Chapter

24

Abdominal Masses

Neuroblastoma

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A 20-month-old girl presents with abdominal distension and a palpable, firm mass in her right abdomen. She is scheduled for open resection of the tumor. Vital signs include: Temp 37.6°C, BP 116/79, HR 88 bpm, SpO_2 100%. Heart and lung sounds are normal. Her CT scan is seen in Figure 24.1.

What Is the Differential Diagnosis for This Child's Abdominal Mass?

Malignancy is a serious concern with a new pediatric abdominal mass. Pediatric abdominal malignancies to consider include neuroblastoma, Wilms tumor (nephroblastoma), lymphoma, rhabdomyosarcoma, renal cell carcinoma, leiomyosarcoma, teratoma, and germ cell tumors. Other organ pathologies that may manifest as an abdominal mass include polycystic kidney disease, Meckel's diverticulum, and hepatic storage diseases. In a neonate one should also consider pyloric stenosis.

What Are the Clinical Characteristics of Neuroblastoma and Wilms Tumors?

Neuroblastoma is the most common extracranial pediatric tumor overall (with 65% of abdominal or adrenal origin), while Wilms tumor is the most common abdominal tumor in children. Both commonly present as palpable, painless abdominal masses without associated symptoms. Neuroblastomas may have associated hypertension, abdominal pain, constipation, or distension, while Wilms tumor often presents with hematuria and hypertension.

Children with localized disease may be asymptomatic other than a palpable mass, whereas those with advanced disease may appear cachectic and have constitutional symptoms (fever, weight loss). Large abdominal tumors may present with mass effect symptoms such as abdominal pain, fullness, bowel obstruction, or lower extremity edema. Mediastinal neuroblastoma can cause all of the most perilous symptoms of mediastinal masses including dyspnea from compression of the trachea or great vessels. Thoracic or cervical tumors may compress the sympathetic trunk causing Horner's syndrome, while larger thoracic tumors may cause superior vena cava syndrome. Metastatic disease can occur in the liver, bone, and skin.

What Is Neuroblastoma?

Neuroblastoma is a malignancy of the primordial neural crest cells that give rise to the sympathetic chains and adrenal glands. Their origin can stem from any part of the sympathetic nervous system, namely the adrenal glands and sympathetic chains of the abdomen, thorax, pelvis, and cervical area. As they are derived of neural crest they are capable of catecholamine secretion, although unlike pheochromocytoma they do not typically present primarily with symptoms of hypertension.

What Is the Incidence and Epidemiology of Neuroblastoma?

There are about 700 new cases of neuroblastoma annually in the United States. They account for around 10% of pediatric malignancies, though they are responsible for approximately 15% of pediatric cancer-related deaths. Most children are diagnosed before the age of 4, with a median age at diagnosis of 19 months.

How Is Neuroblastoma Diagnosed?

A definitive diagnosis of neuroblastoma requires one of the following:

(1) Tumor biopsy

(2) Evidence of bone metastasis seen on bone marrow aspirate with concomitant elevation of serum or

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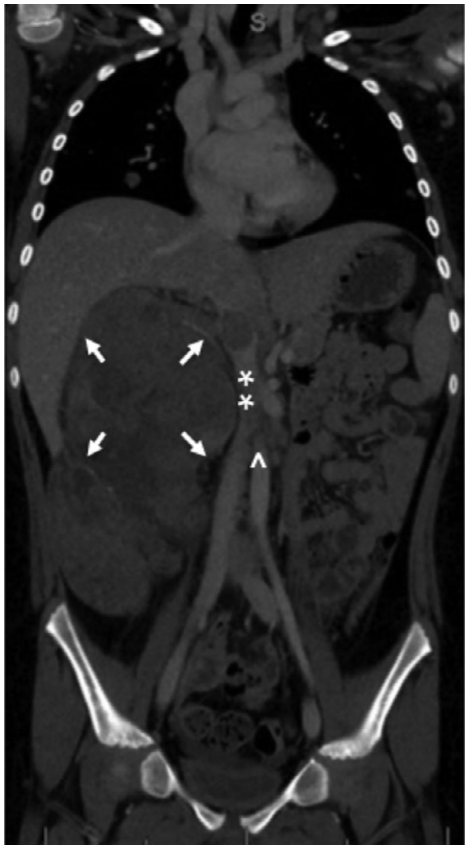

Figure 24.1 CT image of large circumferential neuroblastoma highlighted by arrows. Tumor size resulted in compression of the inferior vena cava (*) and aorta (^).

urine catecholamines/catecholamine metabolites (vanillylmandelic acid [VMA] and homovanillic acid [HVA]).

