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 check out Pediatric Emergency Medicine Fellowship Radiology Rounds, which are currently offered quarterly and are held with the outstanding support of the pediatric radiology specialists at Austin Radiologic Association.

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

**This Month: Let’s Celebrate St Patrick’s Day with the following case. It illustrates how careful one has to be when evaluating a child who appears to have a viral infection that has gone bad. This case reminds me to keep an open mind during respiratory season in that all that coughs is not necessarily a URI...although most of it is!**

**Is RSV season over yet?**

St Patrick’s Day Fact: According to Irish legend, Saint Patrick wasn’t originally called Patrick. His birth name was Maewyn Succat, but he changed his name to Patricius after becoming a priest. He also wasn’t Irish...he was born in England but was captured and sent to Ireland as a slave before escaping back to England.

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**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.

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**PEM Fellow Conference Schedule March 2015**

4th  9AM-10AM: GU Trauma/Scrotal Disorders..Guest Urologist
10AM-12PM: Simulation: Cardiac Emergencies..Sim Faculty

6th  7:30AM-8:30AM: Grand Rounds................................Dr Pines

7th-9th: PEM Fellow’s Conference/Meeting

10th Special Lecture: Academic Medicine/Job Finding

11th  9AM-10AM: Immunocompromised Pt.......................Dr Allen
10AM-11AM: Rheumatologic Emergencies....Guest Rheum
11AM-12PM: M&M............................Drs Kienstra and Yanger
12PM-1PM: Posters and Platforms............................Dr Allen

18th SPRING BREAK NO CONFERENCE

25th 9AM-11AM: Board Review (Cardiopulm)..........Dr Solomon
11AM-12PM: ED Staff Meeting

Guest Lecturer:  Jesse Pines, MD, MBA, MSCE
    Director, Office for Clinical Practice Innovation
    Professor of Emergency Medicine and Health Policy
    The George Washington University

All Lectures are at Dell Children’s Medical Center in Command Rooms 3&4, unless otherwise specified.

Simulations are held at the CEC at University Medical Center - Brackenridge.

*Schedule subject to change.*

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*The Leprechaun is said to be a solitary creature, whose principal occupation is making and mending shoes, and who enjoys practical jokes.*
To celebrate St Patrick’s Day, Chicago has a spectacle all its own. The city has been celebrating Saint Patrick by dumping green dye into the Chicago River since 1962. It takes 40 tons of dye to get the river to a suitably festive shade!

Wilhelm Conrad Röntgen was a German physicist, who, on 8 November 1895, produced and detected electromagnetic radiation in a wavelength range known as X-rays or Röntgen rays, an achievement that earned him the first Nobel Prize in Physics in 1901. In honor of his accomplishments, in 2004 the International Union of Pure and Applied Chemistry (IUPAC) named element 111, roentgenium, a radioactive element with multiple unstable isotopes, after him.

Case History: It’s the middle of a busy shift in the Pediatric Emergency Department. The rooms (and waiting rooms) are chock full of febrile, coughing, sneezing infants and children and there appears to be no end in sight. As you are explaining bronchiolitis for what is probably the thousandth time during the shift to some anxious parents, an infant is quickly brought back. The nurse comments that the child is “a pretty bad bronchiolitic” and she has already summoned the respiratory therapist to the room. The Fellow who is working with you tonight goes in to see the child, freeing you up to conclude your bronchiolitis lecture.

The Fellow comes back to you and reports that the child has had cough for the past several weeks, but this has gotten worse for the past week and for the past three days has had difficulty breathing as well as sleeping at night; his mother reports he appears to be uncomfortable when laying down, which she had not noticed before. There is no report of fever, vomiting, sick contacts, or travel. The child’s past history is not impressive either as he has been healthy all of his life, according to his mother, feeding and growing well.

On exam, this infant certainly appears uncomfortable and is in obvious respiratory distress. As the child is being hooked up and “vitalized”, you give him a quick once over and notice that in addition to tachypnea and moderate intercostal retractions, he is grunting. He does not have nasal congestion and, strangely, he is afebrile. His pulmonary examination on auscultation is even more strange, as he has no evidence of crackles, rhonchi, or wheezing. What kind of bronchiolitic is this? You do notice, though, that he has diminished breath sounds on the left. The nurse also informs you that a pulse oximetry reading is now obtainable and it is reading 85%. You start some oxygen, with little effect on his vitals or respiratory status.

The respiratory therapist shows up and proceeds to give a 10 mg Albuterol neb (why? you wonder). But since it can’t hurt, you decide to see if this helps. It doesn’t. In the meantime, the infant starts to grunt more, become more tachypenic, and overall looks worse. You wonder if you need to intubate this little guy. As you get ready for that, you consider trying high flow oxygen, since a face mask and nasal cannula are not helping. Could this be cardiac? What about a foreign body? Maybe a pulmonary process. You try some high flow oxygen and the child seems to improve enough for you to consider your next step. Your Fellow orders some bloodwork and you both start to consider imaging. Guess a chest xray is in order....
Given this infant’s clinical exam, which is quite concerning, you are able to obtain a chest xray. The films are seen here on the left. Notice anything concerning?

