DCMC Emergency Department Radiology Case of the Month

These cases have been removed of identifying information. These cases are intended for peer review and educational purposes only.

Welcome to the DCMC Emergency Department Radiology Case of the Month!

In conjunction with our pediatric radiology specialists from ARA, we hope you enjoy these monthly radiological highlights from the case files of the Emergency Department at DCMC. These cases are meant to highlight important chief complaints, cases, and radiology findings that we all encounter every day.

If you enjoy these reviews we invite you to check out Pediatric Emergency Medicine Fellowship Radiology Rounds, which are offered quarterly and are held with the outstanding support of the pediatric radiology specialists at Austin Radiologic Association.

If you have any questions of feedback regarding the Case of the Month format, fell free to email Robert Vezzetti, MD, at rmvezzetti@seton.org.

This Month: Abdominal pain AFTER an appendectomy. This, in and of itself, is not uncommon. In this child’s case, though, there’s more than meets the eye.

PEM Fellowship Conference Schedule May 2016

3rd Special Lecture: Maintaining an Academic Presence….Dr Allen
4th 9:15 - Cardiovascular Infections…………………Drs Allen and Berg
10:15 - Board Review Potpourri………………………Dr Pittman
11:15 - TBD
11th 9:15 - US PIV and Central Lines………………….Dr Boeck
10:15 - Pharyngitis/Oral Disease………………………Dr Schwartz
11:15 - TBD
18th 9:15 - QI Measurements……………………….Dr Iyer
10:15 - Board Review: Cardiology…………………..Dr Earp
11:15 - TBD
12:15 - Research Update……………………………..Dr Wilkinson
31st - Journal club/literature review………..Drs Gregg/Harrison/Freisen

Lectures are held in the DCMC Command Rooms 3 and 4. Simulations are held at the CEC Breckenridge. All are welcome to attend!

Schedule subject to change.
May is comes from the Latin ‘Maius’, is named after Maia, a Roman goddess of growth. Greek Mythology claims Maia as well, as the mother of Hermes and the daughter of Atlas. In Greek, μαῖα (maia) is an honorific term for older women related to μήτηρ (mētēr) ‘mother’.

**Case History:** It appears that a Spring pattern is starting to settle in the ED, as the volume of patients, while not drastically down, has diminished somewhat. The usual Spring time cases are coming in: mostly orthopedic issues (fractures, sprains, etc), the Spring time cold, allergies, and abdominal pain. Yep, lots of abdominal pain is seen during this time of year, but one chart catches your eye. It’s a 9 year old female who is here with 4 weeks of abdominal pain. The pain is described as initially right sided, but now seems to be more epigastric and right upper quadrant. You note that she had an appendectomy 5 weeks ago. She denies fevers, weight loss, vomiting, or diarrhea. The child denies dysuria or hematuria. She also denies any trauma. She has been intermittently nauseated since the appendectomy, but has been eating fairly well. Eating and drinking, by the way, does not change the quality or quantity of the pain. The child states that the pain does not radiate anywhere and she describes it as sometimes sharp, sometimes dull, but persistent. In fact, over the past 1-2 weeks, her symptoms has become quite persistent, which led to this Emergency Department visit. She has been to her pediatrician on several occasions and was diagnosed with GERD. The physician prescribed dietary changes and, should this not help with the pain, was planning on trying Zantac.

On exam, the child is afebrile and has age-appropriate vital signs (afebrile, not tachycardic, normal blood pressure). She is no obvious distress and, in fact, smiles at you. Her exam is unremarkable with the exception of epigastric tenderness to palpation, along with minimal right upper quadrant tenderness. Her abdomen seems slightly full but is not tender anywhere else. You do not appreciate guarding or rebound. There are no obvious masses. Her appendectomy sites look well-healed and clearly not infected. She has no back tenderness.

What is going on with this child? Is the pain related to her previous appendectomy? Not likely, as this was done one month ago and a related complication would be extremely rare, and, frankly, very odd. What could be causing this epigastric pain? Is imaging needed? Probably so, given the history and exam, but which modality? Plain film? Ultrasound? CT? One thing is for sure….it’s not her appendix!

In 1886 Reginald Fitz provided the first accurate description of appendicitis. In 1887, Thomas Morton performed the first successful appendectomy in the US. 1889 saw Nicholas Senn diagnose, perform an appendectomy, and have the patient recover successfully. Later that year, Charles McBurney described clinical findings of appendicitis, including a point of maximal tenderness, still known today as McBurney’s point.

Laparoscopic appendectomy is the preferred technique when surgically treating appendicitis. There are numerous advantages, including faster recovery, reduced pain, and minimal scaring. It also has the advantage of allowing a surgeon to view the entire abdomen.

Complications from an appendectomy are few but usually are infectious. A perforated appendix will increase the chance of post-operative infectious complications. Abscess formation is also a common complication, but the rate of abscess formation in laparoscopic appendectomy versus open appendectomy is not higher.

