre senting cystic components (Fig.8,9). There were smal bright echogenic masses eccentrically protruding int the cystic part (Fig.10).

All three tumors showed bright echoic foc scattered throughout with acoustic shadowing representin calcifications.

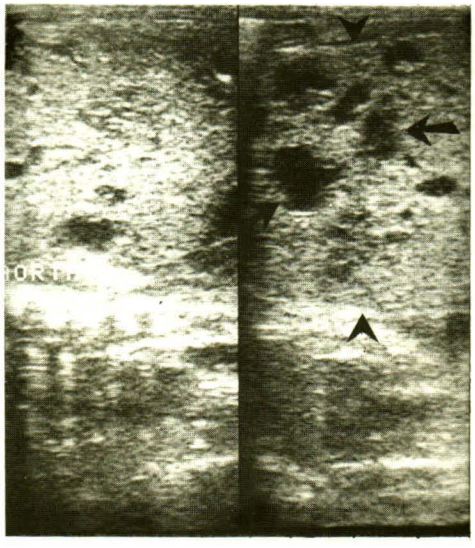


Figure 8. Teratoma
Longitudinal scan showed a heterogenous hyperechoic mass (arrow heads) with multiple cystic components (arrows) located superiorly to the right kidney.

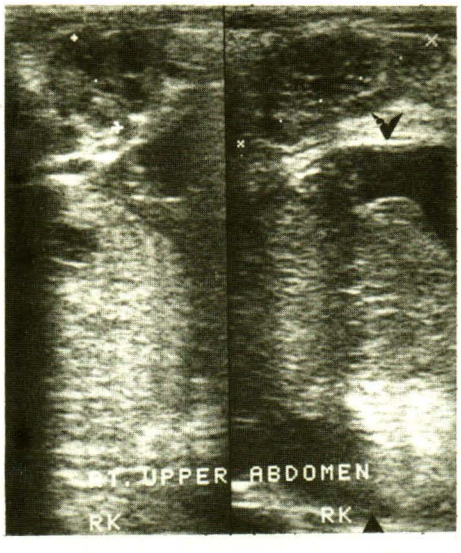


Figure 9. Teratoma
Longitudinal scan showed a heterogenous echoic mass (arrow heads) at inferior aspect of right kidney (R.K.) with multiple cysts (c).

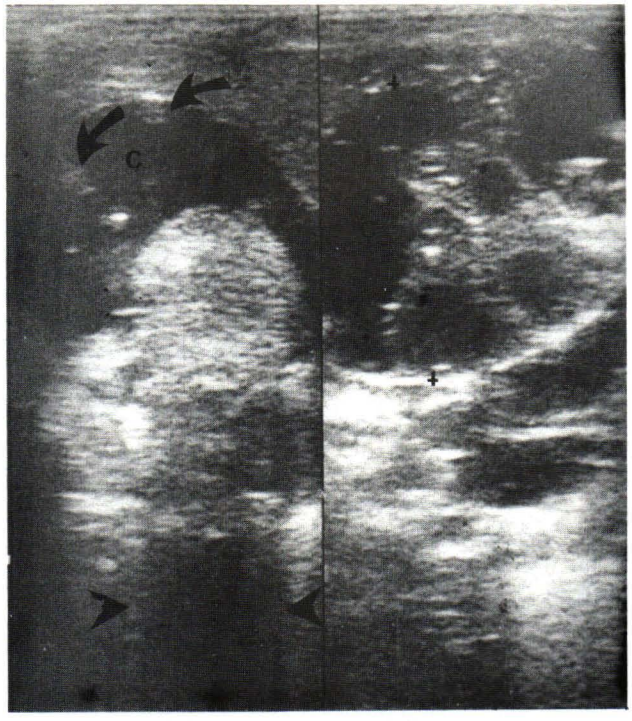


Figure 10. Teratoma
Longitudinal scan showed a slightly low echoic mass (+ +) at superior aspect of right kidney containing multiple cysts (c), calcification : bright echoes with posterior shadowing (arrow heads), and mural nodules : echoic masses outgrowth from inner surface of a cyst (arrows)

