

This is a text version of a podcast from PedsCases.com on “**Approach to Abdominal Mass Part 2.**” These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedcases.com/podcasts.

Approach to Abdominal Mass Part 2

Developed by Kieran Purich and Dr. Ioana Bratu for PedsCases.com.
Oct. 16, 2017

Introduction:

Welcome to the second PedsCases episode on an approach to pediatric abdominal mass. Once again, my name is Kieran Purich, and I am a medical student at the University of Alberta. I would like to take a moment to thank Dr. Ioana Bratu, a pediatric surgeon and Associate Professor at the Stollery Children’s Hospital at the University of Alberta in Edmonton, Canada for her help with the creation of this podcast.

In our last podcast we discussed the differential diagnosis, history and physical exam and initial investigations which should be considered when faced with a pediatric abdominal mass in practice.

Objectives

The objectives for the second episode of this two part series is primarily focused on two specific etiologies of abdominal mass – Wilms’ Tumor and Neuroblastoma. After listening to this podcast the listener should:

- 1) Review the pathology of Wilms’ Tumor and Neuroblastoma
- 2) Describe the typical presentations of Wilms’ Tumor and Neuroblastoma including key aspects on history and physical examination
- 3) Outline the diagnostic work up to confirm or rule out the presence of Wilms’ Tumor and Neuroblastoma
- 4) Discuss the management and prognosis of each tumor

Before discussing these specific causes of abdominal mass let’s review what we learned from Approach to the Abdominal Mass Part 1.

Take a moment to think about the differential diagnosis for an abdominal mass from an anatomic standpoint. Can you recall the organs/ organ systems that we discussed in our previous podcast? They were: renal, hepatobiliary, adrenal, gastrointestinal, pancreatic, spleen and genitourinary.

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In this podcast we will discuss two specific pediatric malignancies - one of renal origin known as Wilms' Tumor, as well as neuroblastoma – a tumor often associated with the adrenal gland.

At the end of this podcast we will also return to the case of Tyler – the 9 month old baby boy who presented with a large, non-tender abdominal mass in the right upper aspect of his abdomen and no other notable symptoms.

Now lets get started:

So what exactly is Wilms' tumor?

Wilms' tumor, also called nephroblastoma, is the most common renal malignancy in children making up 95% of renal tumors in individuals less than 18 years of age. The incidence is 8 Wilms' tumors per million children under 15 in the USA per year.

Two thirds of Wilms' tumors are diagnosed before 5 years of age. Risk factors include African-American race, family history of Wilms' tumor and the presence of a horseshoe kidney. About 10% of Wilms' Tumors are found in congenital syndromes including: WAGR syndrome which includes a combination of anomalies: Wilms' tumor, aniridia, GU anomalies & intellectual disability, as well as two other syndromes: Denys-Drash and Beckwith-Wiedemann syndrome.

Most Wilms' tumors are unilateral solitary lesions, however, bilateral involvement and multifocal loci within a single kidney do occur.

How would it present?

Most patients present with an abdominal/flank mass and no other significant signs or symptoms. Other patients may complain of abdominal pain (30-40%), demonstrate gross hematuria (12-25%), or present with fever and hypertension (25%).

Physical exam

On exam vital signs may show hypertension. On abdominal exam you will feel a firm, smooth mass that is eccentrically located and rarely crosses the midline. One must be very careful to not palpate too aggressively as the renal capsule can be ruptured resulting in tumor spillage. Ensure to look for superficial bruising as acquired Von Willebrand disease can occur in 4-8% of patients with Wilms' tumors at diagnosis.

Initial imaging - Abdominal ultrasound is the first test of choice. MRI or CT is used secondarily to look for bilateral lesions, metastases and to assess resectability. On imaging Wilms' tumor typically presents as a hypervascular, solid, heterogeneously echogenic solitary renal mass. Calcification is present in up to 20% of cases.

Labs include – CBC/D, urinalysis, creatinine, electrolytes, liver function tests, serum calcium (high in rhabdoid tumor or mesoblastic nephroma), coagulation studies including VW assay for Von Willebrand Disease.

Treatment - Patients with suspected Wilms' tumor should be referred to pediatric oncology and surgery for diagnoses by histology & management. Most Wilms' tumors are treated with surgery and chemotherapy and/or radiation.

