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Retroperitoneal tumors in infancy and childhood

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Özet

Yatırılarak tedavi gören tüm hastalarımıza göre abdominal kitle sıklığı % 5 bulunmuştur. Bunların % 2'si retroperitoneal kitle tanısı almıştır. Retroperitoneal kitlelerin büyük çoğunluğunu nefroblastoma (% 22.5), hidronefroz (% 12.5), iliopsoas absesi (% 10), nöroblastoma (% 5), ganglionöroma (% 5) meydana getirmektedir. Abdominal nöroblastomanın serimizde beklenen sıklıkla görülmeyişi retroperitoneal kitle sıklığını düşüren etmen olduğu sonucuna varılmıştır.

Anahtar kelimeler: Retroperitoneal tümörler

Summary

The analysis of retroperitoneal masses detected between 1980-1988 revealed that only 5 % of the total number of the children admitted to the Ankara University Hospital had a surgical abdominal mass.

20 % of these masses were retroperitoneal in origin. The great majority of the retroperitoneal masses are nephroblastomas (22 %), hydronephroses (12 %), iliopsoas abscesses (10 %), neuroblastomas (5 %) and ganglioneuromas (5 %). The reason for the low rates of retroperitoneal massses may be unexpected low incidence of neuroblastoma in the presented series.

Key words: Retroperitoneal tumors

Introduction

The retroperitoneum and masses that originated in this region have an important place in pediatrics and pediatric surgery [17,18].

Materials and Medhods

A retrospective analysis of the patinets with abdominal masses admitted to the department of Pediatric Surgery, Ankara University Hospital from January 1st 1980 to January 1st 1988 was performed with data available in the computers. The following parameters were recorded: Age, sex, symptoms and findings, laboratory examinations and investigations for diagnostic purposes, operative and histopathologic diagnoses, operative interventions and chemoteraphy and radiotheraphy regimens if the masses were neoplasia. The diagnosis was made based on history, physical examination, labarotory findings such as complete blood count, blood smear, biochemical analysis of urine and blood, ultrasound, CAT scan, scintrgrams and excretory urograms.

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Results

As a result of analysis, 202 cases of abdominal masses were recorded.

The patients with retroperitoneal masses were only 40 cases and making 20 % of all abdominal tumors. ın other words 1 % of the patients admitted to the ward had a mass in retroperitoneal space. Most of the cases were nephroblastomas (22.5 %) hydronephroses (12.5), iliopsoas abscess (10 %) neuroblastomas (5 %) and ganglioneuromas (5 %) (Table I).

There were six female and three male patients with nephroblastoma, consisting the 0.23 % of all of the patients with abdominal masses in an eight-year-period. While the youngest patients was one year old, the oldest was aged 13, having an average age of 4.8. Five of the nine nephroblastomas originated from the left kidney. The nephroblastomas were staged as IV (6 cases), II (2), III (1). Three patients with stage IV nephroblastoma were treated by subtotal excisions, but the remaining were treated by nephrectomies, following by chemotherapeutic agents including Vincristine, Actinomycin D, Cyclophosphamide and irradiation.

In a patient with liver metastases incisional biopsy was performed, while in a patient with pulmonary metastases metastasectomy was undertaken a

Table I. Retroperitoneal tumors

Diagnosis	N	%
Wilm's Tumor	9	22.5
Mesoblastic Nephroma	1	2.5
Malignant Mesenchymal Tumor	1	2.5
Hydronephrosis	5	12.5
Pyonephrosis	3	7.5
Multilocular Cystic Kidney	1	2.5
Polycystic Kidney	1	2.5
		52.5
Neuroblastoma	2	5.0
Adrenal Cortical Ca	1	2.5
Ganglioneuroma	2	5.0
Retroperitoneal Teratoma	1	2.5
Pancreatic Islet Cell Adenoma	1	2.5
Rhabdomyosarcoma	1	2.5
Immunoblastosarcoma	1	2.5
Retroperitoneal Lymphoma	2	5.0
Abscess	4	10.0
Hematoma	2	5.0
Hydatid Cyst	2	5.0
Total	40	100

week later after nephrectomy. Where the tumoral mass infilitrated the adjacent sructures, operative borders were marked by silver clips or stainless steel sutures in order to define the irradiation site. One of the nephroblastoma cases expired due to pulmonary embarassment (Table II).

In two seven year-olds, ganglioneuromas were treated by total excisions without further treatment. Neuroblastomas were detected in two patients, one with stage IV and the other with stage III. While the patient with stage III neuroblastoma was treated with total tumor excision, only an incisional biopsy was obtained in the patient with stage IV neuroblastoma. Both cases received chemotheraphy and radiotherapy. In the case with adrenal cortical adenocarcinoma, partial hepatectomy was added the surgical excision of the orijinal tumor due to local invasion into the liver (Table IV).