The PA view of this child’s chest demonstrates opacification of the left hemithorax (red arrows), leading to mediastinal shift to the right (blue arrows). What is visualized of the cardiac silhouette appears normal (yellow arrows). The purple arrow shows oxygen tubing.

The lateral view of the 2 view xray series is equally impressive. What is notable is that the heart size does indeed seem normal (yellow arrows). Again we see the opacity (red arrows).

This is obviously a very concerning xray. What is causing this? Could it be infectious, such as pneumonia or even an empyema. Possibly, but the clinical picture does not fit and there are no signs of effusion on the chest xray. Could there be a cardiac cause? Also a possibility, but the heart size looks normal and the right hemithorax looks fine as well. There are no signs of fluid overload on the films, which would lead one away from a heart failure issue. A foreign body? Maybe, but it would be quite unusual to have a film like this if that were the case. One would expect hyperinflation and most (but not all) foreign bodies tend to involve the right mainstem bronchus. Speaking of which, you can see the trachea on the films being pushed over. One of the top concerns would be some kind of mass; this seems to fit the picture. There is unilateral involvement causing the mediastinal shift. No fever history and no history concerning for a cardiac issue.


Corned beef and cabbage, a traditional Saint Patrick’s Day staple, doesn’t have anything to do with the grain corn. Instead, it’s a nod to the large grains of salt that were historically used to cure meats, which were also known as “corns.”
The chest xray and clinical picture are concerning for a chest mass and further imaging is needed. Typically, CT is next diagnostic study of choice. It is readily available and can provide important anatomic details about the extent of the mass. An important consideration when obtaining further imaging in patients with chest masses is whether or not sedation will be needed. This is critical, as sedating a child with a chest mass like this can lead to respiratory arrest; the mass, after all, is occupying the entire left side of the chest and causing a mediastinal shift. Airway protection and control is a paramount issue, as sedating a child with such a mass will directly impact their airway. If a study is time consuming, like an MRI, then intubation is advisable. The nice thing about a CT is that it is quick as well, making intubation less likely to be needed during this quick study.

This child underwent a CT scan with and without contrast (for a discussion about when and why to use IV contrast with CT scanning, please check out the February 2015 issue of the Newsletter, it’s all there, folks).

There are some obvious very concerning and very abnormal findings on this scan. There is a left chest mass (Red Arrows) with associated effusion (Blue Arrows). The effusion could be hemorrhagic products, which can be seen commonly with chest masses. This is causing atelectasis of the left lung. There is displacement of the heart and mediastinal structures (not seen entirely on these images, but the Yellow Arrow shows this) by the mass, which measures 6 x 6 x 5.7 cm—pretty big chest mass. The epicenter of the mass appears to be the posterior mediastinum. There is also widening of the neural foramen at T2 (Purple Arrow), suggesting tumor extension into the transverse process of T2.

That’s a lot, people. It is clear this child has a chest mass, originating in the posterior mediastinum. Time to get the books (or Internet) out and determine the differential diagnosis for posterior mediastinal masses. On appearance, this mass looks consistent with a germ cell tumor, but the extension into the intraspinal space point to a ganglion cell tumor, such as neuroblastoma or ganglioblastoma. Next steps?
Further imaging is needed. In the meantime, what can cause a posterior chest mass in children? How does one know the mass is posterior in the first place?

The boundaries of the mediastinum include the superior border, which is the thoracic inlet, the inferior border, which is the diaphragm, the anterior border, which is the sternum, and the posterior border, which is the vertebra. The mediastinum is divided into anterior, middle, and posterior components.

How does one know clinically if there is a mediastinal mass? Most are symptomatic to one degree or another. Symptoms, however, can vary and are often quite nonspecific, such as fever, cough, dyspnea. One red flag is recurrent pulmonary infections or what appears to be persistent atelectasis on a chest xray. Dysphagia can result from esophageal compression and dyspnea can result from bronchial compression. Compression of the laryngeal nerve can result in hoarseness.

Superior Vena Cava Syndrome is a specific clinical finding that may occur with mediastinal tumors, due to compression of the superior vena cava. Clinical hints include dyspnea, headache, facial edema (worse in the morning but resolves/improves by the end of the day), upper limb edema, edema of the neck (collar of Stokes), and Pemberton’s sign.

Horner’s Syndrome (remember this one from med school days?) can result from damage to the sympathetic nerve chain of the affected portion of the face.

Irish law, from 1903 to 1970, declared St. Patrick’s Day a religious observance for the entire country meaning that all pubs were shut down for the day. That meant no beer, not even the green kind, for public celebrants. The law was overturned in 1970.

Hugh Pemberton was an English physician who wrote extensively on diabetes, thyrotoxicosis, and peripheral vascular disease. He is responsible for discovering Pemberton’s Sign (aptly named, I think).
This child was admitted and MRI studies were ordered, including the chest, cervical spine, thoracic spine, lumbar spine, and brain. When a lesion is identified, whole studies of the brain, etc are undertaken to look for either other lesions (sometimes the primary lesion is found this way), or secondary metastises.

