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A 4-Year-Old Boy with Constipation and Abdominal Pain

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CASE PRESENTATION

History

A 4-year-old boy was brought to the emergency department with a long history of constipation and recent onset of abdominal pain. His mother stated that he did not have diarrhea, vomiting, or fever, but noted that "his belly is getting hard." He had a small bowel movement 2 days previously. On the day of presentation, he woke up with abdominal pain.

The patient previously had been in good health and had regularly seen his pediatric primary care physician. He had never had surgery or an illness requiring hospitalization.

Physical Examination

On examination, the following vital signs were obtained: oral temperature, 98.5°F; heart rate, 145 bpm; respiratory rate, 30 breaths/min; blood pressure, 108/59 mm Hg. He was pale and appeared to be uncomfortable due to the abdominal pain.

The abdominal examination revealed a large mass in the right side of his abdomen. The mass was firm and slightly tender. He also had guarding. He had bilaterally descended testicles and no evidence of inguinal hernias. His musculoskeletal and neurologic examinations showed normal results, and skin examination revealed decreased turgor and coolness of peripheral extremities, with no rashes, bruising, or petechiae.

- **What is the differential diagnosis of this child's abdominal mass?**
- **What diagnostic evaluations are indicated?**

EVALUATION OF AN ABDOMINAL MASS

Constipation is a common presenting complaint in infants and young children. The most common cause is functional constipation,[1] which is benign and transient. Other potential causes are listed in **Table 1**.

Abdominal masses have a wide range of clinical presentations. They may present with pain, vomiting, constipation or, less commonly, intestinal obstruction. It is not unusual for a child with an abdominal mass to present because a family member notices a protuberant mass. The identification of an abdominal mass in a child is a cause for concern because of the possibility of malignant disease. In addition, even nonmalignant conditions can sometimes be serious and warrant prompt evaluation and treatment.

Key Point

Constipation is not always "benign." Pediatricians should be alert for possible presenting signs and symptoms of childhood malignancy and other serious problems that may present as constipation.

Clinical Examination

Evaluation of an abdominal mass begins with a thorough history and physical examination to detect other possible findings. Special points of focus in the physical examination include location of the mass, size, consistency (eg, firm, tender, mobile), the presence of bruit, and the presence of ascites. Other points to note are whether it crosses the midline and whether it extends into the pelvis.

When a swelling in the abdomen is palpable, first one should make sure that it is not a normal structure. In children, especially those with a thin or lax abdominal wall, feces in the colon may simulate an abdominal mass. The pelvic colon is frequently palpable, particularly when loaded with hard stool. The cecum is often palpable in the right lower quadrant as a soft rounded

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Table 1. Causes of Constipation

Gastrointestinal
Hirschsprung's disease
Drugs
Anticholinergics
Narcotics
Antidepressants
Metabolic
Hypokalemia
Hypercalcemia
Hypothyroidism
Diabetes mellitus
Neurologic
Spina bifida
Muscular dystrophy

Table 2. Selected Causes of Abdominal Masses in Children

Condition	Associated Laboratory Results
Wilms' tumor	Hematuria
Neuroblastoma	Urine VMA ↑, urine HVA ↑, serum ferritin ↑
Non-Hodgkin's lymphoma	Serum urate ↑, bone marrow + for malignant cells
Germ cell tumors	Serum HCG ↑
Hepatoblastoma	Serum AFP ↑
Hepatoma	Serum AFP ↑

AFP = α-fetoprotein; HCG = human chorionic gonadotropin; HVA = homovanillic acid; VMA = vanillylmandelic acid.

swelling with distinct borders. The transverse colon is sometimes palpable in the epigastrium.

Next, one should make sure that the swelling does indeed lie in the abdominal cavity and not in the anterior abdominal wall. With the child lying supine, ask him or her to lift the head and shoulders off the pillow while you press firmly against the forehead. Now, feel the swelling again. If it disappears or becomes much less obvious, then it lies within the peritoneal cavity; if it remains the same size, it is located within the layers of the abdominal wall.

