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A Child with an Abdominal Mass

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CASE PRESENTATION

Initial Presentation and History

A 4-month-old girl was brought by her parents to the acute care clinic for evaluation of irritability, decreased oral intake, and a distended abdomen noted during bathing the night before. The parents had also observed yellow scleral discoloration and clay-colored stools over the previous several days. The child had a negative past medical history, including an uncomplicated vaginal delivery at term. Her prenatal ultrasound examination was normal. She did not have any hospitalizations or surgeries and was not taking any medications. The parents denied weight loss, fever, trauma, travel history, or recent sick exposures.

Physical Examination

The child's vital signs were normal. Height and weight were between the 50th and 75th percentiles for age. She had mild jaundice and a distended abdomen, with a right upper quadrant mass extending below the right costal margin beyond the midline. The mass was slightly tender to touch, had a rubbery texture, and was dull to percussion. The liver edge could not be identified by palpation. The spleen was not enlarged. No ascites were evident. The remainder of the examination was unremarkable.

- **What is the approach to evaluation of a child with an abdominal mass?**

ASSESSMENT

The patient's age is one of the most important factors that help narrow the potential etiologies of an abdominal mass in a child as likely etiologies differ between neonates and infants/children (**Table 1**). Important history components include the length of time since the mass was found, rapidity of growth, and signs of gastrointestinal or genitourinary obstruction. The presence of constitutional symptoms, such as pallor, anorexia, fever, or weight loss, may point toward a malignant lesion, but these findings are not specific. In neonates and young infants, information from prenatal ultrasonography examinations and other prenatal interventions during pregnancy can be helpful. The presence of oligohydramnios or polyhydramnios on prenatal ultrasound might suggest nonacquired disease processes affecting the developing fetal renal system.

A thorough physical examination can be difficult in the very young or uncooperative child. A parent's lap is a good substitute for the examination table, especially with anxious and apprehensive young patients. The infant or toddler should lay supine with the abdomen exposed for inspection for protrusion, bulging, or asymmetry. The examiner's hands should be warm when touching the patient to minimize discomfort and opposition to the examination. Distraction by the physician or parent can be helpful for a more relaxed abdomen. Auscultation for bowel sounds is necessary to assess for intestinal obstruction. Initial light palpation of all 4 quadrants and the flank areas is essential. A second circuit of palpation can allow deeper examination. Percussion helps detect organ or mass size and assists in differentiating the underlying components. Solid masses and fluid-filled cysts are typically dull to percussion, while air-filled structures are tympanitic. The examination should also assess for guarding or tenderness indicative of an inflammatory or infectious process.

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Table 1. Differential Diagnosis of Abdominal Mass in Children

- Neonates
  - Renal
    - Hydronephrosis*
    - Multidysplastic kidney*
    - Mesoblastic nephroma*
    - Renal vein thrombosis†
    - Polycystic kidney disease†
    - Wilms' tumor†
    - Rhabdoid tumor†
  - Pelvic
    - Ovarian cyst
    - Hydrocolpos
    - Hydrometrocolpos
    - Gastrointestinal duplication
- Infants and children
  - Retroperitoneal
    - Neuroblastoma
    - Wilms' tumor
    - Lymphoma
  - Liver
    - Hepatoblastoma*
    - Embryonal sarcoma†
  - Gastrointestinal
    - Duplication
    - Meckel's diverticulum
    - Fecal mass
  - Pelvic
    - Ovarian cysts
    - Teratomas
  - Other
    - Omental or mesenteric cyst

*Common.

†Rare.

Table 2. Suggested Initial Evaluation of Abdominal Mass

- Radiologic imaging
  - Plain abdominal radiograph
  - Sonogram
  - Computed tomography scan or magnetic resonance imaging*
- Laboratory studies
  - Complete blood count with differential
  - Electrolytes (including calcium, phosphorus) blood urea nitrogen, creatinine
  - Uric acid and lactate dehydrogenase
  - Urinalysis
  - Urine homovanillic acid and vanillylmandelic acid*
  - Serum β chorionic gonadotropin and alpha-fetoprotein*

*If clinically indicated.