The remaining malignant retroperitoneal tumors apart from nephroblastomas and neuroblastomas were lesions such as rhabdomyosarcoma, teratoma, malignant mesenchymal tumor of renal origin, mesoblastic nephroma, immunoblastosarcoma, Hodgkin's disease and Burkitt lymphoma. The patient with malignant mesenchymal tumor of renal origin was treated by nephrectomy and chemotherapy and irradiation. The cases with mesoblastic nephroma in whom diagnosis mass was established prenatally as a retroperitoneal was treated by nephrectomy. While surgical treatment of the case with Hodgkin's lymphoma was total "en block" excision of tumor and aorto-femoral grafting, for the remaining cases only biopsies were performed. The case with gastric outlet obstruction because of retroduodenal teratoma, intestinal continuity was established by gastrojejunostomy and added biopsy procedure for histologic confirmation. Table III.

Surgical therapy in four patients (three were females), with iliopsoas abscess, consisted of drainage with laparotomy and antibiotics. The causitive microorganisms were Klebsiella and Staph. aureus.

Five of the retroperitoneal masses were hydronephroses. All of the patients were male and the youngest infant was aged one month, while the oldest one was eight years old. Hydronephroses

Table II. Wilms tumors, age and sex distribution

Case	Sex	Age (Y)	Location	Stage	Intervention
1	F	7	Right	IV	Subtotal excision VAC+Irradiation
2	M	1	Right	III	Right nephrectomy VAC
3	F	1.5	Left	II	Left Nephrectomy VAC+nephrectomy VAC+ırradiation
4	F	6	Left	II	Left nephrectomy VAC+ırradiation
5	F	3.5	Right	IV	Right nephrectomy Biopsy (liver metastases)
6	F	13	Left	IV	Left Nephrectomy Metastasectomy (lung)
7	M	2	Left	IV	Left Nephrectomy VAC+ırradiation
8	M	3	Right	IV	BSubtotal excision VAC+ırradiation
9	F	6	Left	IV	Subtotal excision VAC+ırradiation

Table III. Summary of operative and medical treatment of the patients with neuroblastomas, adrenal cortical Ca and genglioneuromas

Case	Sexs	Age (Y)	Location	Stage	Case
Ganglioneuroma					
1	M	7			Total tumor excision
2	M	7	R.adr.		Total Tumor Excision
Neuroblastoma			R.adr.		
1	M	6	CA	III	Total Tumor Excision
2	M	6	R.adr.	IV	Biopsied
Adrenal Cortical					
1	F	4		IV	Adrenalectomy+Partial Hepatectomy

Table IV. Other retroperitoneal malignant tumors

Diagnosis	Sex	Age (Y)	Intervention
Rhabdomyosarcoma	M	7	Laparotomy+biopsy
M.Mesenchymal renal tm	M	3	Right nephrectomy
Mesablostic neproma	M	5 days	Left nephrectomy
Immunoblastosarcoma	M	8	Lymph node biopsy
Hodgkin's disease	F	10	En block resection + aorto-femoral bypass.
Lymphoma	M	11	Biopsy
Teratoma	M	3	Biopsy+gastrojejunostomy

due to posterior urethral valves were bilateral in two cases. First one was a newborn and the other an eight-month-old infant. Treatment of these cases were endoscopically performed valve excisions and vesicostomies. At 20th postoperative day, the newborn was lost because of renal insufficiency, but the infant survived with a mild hypertension. The remaining three cases with hydronephroses, the right kidneys were affected and the etiology of ureteropelvic obstructions were found to be urolithiasis in one patient and congenital origin in two. Nephrectomy was performed in these cases since excretory urograms and renal scintigraphies demonstrated no excretion but renal cortical severe atrophia.

Three patients had pyonephroses due to renal tuberculosis (one case) and urolithiasis (two), and again, being nonfunctional, nephrectomy was performed (Table IV).

In two patients with retroperitoneal masses laparotomy proved that the origin of the masses were hematomas, organized one was excised and the other case was treated with only drainage procedure. Further investigations did not indicata any coagulophaty, but blunt abdominal trauma. Retroperitoneal hydatid cysts appeared in two cases who were treated with cystectomy and capitonating. A 13 year old female patient having a well capsulated pancreatic islet cell adenoma with no endocrine involvement healed after simple excision. A case of multiloculer cystic disease of the left kidney was treated by nephrectomy, histopatolgical examination showed no malignant degenerations.