Differential Diagnosis

The differential diagnosis of abdominal pain and a mass in a child is extensive. An abdominal mass in a child can have many causes, including benign and malignant tumors originating in various organs (**Table 2**), intestinal disorders, and several renal conditions. The most common neoplasms in the renal area that occur in childhood are Wilms' tumor, neuroblastoma, and congenital mesoblastic nephroma. Wilms' tumor is by far the most frequently encountered renal lesion in young children. Congenital mesoblastic nephroma occurs in neonates and is usually large. Other considerations include a renal cyst, polycystic kidney disease, and hydronephrosis. Rhabdomyosarcoma of the bladder also can present with an abdominal mass. In addition, the presence of an abdominal mass may be due to an enlarged spleen or liver, lymphoma, germ cell tumor, or mesenteric cysts.

Although neuroblastoma occurs more often than Wilms' tumor in infancy, it almost always originates in the adrenal gland. Typically, the child with a neuroblastoma appears ill and has a large mass with a speckled appearance on radiographs owing to calcified foci. The tumor is irregular on palpation and typically crosses the midline of the body. A neuroblastoma may arise from sympathetic nervous tissue anywhere in the body, but it most often develops in the abdomen. Up to 65% of primary neuroblastoma tumors in children arise in the abdomen.[2] The presentation depends on the local effects of the solid tumor and its metastases. Signs and symptoms include anorexia, abdominal pain, and distention. Occasionally there is associated weight loss, fever, or decreased appetite. Pallor or weakness may be caused by anemia from slow hemorrhage into the mass or from infiltration of the bone marrow. Other manifestations of marrow involvement include bleeding, bruising, and sometimes bone pain.

Renal tumors other than Wilms' tumor are infrequent in childhood. Wilms' tumors account for 6% to 7% of childhood cancers, whereas the remaining renal tumors account for less than 1%.[3] The most common non-Wilms' renal tumors are clear cell sarcoma of the kidney, rhabdoid tumor of the kidney, renal cell carcinoma, mesoblastic nephroma, and multilocular cystic nephroma. Collectively, these tumors account for fewer than 10% of the primary renal neoplasms in childhood.

Although cancer can develop in children of any age, certain malignancies have a predilection for specific age groups. For example, neuroblastoma, retinoblastoma, and Wilms' tumor most commonly occur in children between birth and age 4 years.

Laboratory Examination

Laboratory studies are helpful in the evaluation of differential considerations of abdominal pain and/or

an abdominal mass. Initial studies include a complete blood count and chemistry panel (ie, levels of electrolytes, liver transaminases, total and direct bilirubin, albumin, calcium, phosphorus, uric acid, and lactate dehydrogenase). An elevated leukocyte count with a left shift may be seen because of infection caused by obstructive effects of an underlying malignant process. Anemia can be caused by chronic disease or may indicate hemorrhage into the tumor. Urinalysis can detect hematuria or proteinuria, either of which may be seen with a renal tumor. Tumor markers are useful in determining the diagnosis and sometimes the prognosis of certain tumors. Serum neuron-specific enolase, as well as spot urine tests for homovanillic acid and vanillylmandelic acid should be obtained if neuroblastoma or pheochromocytoma is suspected. Quantitative β-human chorionic gonadotropin levels can be elevated in patients with liver tumors or germ cell tumors. α-Fetoprotein is excreted by many malignant teratomas and by liver and germ cell tumors.

Imaging Studies

Abdominal radiographs are the initial imaging study for most abdominal masses and provide information as to the location of the mass as well as the presence or absence of calcifications, which are commonly seen in patients with neuroblastoma.

When an abdominal mass is detected in a child, a radiologist with pediatric experience should perform an ultrasound examination of the abdomen. If the ultrasound examination does not clearly identify the origin of the mass, abdominal computed tomography (CT) scanning should be performed. CT scanning is currently the most powerful and versatile imaging procedure for the evaluation of an abdominal mass.

If the mass is confirmed to be of renal origin, immediate referrals to a pediatric oncologist and a pediatric surgeon are indicated. CT scanning is essential to developing a differential diagnosis and to determine the size and location of a primary tumor and the presence and extent of metastatic disease. In addition, the contralateral kidney requires imaging to confirm its presence, function, and possible disease involvement. Because renal malignancy often spreads to the lung and bones, in patients with suspected Wilms' tumor, bone scan and CT scan of the chest should be performed to exclude metastatic disease.