Key Point

The patient's age is among the most important factors that help narrow the potential etiologies of an abdominal mass since its causes differ between neonates and infants/children.

DIAGNOSTIC STUDIES

Plain abdominal radiographs should be the first imaging studies performed in the evaluation of a suspected abdominal mass. Such radiographs may help delineate the location and density of the mass and can also provide valuable information regarding intestinal obstruction, including the presence of multiple air fluid levels or absence of air in the rectum. Calcifications might indicate the presence of a tumor-like neuroblastoma or teratomas or lithiasis in the renal or biliary tract. Sonography is a very useful adjunct study in the work-up of abdominal masses. It is usually inexpensive and readily available, does not involve radiation exposure, and seldom requires sedation. Sonography can identify the organ of origin for the abdominal mass as well as the type of tissue components present (solid versus cystic). More specific anatomic information can be obtained by computed tomography (CT) scan or magnetic resonance (MR) imaging. When a malignant lesion is suspected, CT scan of the chest, abdomen, and pelvis can be done to determine mass extension and infiltration into adjacent organs and vessels. MR imaging of the brain and spine is warranted in patients with neurologic deficits.[1,2]

Useful laboratory studies include a complete blood count with differential and a chemistry panel with assessment of electrolytes and uric acid and lactate dehydrogenase levels (**Table 2**). Anemia, neutropenia, or thrombocytopenia may suggest bone marrow infiltration. When abnormalities in more than one bone marrow cell line are uncovered, further evaluation with a bone marrow aspiration and/or biopsy is usually recommended. Elevated levels of uric acid and lactate dehydrogenase are suggestive of rapid cell turnover associated with malignancies. Electrolyte abnormalities can be caused by kidney involvement or tumor lysis syndrome. Lesions involving the urinary system may present with proteinuria or hematuria on urinalysis.

Other tests should be tailored to the nature and location of the abdominal mass. Elevated levels of

homovanillic acid and vanillylmandelic acid in urine can be seen in cases of neuroblastoma or pheochromocytoma. Serum β chorionic gonadotropin and alpha-fetoprotein are used as tumor markers that aid in diagnosis and follow-up of certain tumors, such as teratomas and liver and germ cell tumors.[1,2]

Work-up for an abdominal mass may be initiated locally since radiologic imaging is widely available. This work-up may aid in directing the patient to a primarily surgical or medical service if specialized care is needed. Available information, including scans and radiographs, should be sent promptly with the patient upon transfer to specialized centers. Providing this information gives pediatric radiologists an early opportunity to discount false readings and will minimize the risk of repetitive studies and decrease anxiety and cost.

Key Point

Plain abdominal radiographs should be the first imaging studies to evaluate an abdominal mass, while sonography is an inexpensive, radiation-free ajunct imaging modality that can determine the origin and extent of an abdominal mass.

CASE PATIENT: WORK-UP

A plain abdominal radiograph showed a normal bowel gas pattern and no evidence of calcifications or fecal masses. An abdominal sonogram subsequently revealed an 8-cm cystic mass located between the stomach and the liver, compressing the common bile duct, which was dilated proximally, indicating possible obstruction. Results of laboratory evaluation showed a hemoglobin of 8.5 g/dL and normal platelet and leukocyte counts and differential. Serum electrolytes and uric acid and lactate dehydrogenase levels were within normal limits. She had elevated levels of total and direct bilirubin (6.7 and 4.4 mg/dL) and γ-glutamic transferase, and normal transaminase levels, pointing toward biliary injury. Serum β chorionic gonadotropin and alpha-fetoprotein levels were within normal limits, and the urinalysis did not show hematuria or proteinuria. CT scan of the abdomen showed a 7.3 cm × 7.5-cm cystic mass with an intracystic fluid level, pushing against the stomach, pancreas, and porta hepatis (**Figure**).

- **What are the most common causes of abdominal mass in children, and how are they managed?**

MANAGEMENT

As discussed above, age is the most important point in differentiating the potential causes of abdominal mass. This section reviews the approach to the most commonly found masses in neonates and in infants and children.