LYMPHOMA

There were two patients :

Case No. 1 : A 8×11×15 cm-mass proved to be nonHodgkin's lymphoma was located around the aorta and inferior vena at the lower mid abdomen of a 12-year-old boy. An inhomogeneously hypoechoic solid pattern with lobulation was seen. This mass had well-definded, low echoic margin (Fib.11). Bilateral hydronephroses and hydroureters resulted from obstructed ureters by the mass were also noted.

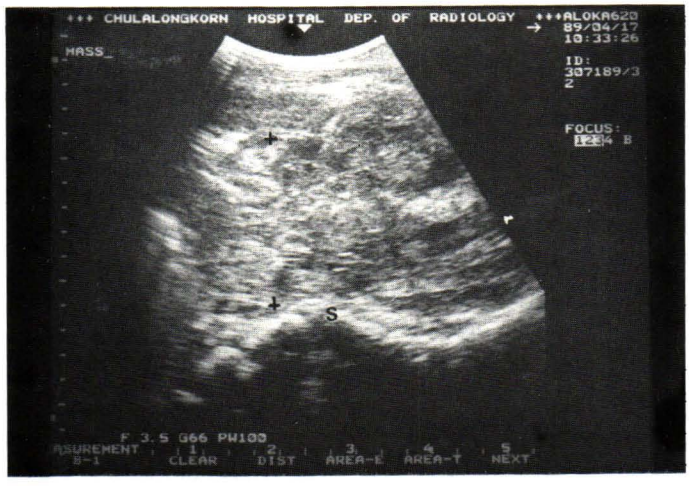
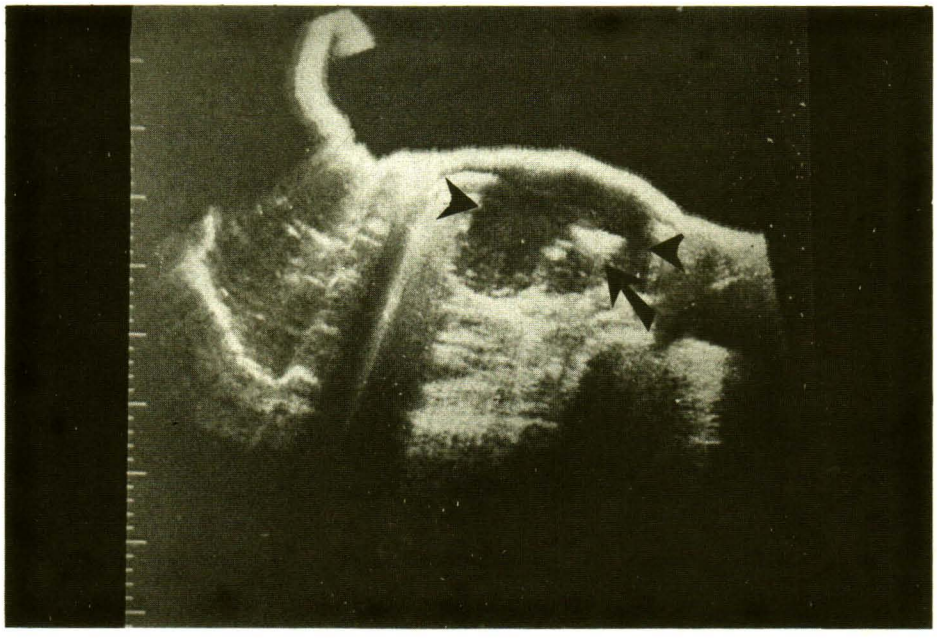


Figure 11. Lymphoma of periaortic and pericaval lymph nodes.
Longitudinal scan showed lobulated inhomogenous hypoechoic mass with low echoic rim (+ +) lying anterior to lumbosacral spine (s).