Single site laparoscopic appendectomy is often used, but sometimes there are up to three very small incisions, depending on the patient as well as the surgeon.

In 1901 the Prince of Wales, Albert Edward, underwent an emergency appendectomy, just two weeks before his scheduled coronation as King Edward VII.
Epigastric and Right Upper Quadrant Pain: Considerations

Abdominal pain is, as we all know well, not uncommon in children. Most of these cases have benign causes of the pain. Acute causes of epigastric pain include gastritis, GERD, and pancreatitis. Chronic causes of pain often do not have an identifiable cause, but include GERD, pancreatitis, and gallbladder issues. Does this child meet criteria for chronic pain? The AAP published guidelines in 2005 regarding the definition of chronic abdominal pain, as well as various causes, including functional abdominal pain. This child is pretty specific about the pain. This raises concern for either pancreatitis or gallbladder issues.

So, you decide to obtain some lab work, given the exam. Specific tenderness to the epigastric area is suggestive of pancreatitis, although the right upper quadrant tenderness certainly could be gallbladder related. That would be strange, as the child does not fit a typical body habits for gallbladder illness and the pain is not related to eating/foods. Could a plain film help? You get one, while waiting for the lab work to result.

The film is seen on the right. Not very remarkable. There is somewhat prominent stool. Could this all be constipation? There is not sign of free air or bowel obstruction. There are no obvious masses. Now what?

While you are contemplating the KUB, the child’s lab work is resulted. Her CBC shows a WBC of 6.2 (46S 34L), H&H of 9.8/29.8, platelets of 245. Hmmm..a little anemic. Her CMP looks normal. The Amylase and Lipase, though, are elevated at 137 and 181 respectively. Those are slightly elevated. Could this be pancreatitis? From what? Is it related to the appendectomy? These values a slightly elevated, but does this child need additional imaging?

Pancreatitis In Children - Pediatric pancreatitis is not common, but can be associated with significant morbidity and mortality. It can be classified as acute, chronic, necrotic, or hemorrhagic, depending on the etiology. Predominant causes of pancreatitis in children include abdominal trauma (majority of cases), tumors, pancreatobiliary system abnormalities, drugs/toxins, viral infections (CMV, mumps, rubella, coxsackie B), metabolic disorders, and hereditary conditions (caused by alterations in chromosome 7, including those children with cystic fibrosis). Depending on the etiology, most children with pancreatitis do well.

There is certainly a large amount of stool demonstrated on this film (red arrows). Perhaps discharge with Miralax or an enema is in order. Contestation certainly can cause abdominal discomfort, but this has been going on for the past month. It would be very unusual, with a film like this, to have these symptoms (particularly epigastric and RUQ pain) for this length of time.

The first known observance of Mother’s Day in the U.S. occurred in Albion, Michigan, on May 13, 1877, the second Sunday of the month.
Pancreatic Pseudocyst - Sometimes pancreatitis is complicated by a pseudocyst, which is the formation of a fibrous-walled cavity that is filled with pancreatic enzymes. This can be seen following abdominal trauma and in children with ventriculoperitoneal shunts, as well as in children with both acute and chronic pancreatitis. Pseudocysts are managed both medically and surgically. Pseudocysts from trauma are typically surgically managed, as are those associated with VP shunts.

Here is a nice CT example of a CT demonstrating both the pancreas and an adjacent pancreatic pseudocyst. These are usually pretty well-defined.

OK, back to our patient. With the child’s exam and the elevated amylase/lipase, you decide to obtain an ultrasound. A selected image is noted to the right. Try to find the pancreas! It’s a little hard, because there is a large mass that is obscuring our view of a normal pancreas (red arrow). Look below for a comparison of what a pancreas should look like in ultrasound. This finding is either a mass or possibly a very large thrombus of the superior mesenteric artery. Either way, this is a very concerning finding. More imaging?
So, in light of the ultrasound findings, a contrast CT scan of the abdomen is obtained. Selected images are seen to the left. The most striking finding is the mass, which measured 3.2 x 4.1 x 3.9 cm (red arrow). There is also occlusion of the splenic vein and possible thrombus formation in the superior mesenteric vein (blue arrow), although this is only limitedly seen.

These CT findings are very concerning for a pancreatic tumor. This child needs admission and consultation with Pediatric Hematology-Oncology. She will also need a Pediatric Surgery Consultation, both to explore surgical treatment options and, more likely, the placement of a Port-a-cath for chemo, etc. Additional labs are obtained: Uric Acid, LDH, and Alphafetoprotein (AFP). These are resulted; all are normal except for the AFP, which is very elevated at 16,000.

Alphafetoprotein is elevated in yolk sac tumors, embryonal cell carcinomas, neuroblastomas, mixed germ cell tumors, and some teratomas. Levels will often correlate with tumor size. Levels can be monitored during and after treatment as one of many ways to gauge a patient's response.