Prognosis is determined by: tumor histology, tumor stage, molecular and genetic markers as well as age (<2 years is favorable). The 5-year survival rates with multimodal treatment currently approaches 90%.

Recurrence varies from 15 - 50% depending on histology at diagnosis and original staging of the patient.

It is important to follow up long-term with these patients, as there is risk of recurrence and late complications of chemotherapy and radiation therapy. There is a 1-15% risk of long-term renal failure over their lifetime. Most tertiary oncology centres will have a clinic dedicated to monitoring survivors of childhood cancer throughout their life specifically assessing for late effects of treatment and/or cancer recurrence.

Now lets move on to neuroblastoma.

What is neuroblastoma?

Neuroblastoma is a term that encompasses multiple tumors arising from neural crest cells destined to be sympathetic ganglion cells. These tumors have potential to secrete catecholamines and can appear in various locations. Tumors within this group can vary from benign to aggressive disease.

Neuroblastoma is the most common extracranial solid tumor of childhood, and is one of the most common childhood malignancies. The median age at diagnosis is 19 months.

Neuroblastoma is slightly more common in boys, and in some cases is speculated to be associated with maternal/fetal factors such as: opiate consumption in pregnancy, folate deficiency, gestational diabetes and congenital abnormalities. It is more common in patients with Turner syndrome, central hypoventilation, Hirschprung disease and NF type 1.

Clinical Presentation

Neuroblastomas can present in numerous ways. The adrenal gland is the most common site for occurrence (40%). Followed by the abdomen (25%) and the thorax (15%). These tumors can also arise from cervical and pelvic ganglia. For our podcast we will focus on those, which present as an abdominal mass

Parents may comment on a newfound fixed, firm abdominal mass. (2/3 of primary neuroblastomas arise in the abdomen) which may be accompanied by abdominal pain, constipation, back pain, scoliosis, bladder dysfunction, fever, weight loss, and bone pain. Far less often, patients may demonstrate flushing and sweating due to the tumor's secretion of catecholamines.

Metastasis occurs by hematogenous and lymphatic spread, often affecting bone, bone marrow, skin and the liver. This may cause pain, blood count abnormalities, fever, bone pain, periorbital ecchymosis, ptosis, proptosis, a diffuse bluish red non-tender subcutaneous rash termed "blueberry muffin baby", heterochromia iridis (different colors of iris or portions of iris), nasal obstruction and/or limping.

Physical exam - On physical exam there may be hypertension or tachycardia noted due to catecholamines secreted by some of the tumors. It is important to determine the masses location and characteristics and to feel for associated lymphadenopathy. A head to toe physical exam should be completed to look for any signs of metastases as was mentioned previously.

Imaging - As with most abdominal masses the initial study should be an ultrasound and if the results are concerning for malignancy an urgent consultation with a pediatric oncologist and or a pediatric surgeon is warranted. CXR may also be completed to ensure there is no thoracic disease.

In order to evaluate lymph node involvement, presence of metastases, and resectability a CT or MRI will be ordered by the oncologist or surgeon. These modalities will generally show a heterogenous mass with anechoic areas due to necrosis or hemorrhage. Calcifications are present in about 85% of neuroblastomas on CT.

CT and MRI allow for accurate assessment of the size, location and vascular encasement, which are critical to determine resectability. A nuclear medicine test called I123-MIBG scan is also used to delineate the lesion as a neuroblastoma and look for metastasis.

Other investigations – Standard laboratory workup, which includes CBC/D, creatinine, and electrolytes, should be completed. In addition, neuroblastomas have the ability to secrete the catecholamines norepinephrine, epinephrine and dopamine, which degrade into end products Vanillylmandelic Acid (VMA) and Homovanillic acid (HVA) These can be detected in the serum and urine of 70-90% of patients with neuroblastoma.

Otherwise the tests to consider are similar to our previous abdominal mass presentations.

Diagnostic evaluation for a neuroblastoma requires either:

1. Unequivocal histologic diagnosis.

2. Evidence of metastases to bone marrow on aspirate with concomitant elevation of serum or urinary catecholamines and metabolites.