Case No. 2 : Sonogram in a 3-year-old girl revealed a right-lower-quadrant mass, measuring about 6.5×9×12 cm. The mass was complex with a nearly entire anechoic area and an eccentric, brightly reflective area producing a target-like lesion. (Fig.12) An acoustic shadowing produced by intertinal gas was noted. No liver metastasis was identified in both patients.



Discussion

The usefulness of ultrasound in evaluation of masses in pediatric patients is well documented. Our results confirm its value. Let us discuss each entity separately.

WILMS' TUMOR

Wilms' tumor (nephroblastoma) is one of the most common malignant abdominal tumors in childhood[1] It has approximately the same overall incidence as neuroblastoma and accounts for approximately 10 percent fo all childhood malignant tumors[2] It frequently occurs at less than 8 years of age with an average age at the time of diagnosis of 2 1/2 to 3 years.[1-3] There is no sex predilection. Most of our children were less than 2 years old; but contradictory to most series, there was a male predominance. It may be associated with congenital anomalies, and some cases are familial.[1,7] Heritable tumors tend to have a higher frequent of bilaterality.[1]

Most tumors are large, predominantly solid and well circumscribed,[1,4,5] as they were in our study. The margins appear as echolucent lines, echodense lines, or abrupt changes in tissue texture.[4] The discrete hypoechoic rim representing a thin rim of compressed renal tissue pathologically[4] was seen in ten of our patients. However dense fibrous pseudo-capsule, frequently seen,[1,4] were found only in parts of two tumors.

The textures of Wilms' tumor are more variable sonographically. The echogenicity of the tumor itself is similar to liver parenchyma. The intensity of echoes varies from hypoechoic to hyperechoic;[4,5] our findings concur with these series. Wilms' tumor has a heterogeneous cellular growth pattern consisting of fibrous tissue, cartilage, fat, smooth muscle, striated muscle, myxomatous stromas, ect.[2,4] The random pattern of cellular components is related to the echogenic heterogeneity.[4] Nine of our cases were echogenically heterogeneous which agreed with these authors.

Small anechoic areas are seen in most cases corresponding to areas of necrosis and hemorrhage pathologically.[1,2,4-6] Our findings, which showed five cases with anechoic areas, concurred with them. Calcification is uncommon,[1,2,4] as was detected in two cases. Evidences of compressed normal renal parenchyma and

distorted or dilated pelvicalyceal system may be demonstrated.

All of our tumors were intrarenal. The diagnosis of Wilms' tumor by ultrasonography at first glance seems easy : a solid renal mass in a child is Wilms' tumor.[4] However, when the mass is very large, it is difficult to prove that it arises from the kidney, making differentiation from neuroblastoma not always possible.[1,5] Rarely, Wilms' tumor may be entirely extrarenal.[1,8]

Mesoblastic nephroma is the most common solid real mass in a neonate and has different clinical, pathologic, and biologic characteristics from Wilms' tumor.[1,4] Cystic Wilms' tumors are also rare.[5,9] The differential diagnosis includes hydronephrosis, multicystic kidney, adult polycystic kidney, multiple simple cysts, etc.[2,5]

NEUROBLASTOMA

Neuroblastoma is one of the most common solid tumors of childhood, occuring with approximately the same frequency as Wilms' tumor.[1,2] Neuroblastoma and its more fully differentiated forms respectively, ganglioneuroblastoma and ganglioneuroma, arise from primitive sympathetic neuroblasts of the embryonic neural crest. Spontaneous regression from the more malignant to the more benign forms may occur.[1] They may occur throughout the length of the sympathetic chain (neck to pilvis) and adrenal gland. Three of our cases were adrenal in origin and the remainder originated in the paravertebral region. Sixty-five percent of neuroblastomas are located in the abdomen; approximately two-thirds of these arise in the adrenal gland. The remainder of abdominal or pelvic tumors almost always originate in the paravertebral sympathetic chain or the presacral area.[2]

When initially diagnosed, 50 percent of patients are less than 2 years of age, 75-85% of patients are less than 4 years of age,[1,2,10,11] as all of our patients.