Pancreatic tumors are very rare in children, accounting for approximately 0.2% pf pediatric malignant deaths. Neoplasms can be divided into epithelial and non epithelial types. These are further divided into exocrine and endocrine origin. Exocrine tumors are usually pancreatoblastoma, which is seen almost exclusively in children. This tumor is the most common pancreatic tumor in children. There had been a history of difficulty in nomenclature and staging, such that there is really no consensus regarding staging of pancreatoblastomas in children. As a general rule, clinical outcome is better versus adults.
The child was admitted and appropriate subspecialty consultation was obtained. Due to the results from the bloodwork as well as imaging tests, the diagnosis of pancreatoblastoma was made. Additionally, the child underwent biopsy of the lesion. The results of this confirmed the diagnosis. The lesion was positive for AFP and KerAE1, both markers consistent with embryonal tumors, as well as Keratin 1/3 and OCT 3/4. Flow cytometry was performed and this demonstrated no abnormal lymphoid proliferation.

Pancreatoblastoma usually occurs in the first decade of life. Asymptomatic abdominal masses are reported, but so are nonspecific symptoms (abdominal pain, malaise, weight loss, vomiting, diarrhea).

Imaging Features:
They are typically solitary masses and well-defined. Approximately half will arise from the head of the pancreas. The tumor is usually large when discovered and often will not invade surrounding tissue (radiographically).

On ultrasound, these lesions appear as well-circumscribed heterogeneous masses. They have both solid and cystic components.

On CT, these lesions also appear heterogeneous which may reflect internal cystic areas. The septa may enhance and the lesion appears multiloculated.

MRI has been used to investigate pancreatoblastoma, but its use is limited and there are few descriptions in the literature. On T1 images, there is often low signal intensity (demonstrating areas of necrosis) and high signal intensity on T2 weighted images.

Adjacent organ involvement as well as distal metastases may occur. This is not common though. When metastases occurs, it typically involves the liver and abdominal lymph nodes.

One should also consider a differential of neuroblastoma, Wilms tumor, and hepatoblastoma in children with pancreatic tumors, especially if it is not clear that the tumor is arising from the pancreas. Lymphomas may also present with pancreatic lesions.

Cinco de Mayo is a regional holiday in Mexico, primarily celebrated in the state of Puebla, with some limited recognition in other parts of Mexico. The holiday commemorates the Mexican army’s unlikely defeat of French forces at the Battle of Puebla on May 5, 1862, under the leadership of Mexican General Ignacio Sequin.
However, a common misconception in the United States is that Cinco de Mayo is Mexico’s Independence Day. Mexico’s Independence Day is actually September 16.

Case Resolution: This child was admitted to the hospital and both Pediatric Surgery and Pediatric Hematology-Oncology were consulted. Initially an MRI was planned, but the ultrasound and CT images provided enough detail to prompt a biopsy of the mass. The results were indeed consistent with pancreatoblastoma. Given the size and location, the mass is not surgically resectable at this time. Instead, a PICC line was placed and chemotherapy was planned with Cisplatin and Doxorubicin. After chemotherapy, surgical options will be re-explored. Should the mass be unresponsive, other chemotherapeutic or radiation therapy options will be explored.

Teaching Points
1. Abdominal pain in children is extremely common and often the cause is readily apparent. There are times, though, that the etiology is not obvious, prompting workup. The workup for abdominal pain should be directed by history and physical examination. This will often obviate the need for further testing.
2. Persistent or worsening abdominal pain, especially accompanied by fevers, weight loss, bloody stools, abdominal distention, persistent vomiting, or other systemic signs of illness should be evaluated.
3. Epigastric pain in pediatrics can be the result of acute processes, such as gastritis, streptococcal throat infections, pneumonia, trauma, hepatobiliary issues (acute cholecystitis or hepatitis for example) issues, or acute pancreatitis. More chronic causes include gastroesophageal reflux disease, pancreatitis, hepatobiliary issues (chronic gallbladder disease or chronic hepatitis), and ulcers.
4. Laboratory evaluation of epigastric pain includes CBC, CMP, and Lipase. Some studies have concluded that Lipase is more sensitive and specific while others have advocated improved detection of pancreatic disease by utilizing both assays. Amylase does appear to rise faster in the setting of trauma.
5. Imaging options for suspected pancreatic or hepatobiliary disease typically include ultrasound (as a first line test), IV contrast CT scan, and MRI. CT scanning is excellent in the setting of trauma and as a quick, first line test when evaluating masses detected by ultrasound. MRI is often reserved as a more definitive option to evaluate anatomy but seldom used as a first line test.
6. Pancreatoblastoma is a very rare cause of pediatric tumors, but the most commonly encounter pancreatic neoplasm in children. Treatment is typically total resection, but chemotherapy and radiation therapy can play a role as well.
7. Most children do well with complete resection. Metastasis is indicative of worse prognosis.

References