Pediatric surgery, Pediatric critical care, and Pediatric hematology-oncology were all consulted. The child was admitted to the PICU.

A Ct of the abdomen/pelvis was also obtained. Again, this is a quick way to look for other lesions/masses and get a sense about the extent of the primary disease.

The images show ascites (violet arrow) and portions of the chest mass/atelectasis (purple arrow), but are otherwise negative.

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Here is the MRI. The chest mass is again seen quite easily. The MRI of the thoracic spine is interesting, as well. At T2 the posterior elements are abnormal (wide and enhancing yellow arrow), suggesting extension of the tumor. However, there is no cord compression or involvement.

The next step was to identify what this mass is and then develop a treatment plan. The child was taken to surgery and the mass was biopsied. The pathology came back suggestive of Ewing’s Sarcoma or a Primitive Neuroectodermal Tumor (PNET). The definitive diagnosis was made by FISH study, which confirmed that the tumor was, indeed, Ewing’s Sarcoma.

According to legend, St. Patrick drove all the snakes, or in some translations, “toads,” out of Ireland. In reality, this probably did not occur, as there is no evidence that snakes have ever existed in Ireland, the climate being too cool for them to thrive. Despite that, scholars suggest that the term “snakes” may be figurative and refer to pagan religious beliefs and practices.
FISH (Fluorescence In Situ Hybridization) is a technique developed in the 1980’s. This technique uses fluorescent probes that bind to specific parts of a chromosome to which the probe shows sequence complimentarity. These are highly specific DNA or RNA sequences. FISH is not just limited to identification of tumors; it can also be used to identify pathogens and in genetic counseling.

This child was ultimately diagnosed with Ewing’s Sarcoma, confirmed by FISH. This is a malignant tumor that arises out of bones or soft tissue. Common bony sites for Ewing’s include the humerus, ribs, pelvis and femur.

The genetic location responsible for the majority of these tumors is often shared with primitive neuroectodermal tumors (PNET) as well, and sometimes these lesions are collectively known as Ewing Family of Tumors, but they are distinct disease processes. Ewing’s is most commonly seen in teenagers. The vast majority of Ewing’s sarcomas are caused by genetic translocations between chromosomes 11 and 22.

On imaging, these tumors can appear to affect the bony cortex or even appear as soft tissue masses without bony destruction and radiographs frequently will not show any cortical destruction.

1/3 of Ewing’s Sarcomas are metastatic on discovery and symptoms are often vague, including fevers, pain at involved bone sites, anemia, fatigue, and increased inflammatory markers.

Ewing’s sarcoma, on staining, will have clear cytoplasm and be PAS positive. Imaging studies to define the extent of the disease include x-rays, CT scanning, MRI, and bone scintigraphy.

Treatment options for Ewing’s sarcoma include multidrug chemotherapy, local disease control (resection), and sometimes radiation therapy. Typically after chemotherapy, the tumor then is resected, although sometimes radiation is used in conjunction with resection. Treatment time lengths vary depending on the stage of the disease as well as the response of the patient. Progress/response is followed clinically, but also with bone scans and CT/MRI.

**Case Resolution:** This child had extensive multi-specialist involvement. A treatment course of chemotherapy was prescribed after which he was a candidate for surgical resection. This proved to be difficult, however, due to the involvement of the thoracic vertebrae, requiring resection of portions of the involved bone. Pediatric Neurosurgery determined that the cord was not involved, which was good news indeed. The child had local tumor resection done in Boston, with collaboration among specialists at DCMC and Texas Children’s Hospital in Houston. As of this writing, the child continues to be followed clinically and radiographically, primarily by Pediatric Hematology-Oncology at Dell Children’s; there has been no recurrence of disease and he appears to be doing well. Luck of the Irish indeed!

**Teaching Points:**

1. **All that coughs during the cold and flu season is not necessarily viral, although most of it is!**
2. **If the clinical picture doesn’t make sense, then look at it again. Chest x-rays, for example, that look like infectious issues may not be, especially if the clinical picture doesn’t make sense. Step back, and take a fresh look!**
3. **When a pulmonary mass is suspected, then imaging with CT scanning is usually the next step. MRI is also an imaging modality that should be obtained in patients with suspected tumors. MRI imaging includes not only the area of involvement, but other areas to rule out metastatic lesions. Bone scanning is also employed in many malignant lesion workups to rule out bony involvement.**
4. **Ewing’s sarcoma is typically a tumor of the bones, but can be a soft tissue lesion as well (alone or in conjunction with a bone lesion). It is the second most common form of pediatric bone cancer (the first is osteosarcoma).**
5. **Tumors that are suspicious for Ewing’s may in fact be PNET lesions. FISH studies and biopsy can distinguish the two.**
6. **Treatment for Ewing’s includes chemotherapy, local tumor resection/control, and sometimes radiation.**
7. **5 year survival rates for localized disease is 70%-80%; with metastatic disease it may be 10%-20%.”

**REFERENCES**