Since the greatest survival is in patients with surgical resection of early-stage tumor, diagnostic imaging and accurate staging remain important for improvements in treatment of this disease.[10] Neuroblastoma provides a variety of sonographic appearances because their tissue patterns depend on their growth characteristics and on the degree of hemorrhage, necrosis, and/or calcification.[12,13] Generally it is an extrarenal solid mass with mixed echogenicity, as they were all in our small series. Anechoic areas representing necrotic tumor can predominate.[12,13] The characteristic sonographic feature of hyperechoic lobules in the mass correlated pseudocapsule described by Amundson et al was not seen in our study.[13]

Tumor calcification (bright echoes with acoustic shadowing) is present in 70 percent of tumor and, when present, is the useful sonographic evidence.[1,12] Tumor margins are generally poorly defined as in our two cases. Since the majority of cases of neuroblastoma have metastasis at the time of diagnosis and one of the common site is the liver; the presence of a suprarenal mass with hepatic lesions should strongly suggest the diagnosis of neuroblastoma.[10] Our series showed the liver metastasis in three.

Differentiation from other solid tumors, such as Wilms' tumor or lymphoma, may not be always possible on the basis of sonogram.[4,12] In the newborn presenting with a suprarenal mass, adrenal hemorrhage should be differentiated from necrotic neuroblastoma by performing serial sonograms.[10,12]

Ultrasonography has limited value in evaluating the disease, especially when compared with the role played by CT in the assessment of the primary tumor and the extent of spread.[2,10-12,14,15] Sonography can be used to detect and follow liver metastasis. It may be the preferred imaging method when artefacts created by surgical clips are encountered in CT.[12]

TERATOMA

Teratomas are embryonal neoplasms that composed of several recognizable somatic tissues, arising from pluripotetial embryonal cell.[16] In the literatures[17-19] they arise, in order of frequency, in the ovaries, testis, anterior mediastinum, retroperitoneum and sacrococcygeal region; but some authors[20,21] reported a different order. More than 170 cases of retroperitoneal teratomas[16-21,24-28] and an additional 2 cases from Thailand[22,23] have been reported. They frequently present in infancy and childhood[16-18,20,25] and a few, more than 40 percent, are in the first decade of life[25] as in our patients.

Teratomas arise independently from tissue in the retroperitoneal space and not from adjacent structures or organs.[26] They are most commonly found in the left retroperitoneal space, typically in the region of the upper pole of the kidney.[18,24] All our cases were in the right, two at the typical region.

Accurate preoperative diagnosis is still important since retroperitoneal teratomas may be confused with sarcomas at surgery causing a lesion that could have been removed to be given up as unresectable.[24] Plain film findings of teratoma include teeth or bone and fat. The calcifications, although present in 74% of benign teratomas, are observed in more than a quarter of malignant ones.[26]

Sonographically the teratoma may be a predominantly cystic, solid, or complex mass.[23,25,27-30] The

contents of the tumor may include bright echogenic structures with shadowing representing calcifications, a fat-fluid or fluid-debris level,[25,27] and densely echogenic area which may represent fatty tissue[29] or those of simple fluid. All masses of our series were complex mass predominantly of solid. The cystic components vary from those of simple fluid to those of complex fluid characterized by internal echoes within the fluid. No layering was found in our series.

One of the sonograghic features in our series is noteworthy. The dermoid plug, mural nodules or Rokitansky protuberance, an outgrowth from inner surface of a cyst, containing hair and other atypical tissues, have been described sonographically[30,31] as a round hyperechoic mass which form an acute angle with the wall of the cyst.