Table V. Summary of the patients with hydronephrosis and pyonephrosis including age and sex dirtributions and surgical treatment

Case	Sexs	Age (Y)	Location	Etiology	Intervention
Hydronephrosis					
1	M	8M	Bilateral	Post. Urethral Valve	Valve Excision Vesicostomy
2	M	1M	Bilateral	Post. Urethral Valve	Valve Excision
3	M	5	Right	Congenitall	Vesicostomy
4	M	8	Right	Urolithiasis	Nephrectomy
5	M	16	Right	Congenital	Nephrectomy
Pyonephrosis					Nephrectomy
1	M	4	Right	Tuberculosis	Nephrectomy
2	M	10	Right	Urolithiasis	Nephrectomy
3	M	11	Left	Urolithiasis	Nephrectomy

Discussion

Review of large series of abdominal masses shows that 75-80 % of the surgical masses in infancy and childhood is retroperitoneal in nature, but the remaining 20-25 % is intracoelemic [12,17,18]. Kirk (11) et al, reported that, of the surgical masses in infancy and childhood, 78 % was retroperitoneal (being 55 % of renal) and only 10 % was hepatobiliary. In the presented series, of the abdominal masses, only 20 % was retroperitoneal. The reason for the low rates of retroperitoneal masses may be explained by unexpected low rate of neuroblastoma, whic is expected to be the ame with nephroblastoma [13,14]. These differences led to a significantly higher rate of intra-coelemic masses in the presented series than that of others (80 % versus 10-25 %). Of the retroperitoneal masses, approximately two-thirds are of renal origin, slightly less than half being Wilms' tumors and a few mesoblastic nephromas [3,11,17,18].

Approximately half of the retroperitoneal masses were of renal origin in the presented series, and half of the renal tumors were benign masses such as hydronephroses, pyonephroses and multicystic and polycystic disease of kidney. While two-fifths of the hydronephroses apeared in infancy, the remainder three-fifths were diagnosed in childhood.

Hyronephroses migh be detected inutero, but usually are detected in infancy and childhood [10]. In some reports it had been reported that the frquency of hydronephroses has its peak in the first year of life, indeed in the first six months [10,19], but in large series the peak incidence appeared in five year-old children [20]. The causes of hydronephroses those appeared in childhood are usually due to obstruction of ureteropelvic junction which is presumed to occured during intrauterine development.

Hydronephroses due to ureteropelvic junction obstruction are generally affected unilaterally and both kidneys equally [4,9,10,15]. In the presented series, on the contrary, right kidneys were affected in all cases with hydronephroses resulting from ureteropelvic junction obstruction.

One-thirds of the benign renal masses were pyonephroses in this series. It has been reported that pyonephrosis is a rare complication of corrective surgery of hydronephroses in childhood [4,9]. While hydronephroses appeared in five cases; unfortunately, three pyonephroses considering the hydronephroses may be result of the late diagnosis due to parental and professional delay and insufficent sanitation measures.

Howewer amongst the presented patients the causes of pyonephroses and hydronephroses have been found by detecting the renal pelvic stones leading pyeloureteral obtructions.

Retroperitoneal abscesses usually appear as an iliopsoas abscesses and their etiology are not very clear; the inflamation in the adjacent tisues and organs, as well as those of the posterior mediastineum, which may infiltrate via esophagial and aortic orificies are implicated [7,8].

Howewer, in the presented patients with ilopsoas abscesses, no inflamation could be demonstrated of the adjacent tisues and organs and posterior mediastinum, which led us to consider the pesented cases as primary ilopsoas abscess [7,8].

Although ultrasound is less accurate specifically that axial tomography and other investigative methods, this method may be useful as a guide for diagnostic puncture and evacuation [7,8,16].

The most important factor hindering the total excision of tumors in the presented cases with Wilm's tumors was that six of nine cases were in an advanced stage (stage-IV). But with subtotal excision leaving a minimal tumoral tisue led the patients to maximally benefit from chemotreraphy and irradiation.

The mean age of patients with nephroblastoma was 4.8 years which is significantly higher that the review of the large series [2,5]. Although the presenting symptom of Wilms' tumor is usually an abdominal mass not extending the midline, of a female patients aged 13 years, having a nephroblastoma arising from the upper pole of left kidney with multiple pulmonary metastases, the ab-

dominal mass was extended the midline due to hemorage in the medial part of the mass.

The review of repots showed that CAT scan and nuclear magnetic resonance imaging are useful differing the Wilm's tumor from neuroblastoma and other retroperitoneal masses and may demonstrate more accurately intraabdominal spreads of tumors and invasion of lymph nodes [1,6].

In conclusion: The rate of retroperitoneal masses among the patients with surgical abdominal masses was 20 %, which is quite different from the literature (75-80 %). Follow up of the patients with malignant masses was not adequate due to insufficient multidisiplinary approach which is needed for reinforcement and reorganization. Although one of the masses was detected prenatally, the remaining was diagnosed with a significant parental and professional delay. This concept which is life saving has to be emphasized in every occasion.

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