DIAGNOSIS OF CASE PATIENT

Laboratory and Radiographic Studies

The results of laboratory studies of the case patient are shown in **Table 3**. A noncontrast CT scan of his abdomen was obtained and showed a large renal mass (**Figure**). A contrast-enhanced CT scan of his abdomen and pelvis showed a mass arising from the upper pole of the right kidney and occupying the right lumbar and right upper quadrant region. The patient was transferred to a tertiary care center, where the tumor was biopsied and a diagnosis of Wilms' tumor was confirmed. The contralateral kidney was not involved.

Table 3. Laboratory Values of Case Patient

Variable	Result	Normal Range
Blood/serum		
Leukocyte count (× 10^3/mm^3)	29.9	5–17
Neutrophils	73.1%	30%–65%
Platelet count (× 10^3/mm^3)	631	150–400
Hemoglobin (g/dL)	8.8	10.2–15.2
Hematocrit	27%	34%–48%
Electrolytes (mEq/L)		
Sodium	138	137–147
Potassium	3.9	3.6–5.2
Chloride	103	99–112
Carbon dioxide (mEq/L)	21	23–32
Blood urea nitrogen (mg/dL)	9	1–22
Glucose (mg/dL)	196	70–118
Creatinine (mg/dL)	0.4	0.1–1.4
Lactate dehydrogenase (U/L)	737	105–215
Urine		
Ketones	Negative	Negative
Specific gravity	1.013	1.001–1.035
Protein	Negative	Negative
Blood	Negative	Negative
Bilirubin	Negative	Negative
Glucose	Negative	Negative
Feces		
Occult blood	Negative	Negative

WILMS' TUMOR

Epidemiology and Pathogenesis

Wilms' tumor is the fourth most common malignancy in children.[4] Nearly 500 cases are reported annually in the United States.[5,6] It is the most common intra-abdominal solid tumor of childhood. It also is the most common renal malignancy of childhood.[7] It occurs with an annual incidence of 7 cases per million

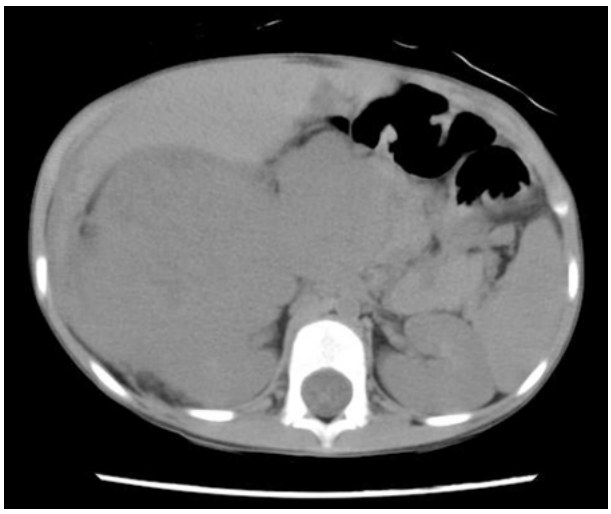

Figure. Abdominal computed tomography scan of the case patient, showing a large right renal mass displacing the liver and colon.

children younger than 15 years. The peak incidence of Wilms' tumor is at age 3 to 4 years,[8] and 80% of patients present before age 5 years.[9] Bilateral Wilms' tumor occurs in approximately 5% to 8% of patients.

Wilms' tumor is thought to arise in rests of mesodermal cells within the kidney. Grossly, Wilms' tumor typically is an intrarenal solid or cystic mass that displaces the collecting system. The tumor extends into the renal vein in 40% of cases. It occurs with increased frequency in patients with aniridia, hemi-hypertrophy, cryptorchidism, hypospadias, and other genitourinary abnormalities. The risk of Wilms' tumor is markedly increased in children with Beckwith-Wiedemann syndrome, Denys-Drash syndrome, Bloom syndrome, and WAGR syndrome (*W*ilms' tumor, *a*niridia, *g*enitourinary malformation and mental *r*etardation).[10–12] Children with these syndrome anomalies should be checked periodically for Wilms' tumor.