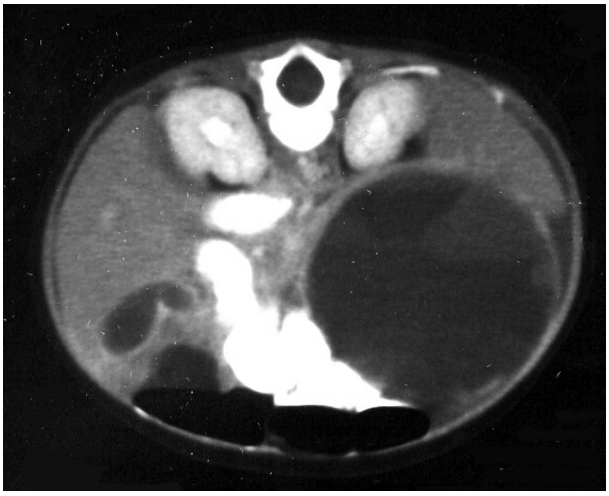

Figure. Abdominal computed tomography scan revealing a well-circumscribed cystic mass compressing the stomach and pancreas.

Neonates

Over half of palpable masses in neonates originate from the genitourinary tract. Hydronephrosis and multidysplastic kidney are the most common etiologies.[3] Hydronephrosis results from obstruction at the ureteropelvic junction, ureterovesicular junction, or the bladder outlet and can be unilateral or bilateral. With hydronephrosis, sonography shows a dilated renal pelvis surrounded by and communicating with several cystic structures (calyces). Renal scintigraphy can demonstrate the level of obstruction and assess renal function. The management of children with hydronephrosis may be supportive as many cases resolve spontaneously. Decompression may be required when there is risk of significant renal compromise or ongoing infection. Resection of nonfunctioning kidneys is indicated for complications such as infections and severe hypertension.[4–6] Multidysplastic kidney is the most common form of renal cystic disease in the first year of life.[3] The exact cause of multidysplastic kidney is unknown but may be related to an obstructive developmental defect. Most cases are sporadic and unilateral and usually present with such symptoms as abdominal mass or pain, hematuria, or urinary tract infection. The affected kidney is nonfunctional and has cysts of various sizes, causing a "grape cluster" appearance on sonography. Outcome is generally favorable after excision if the contralateral kidney is normal.[1,4,5]

Mesoblastic nephroma is the most common solid renal tumor in neonates; it results from proliferation of early nephrogenic mesenchyma. Most cases present as

masses or, less commonly, with hematuria and may be detected prenatally. Associated paraneoplastic syndromes have been described, mainly hypercalcemia and hypertension.[7,8] Mesoblastic nephroma most often is a benign lesion that is successfully treated by surgical resection. Several instances of metastasis have been described.[9,10] Other rarely encountered masses in neonates derived from the genitourinary system include polycystic kidney disease, renal vein thrombosis, Wilms' tumor, rhabdoid tumor, hydrocolpos, and hydrometrocolpos.[11,12]

Ovarian cysts commonly occur in neonates and may also be diagnosed prenatally. They are mostly functional cysts stimulated by fetal, placental, and maternal hormones. A significant proportion of cysts undergo spontaneous regression within the first few months of life, and the incidence of malignancy in these cysts is extremely low. Large-sized cysts, measuring more than 5 cm, may be percutaneously aspirated to minimize the risk of torsion. Surgical intervention in neonates is generally discouraged as a primary therapy and is reserved for persistent or recurrent cases.[13,14]

Abdominal masses in neonates may originate from the gastrointestinal tract. Duplications are cystic congenital abnormalities of the gastrointestinal tract that can occur at any level from mouth to anus but most commonly involve the ileum, followed by the esophagus and duodenum. Duplications vary in size, have spherical or tubular shapes, and may or may not communicate with the enteric lumen. They can present as asymptomatic masses or with signs of obstruction, bleeding, and perforation. Malignant transformation has rarely been reported.[15] Sonography is helpful in establishing the diagnosis. Meckel scan can be useful because of the frequent presence of gastric mucosa. Occasionally, duplications are only recognized at the time of surgery. Surgical resection is usually curative.[16,17]

Infants and Children

Malignant lesions are more commonly encountered as the cause of abdominal masses in infants and children than in neonates. The most common tumors are neuroblastomas, Wilms' tumors, and lymphomas.