Although we can detect calcifications in all of our cases, CT is superior to sonography in detecting calcification and delineating bone and tooth fragments.[25,27]

Since fatty tissue can be eighter echogenic or echofree depending on tissue interface with differences in acoustic impedance,[29] sonograms did not permit confident differentiation among fat, other forms of soft tissue, and calcific deposits. CT is more specific than ultrasonography in the diagnosis of fatty tumors.[25,27]

Apart from other retroperitoneal tumors, retroperitoneal teratomas must be differentiated with clinical significance from a rare entity, the so-called fetus in fetu[32,33] which is almost always benign and retroperitoneal in location. The clue to the differentiation between these two conditions is the recognition of the vertebral column.

LYMPHOMA

Lymphomatous involvement of paraaortic nodes is seen in approximately 25% of patients newly diagnosed with Hodgkin's disease and in 40% of nonHodgkin's lymphoma patients,[34] Ultrasound has been shown to detect retroperitoneal nodal lymphoma with an 80-90% level of accuracy.[35,36] Brascho et al[35] reports a 98% accuracy in the sonographic estimation of nodal size in retroperitoneal nodes greater than 2 cm in diameter, and both Brascho[35] and Rochester et al[36] report sonographic accuracy in the detection of retroperitoneal lymphadenogpthy compatible to that of lymphangiography. Admittedly, approximately 10% of normal size lymph nodes contain disease, and this pathology would likely be detected by lymphangiography only.[37]

The sonographic appearance of lymphomatous involvement of paraaortic nodes has been characterized as low echo or anechoic nodal enlargement.[34] A "dumbbell" or mantle-shaped mass draped over the prevertebral vessels is seen in bilateral involvement. Unilateral involvement of the paraaortic or paracaval chains is seen as sausage-shaped mass on a parasagittal scan.[38] If only a segment of a lymph node chain is involved, it is difficult to differentiate this lesion from other retroperitoneal tumors.

Adenopathy, secondary to nonlymphomatous neoplasms, or inflammatory processes such as retroperitioneal fibrosis may be indistinguishable from lymphoma.[39] Basically the masses seen in retroperitoneal fibrosis often have smoother anterior borders than lymphomatous masses, but this is not a reliable sign. Inflammatory masses may be lobulated; and nodal mass in lymphoma may be atypically more echogenic and inhomogeneous.[34]

Ten to twenty percent of newly diagnosed cases of nonHodgkin's lymphoma will have the gastrointestinal tract as their initial stie of involvement.[40] lymphomas of the gastrointestinal tract occur most frequently in the mid-sixth decade. They are, however, the commonest tumor of the gastrointestinal tract in chilhood, especially in children younger than 10 years.[41] The most common gastrointestinal site for lymphoma is the stomach (48-50%), followed by the small intestine (30-37%), the ileocecal region (12-13%7) and the colon (10-12%).

The sonographic appearance of intestinal lymphoma is quite characteristic.[41] A mass is apparently arising from the bowel as a complex mass with completely sonolucent areas representing markedly thickened bowel wall. A central or slightly eccentric, densely reflective area is indicative of intestinal mucosa and/or mucus. The lumen of the bowel can be indentified by variable degrees of acoustic shadowing due to gas. These findings produce a characteristic target lesion to allow differentiation from other abdominal masses.

Conclusion

Ulrasound is of value to distinquish intraabdominal mass of which it arises. This series studed the ultrasonographic characteristics of retroperitoneal tumors providing some specific clues in each entity of disease that would help to make the specific diagnosis.

The results confirmed that each tumor had the specific ultrasonographic feature concurring with previous reports. Wilms' tumor often showed a solid intrarenal mass with liver parenchymal like echogenicity. Neuroblastoma was predominantly heterogenous, echoic extrarenal mass with frequent calcification. The characteristic echoic lobule described by Amunson et al was not found in our series. All teratoma showed a retroperitoneal mass containing solid, cystic components with

calcification. One case of nodal lymphoma of paraaortic area presented with inhomogenous low echoic mass dissimilar from many reports as a low or anechoic mass. The other intertinal lymphoma at cecum showed the characteristic target lesion.

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