Treatment- Low risk tumors may be treatable by surgical resection alone. Beyond this treatment becomes more complex and is made up of different modalities which may include surgery, chemotherapy and stem cell transplantation.

Prognosis- since neuroblastoma is a heterogenous entity it is difficult to give a generalized prognosis. They are staged using the International neuroblastoma staging system (INSS) and this is a large predictor of prognosis. In general prognosis is better in younger patients with localized disease. As children age the 5-year survival steadily decreases (83% at 1 year and 40 % at 5-9 years).

As mentioned previously, many of these treatments have long-term effects. For neuroblastoma these include: renal issues, poor growth, thyroid dysfunction, infertility and secondary malignancies. Long-term followup is recommended at a Childhood Cancer Survivor Program

Case Summary - Now lets return to Tyler – A 9 month old with the new found 5-6 cm right upper quadrant abdominal mass. The mass is smooth, non-tender, firm, fixed and does not cross the midline. You are concerned and believe that this tumor could be of renal or hepatobilliary origin. You send the patient for blood work including CBC/D, urinalysis, creatinine, electrolytes, liver function tests and serum calcium as well as an ultrasound of the abdomen. The ultrasound shows a single mass 6 cm x 5 cm x 3 cm originating from the right kidney. You are worried about a Wilms' tumor and make a phone call to pediatric surgery for consultation.

The pediatric surgeon arranges for more testing and consults with a pediatric oncologist and pediatric radiologist to optimize management

The MRI shows a solitary renal mass with heterogenous echogeneity. The surgeon deems it is resectable. The child undergoes a nephrectomy and histology confirms a Wilms' tumor. Because of staging he undergoes additional treatment after surgery that uses adjuvant chemotherapy. After a tumultuous time for the family he recovers well and returns to your office regularly for follow-up in concert with the pediatric oncology team.

Conclusion:

Let's finish with a few key take-away points

1. Pay close attention to the differential diagnosis and remember to consider all causes of abdominal mass. Malignant causes are rare but are essential to include on all differentials. Remember to think about Wilms' Tumor & Neuroblastoma.

2. Wilms' Tumor is the most common pediatric renal malignancy in children and may present with hematuria, abdominal pain and hypertension. However, often it presents as an isolated abdominal mass with no other symptoms.

3. Neuroblastoma is the most common extracranial solid tumor malignancy of childhood and arises from neural crest cells. It can occur in various locations; however, the most common location is the abdomen. These tumors may secrete catecholamines, which can lead to hypertension, tachycardia, flushing and sweating.

4. Consult a pediatric oncologist promptly if you detect a concerning abdominal mass in a pediatric patient.

That concludes our series on pediatric abdominal mass. Thank you for listening.

Resources

1. Chu, C., Rasalkar, D., Hu, Y, Cheng, F., Li, C., Chu, W. Clinical presentations and imaging findings of neuroblastoma beyond abdominal mass and a review of imaging algorithm. 2011 The British Institute of Radiology 84 P. 81-91.
2. Davidoff, A. 2008. Pediatric Abdominal Masses. Common Surgical Diseases, DOI: 10.1007/978-0-387-75246-4_63
3. Dingeldein, M. 2015. Pediatric Abdominal Masses. Common surgical diseases. DOI: 10.1007/978-1-4939-1565-1_72
4. Kaste, S., McCarville, B., 2008. Imaging Pediatric Abdominal Tumors. Seminars in Roentgenology. Doi:10.1053/j.ro.2007.08.007
5. Kocaoglu, M., et al., 2010., Pediatric abdominal masses: diagnostic accuracy of diffusion weighted MRI. Magnetic resonance Imaging **28** p. 629-636.
6. Postisek, N. and Antoon, J. 2017. Abdominal Masses. Pediatrics in review **38** p.101-103
7. Up to Date: Clinical assessment of child with suspected cancer
8. Up to Date: Clinical presentation, diagnosis and staging evaluation of neuroblastoma
9. Up to Date: Epidemiology, pathogenesis, and pathology of neuroblastoma
10. Up to Date: Treatment and prognosis of neuroblastoma
11. Up to Date: Presentation, diagnosis and staging of Wilms' tumor
Up to Date: Treatment and prognosis of Wilms' tumor.