Neuroblastoma. Neuroblastoma is the most common malignancy in infants and the most common extracranial solid tumor in childhood.[18] Neuroblastomas arise from neural crest cells within the sympathetic chain or adrenal medulla, with 60% to 70% of cases originating within the abdomen.[1,19] Approximately 550 new cases are diagnosed in the United States every year.[19] Presenting symptoms include a palpable mass, pain, weakness, and failure to thrive. Other associated signs include periorbital ecchymoses, exophthalmos, and Horner's syndrome (miosis, ptosis and anhidrosis). Urinary catecholamines are elevated in 90% to 95% of cases. CT scanning or MR imaging is needed to delineate the tumor size, extent of invasion of adjacent structures, and presence of metastatic disease. Bone scan and skeletal radiographs are helpful in defining bony involvement. Bone marrow invasion should be evaluated by bilateral bone marrow biopsies. The outcome depends on the tumor stage and the patient's age at diagnosis, but the behavior of neuroblastoma is not always predictable. N-*myc* oncogene amplification occurs in approximately 20% of such tumors and is strongly associated with poor prognosis.[2,20] Chemotherapy and surgical resection followed by radiation therapy may be employed. A variant of neuroblastoma affecting infants, with dissemination limited to bone marrow, skin, or liver, has an exceptionally good outcome with spontaneous tumor regression. Patients with neuroblastoma should be immediately referred to a tertiary center with pediatric oncology and surgical expertise.[1,2]

Wilms' tumor. Wilms' tumor is the second most common abdominal tumor in childhood and the most common primary pediatric renal malignancy. It is an embryonal renal neoplasm, with 450 new cases reported annually in the United States.[3] Presentations include a flank or abdominal mass, left-sided varicocele, hematuria, and hypertension. Such masses can be quite large at diagnosis because they can go unnoticed due to their retroperitoneal location and are usually painless unless hemorrhage or rupture occurs. Wilms' tumor may occur in association with other congenital anomalies or syndromes including sporadic aniridia, isolated hemihypertrophy, cryptorchidism, Beckwith-Wiedemann syndrome, Denys-Drash syndrome, and WAGR complex (Wilms' tumor, aniridia, genitourinary malformations, mental retardation). In such associations, Wilms' tumor is more likely to be bilateral and may present at a younger age.[21] Approximately 15% of patients will have metastatic disease at diagnosis, most commonly affecting the lungs followed by the liver and regional lymph nodes.[22] Sonography is the best initial imaging technique to confirm the kidney as the organ of origin and to estimate the tumor size. Major blood vessels should be assessed to determine the extent of intravascular tumor thrombi if present. A CT scan with contrast is helpful to determine the degree of kidney invasion and evaluate for metastasis. The contralateral kidney should be assessed carefully for possible involvement. Treatment includes surgery, if possible, radiation, and chemotherapy. Four-year survival rates range

from 95% for patients with low stage and favorable histology to less than 25% for advanced initial disease and unfavorable histology.[21] Similar to neuroblastoma, such patients should be cared for in a specialized pediatric center.

Lymphomas. Lymphomas are the third most common malignancies in childhood. Sixty percent are non-Hodgkin lymphomas and one third of these have abdominal disease.[1] Signs and symptoms include abdominal pain, gastrointestinal obstruction, or a palpable mass. Intussusception, secondary to a lymphomatous lead point, can occasionally be the presenting picture. Sonography, often confirmed by CT scanning, can determine the primary lesion and other organ involvement. These lesions tend to grow rapidly but are usually responsive to chemotherapy. Surgical excision and occasionally radiation may also be employed. Overall survival rates range between 75% and 95%.[23] Peripheral blood stem transplantation may be an option for patients with advanced or recurrent disease.