Key Point

A number of genetic and familial conditions are associated with an increased risk of childhood cancer; however, most children with Wilms' tumor present without associated anomalies.

Clinical Manifestations

The most frequent sign of Wilms' tumor is an abdominal or flank mass, which often is asymptomatic. Masses are frequently discovered by parents or on routine physical examination. The tumor often is quite large at the time of diagnosis, with replacement of most if not all of the involved kidney. The mass is generally smooth and firm and rarely crosses the midline. Masses vary greatly in size at the time of discovery. About half of affected children have abdominal pain, vomiting, or both. Abdominal pain occurs in 30% to 40% of cases.

Wilms' tumor may present with renal signs and symptoms, such as hypertension, gross hematuria, and flank pain. Hypertension may be due to renin secreted by the tumor or to renovascular hypertension from displacement of the kidney or renal artery by tumor growth. Wilms' tumor presenting only with microscopic hematuria is exceedingly rare.[13]

Key Point

Wilms' tumor must be suspected in any young child with an abdominal mass.

Diagnostic Imaging

CT offers several advantages in evaluating a possible Wilms' tumor. These include confirmation of intrarenal tumor origin, which usually rules out neuroblastoma; detection of multiple masses; determination of the extent of tumor, including vena cava involvement; and evaluation of the opposite kidney. On CT scans without contrast enhancement, areas of low density indicate necrosis. Areas of hemorrhage and small focal calcifications generally are less common and less prominent in Wilms' tumor than in neuroblastoma.

Pulmonary metastases are evident on chest radiographs in 10% to 15% of patients with Wilms' tumor at the time of diagnosis.[14] Chest CT provides better delineation of tumor metastases than do plain film radiographs.

Wilms' tumors often are very large at presentation and can cause severe distortion of adjacent organs, including the inferior vena cava. Determination of whether there is direct invasion of the inferior vena cava or adjacent structures can be difficult but is clearly important to know prior to surgery. Magnetic resonance imaging can be used to confirm these findings if needed and to define further the extent of tumor extension. Magnetic resonance imaging also is useful in identifying tumors and assessing the extent of disease.

Treatment

Staging of Wilms' tumor is based on whether the primary tumor is confined to the renal capsule; the presence or absence of abdominal lymph node involvement; and hematogenous dissemination to distant sites (eg, liver, lung, bone, brain). Treatment includes surgical

resection and chemotherapy for virtually all affected children and additional radiotherapy for those with advanced disease or adverse prognostic features. This approach leads to cure rates exceeding 80%.[15]

Nephrectomy followed by postoperative adjuvant chemotherapy remains the mainstay of treatment for unilateral Wilms' tumor. Surgical resection of the primary tumor is important for both diagnosis and treatment. The National Wilms' Tumor Study Group advocates surgical resection at diagnosis, which yields the most accurate staging information. The International Society of Pediatric Oncology recommends preoperative chemotherapy or radiation therapy, with the aim of decreasing tumor size and promoting fibrosis, hence reducing the risk of intraoperative tumor spillage. Presurgical treatment with chemotherapy may be used to promote shrinkage of the tumor and improve outcome.

Renal failure is a concern for patients with bilateral Wilms' tumor, with the incidence of end-stage renal disease approaching 15% at 15 years posttreatment.[16] Therefore, attempts at renal-sparing procedures are warranted in children with bilateral Wilms' tumor.

Prognosis

The prognosis is influenced by tumor size, stage at presentation, and histology. The collaborative treatment of Wilms' tumor has resulted in an excellent survival for most children with the disease. More than 60% of patients with all stages of Wilms' tumor generally survive. Patients with stage I through III disease have a cure rate varying from 88% to 98%.[17,18]

CONCLUSION

The complaint of constipation warrants the physician performing a thorough history and physical examination. If an abdominal mass is present, appropriate imaging should be obtained. If the diagnosis of cancer is suspected, consultation with a pediatric hematologist or oncologist is indicated. The combined medical expertise of pediatric surgeons, pediatric oncologists, pathologists, radiologists, and radiation therapists is necessary for the diagnosis and treatment of childhood cancers. Rapid diagnosis helps to ensure that appropriate therapy is given in a timely fashion and optimizes the chances of cure. **HP**

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