What Are the Currently Used Staging Systems for Neuroblastoma that Guide Treatment?

Ultrasound may be the initial study when an abdominal mass is discovered; however, CT or MRI is required for staging and evaluation of the extent of the mass if neuroblastoma is considered the likely diagnosis. A nuclear uptake scan is performed to evaluate for metastatic bone disease. Bone marrow biopsies should also be performed.

The International Neuroblastoma Risk Group Staging System (INRGSS) stages the patient based on radiology, clinical exam, and histology (Box 24.1). Surgical and histopathological factors are not part of the staging.

Box 24.1 International Neuroblastoma Risk Group Staging System (INRGSS)

Stage L1: Localized disease without image-defined risk factors

Stage L2: Localized disease with image-defined risk factors

Stage M: Distant metastatic disease (excluding stage MS)

Stage MS: Metastatic disease "special" in children younger than 18 months with metastases limited to liver, skin, and/or bone marrow

What Are the Commonly Used Chemotherapy Drugs and Relevant Side Effects That May Influence Your Anesthetic Management?

Chemotherapy can cause both acute side effects as well as lasting toxicities. Acute toxicities include nausea/vomiting, alopecia, myelosuppression and subsequent infection, and mucositis. Chemotherapeutic agents with well-known chronic side effects include doxorubicin (cardiotoxicity, arrhythmia), vincristine (peripheral neuropathy), and bleomycin (pulmonary fibrosis). The cardiovascular effects, specifically cardiomyopathy should be evaluated prior to anesthesia by echocardiography. Pulmonary involvement of chemotherapeutics, specifically in the presence of severe pulmonary fibrosis may warrant additional preoperative evaluation. In situations of severe end organ involvement, certain procedures may best be accomplished with regional anesthesia when appropriate (e.g., line placement).

What Major Anesthetic Concerns Do You Have for the Surgical Resection of the Tumor?

Hypertension: Hypertension may be present prior to the procedure due to catecholamine secretion from the tumor, but its absence does not necessarily mean that the tumor does not have catecholamine-secreting

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properties. If present preoperatively, blood pressure should be corrected with oral antihypertensive medications. As the patient may also be intravascularly volume depleted, intravenous hydration should also be used. During the procedure, hypertension may occur at any point, but has most commonly been reported at induction and with tumor manipulation. After removal, hypotension may be observed with the sudden absence of catecholamine secretion. Intravenous antihypertensive and pressor agents can be used intraoperatively, as indicated, and arterial and central venous catheters should be placed if hemodynamic lability is anticipated.

Bleeding and third space fluid losses: Neuroblastoma may surround or border large vessels. Large bore intravenous access should be obtained in the upper extremities given the potential for inferior vena cava (IVC) damage during resection or reduced IVC flow from tumor invasion. Blood products should be typed and cross-matched. Monitoring with invasive arterial blood pressure and central venous pressure should be considered.

Difficult ventilation: There may be respiratory compromise from the increased abdominal pressure caused by a large abdominal tumor. Ventilation may also become difficult during the case due to surgical retraction.

As the Surgeon Exposes and Manipulates the Tumor, the Patient Becomes Suddenly Extremely Hypertensive. What Are Your Next Steps?

Unlike pheochromocytoma, routine preoperative alpha blockade is not indicated for patients with neuroblastoma. However, intraoperative hypertension is not uncommon and may need to be treated. Case studies have reported successful use of beta blockers (labetalol, propranolol, esmolol), hydralazine, clevidipine, sodium nitroprusside, fenoldopam, and magnesium sulfate. Reported treatments for preoperative hypertension, if present, included phenoxybenzamine, enalapril, doxazosin, and labetalol.

What Other Anesthetic Strategies May Be Beneficial for This Child?

Resection of large abdominal masses often require large incisions. A caudal, lumbar, or thoracic epidural should be considered for intraoperative and postoperative pain control. Alternatively, bilateral paravertebral catheters may be considered.

Suggested Reading

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