Ovarian masses. Masses of ovarian origin, mostly ovarian cysts, are also encountered in childhood. Such cysts are occasionally associated with sexual precocity. Sonography can help establish the diagnosis. Management of ovarian cysts is based on the cyst size and composition and the patient's symptoms. Ovarian malignancies may be present in approximately 30% of cases, especially when solid or complex structures are noted within the mass.[24] The most common of these malignant tumors are teratomas. Ovarian malignancies in children are frequently found at an early stage and tend to respond favorably to chemotherapy.[13,14,25]

Hepatic masses. Abdominal masses arising from the liver are often malignant and include hepatoblastomas, embryonal sarcoma, hepatocellular carcinoma, or metastatic disease.[26] Less frequently occurring benign masses of hepatic vascular origin include hemangioendotheliomas and hamartomas.[27] Hepatoblastomas are the most common liver malignancies in this age-group, accounting for at least 75% of cases.[28] They may be congenital or familial and usually present with a rapidly enlarging abdomen in an otherwise asymptomatic child. They are derived from undifferentiated embryonal tissue and have been associated with prematurity, Beckwith-Wiedemann syndrome, and familial polyposis. Serum alpha-fetoprotein levels are almost always elevated, which aids in diagnosis and in monitoring response to therapy. Metastasis is present in 20% of cases at diagnosis with the lungs being the most common site, followed by the brain, bone, or bone marrow. Sonography is the initial imaging modality of choice, revealing a hyperechoic, solid intrahepatic mass. Sonography also permits assessment for vascular invasion. CT scanning may follow to determine disease extent and look for metastasis. Evaluation by this radiologic technique often demonstrates a delineated hypoattentuated mass compared with the surrounding normal tissue. A biopsy is usually recommended for diagnosis. Treatment consists of surgical resection and chemotherapy. Liver transplantation is occasionally performed in selected patients. Long-term survival varies depending on the stage of the disease and the success of initial surgical resection.[26,29]

Embryonal sarcoma is a mesenchymal malignancy that accounts for approximately 10% of all hepatic tumors in children.[29] Presentations include an abdominal mass, swelling, or pain. Serum alpha-fetoprotein levels are usually normal. Sonographic evaluation reveals a large predominantly solid mass with hypoechoic areas representing cystic areas. CT scan demonstrates a hypovascular low-attenuated mass with septa. Treatment with multiple modalities including adjunct chemotherapy, radiation, and surgical resection provides a 70% to 80% survival rate at 4 years.[26,29]

Nonhepatic gastrointestinal abdominal masses in children also include duplications, Meckel's diverticulum, fecal masses caused by severe constipation, and omental or mesenteric cysts.

Key Point

Abdominal masses in neonates are most often benign and of genitourinary origin. Malignant abdominal masses are more likely to be encountered beyond the neonatal age and mainly include neuroblastomas, Wilms' tumors, and lymphomas.

CASE PATIENT: DIAGNOSIS AND MANAGEMENT

The exact origin of the mass could not be determined radiologically. The etiology was not evident from other noninvasive tests. The preoperative diagnosis was possible gastrointestinal tract duplication. Jaundice was thought to be caused by common bile duct obstruction. At exploratory laparotomy, a large mass adherent to the posterior gastric wall was observed. A large blood clot was present within the mass. The mass was completely resected and did not communicate with the gastric lumen. A small perforation was found in the common bile duct, probably from the mass effect causing ischemia. The perforation was repaired. Histologic examination showed a gastric duplication cyst. The patient had an uncomplicated postoperative course. She has continued well over several months of follow-up.

CONCLUSION

The neonate, infant, or child with an abdominal mass needs rapid clinical evaluation. Age, history, and physical examination provide initial guideposts to diagnosis. Imaging studies, particularly sonography, may provide a specific diagnosis. If the initial evaluation indicates possible malignancy, more complex testing of blood, bone marrow, serum chemistries, and urine may be required. CT scanning may be also be useful in identifying the type and extent of abdominal masses in children. Outcome varies widely depending on the malignant or benign nature of the existing mass but is generally more favorable in neonates. **HP**

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