"DOCENDO DISCIMUS"

VOL 6 NO 11 NOVEMBER 2019

DCMC Emergency Department Radiology Case of the Month

These cases have been removed of identifying information and 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 or feedback regarding the Case of the Month, feel free to email Robert Vezzetti, MD at rmvezzetti@ascension.org.

This Month:

Pediatric neurologic complaints are not terribly uncommon, but they are anxiety-provoking, both for parents and clinicians, and are seen most often in an acute care setting. The differential diagnosis, etiologies, and variety of presenting symptoms can be broad. The case this month is due to an emerging, and somewhat mysterious, etiology.

dell children's medical center of central texas
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FELLOWSHIP – PEDIATRIC EMERGENCY MEDICINE

PEM Fellowship Conference Schedule: November 2019

6th - 9:00 CNS Infections
10:00 Abdominal Surgical Emergencies
11:00 Grand Rounds: Medicaid Matters

12th - Journal Club

13th - Simulation: Preparation for National USACS Conference

20th - 9:00 M&M
10:00 Board Review
12:00 ED Staff Meeding

27th - NO CONFERENCE HAPPY THANKSGIVING!!

Grand Rounds Speaker: Ryan van Ramshorst
Chief Medical Director Medicaid/CHIP

Simulations are held at the Seton CEC.
Lectures are held at DCMC Command Rooms 3&4.
Locations and topics are subject to change.
All are welcome!

Dia de los muertos. This holiday begins Oct 31 and runs to Nov 2. While largely associated with Mexico, this holiday is celebrated throughout Latin America and parts of the US.

The fare at the first Thanksgiving likely did not include turkey. Rather, lobster, seal, and swan were on the colonists' menu, while the Wampanoag tribe brought venison to the celebration.

Case History

Fall has started to make an appearance in the Pediatric Emergency Department, as plenty of viral upper respiratory tract infections are coming in, along with bronchiolitis, scattered cases of viral gastroenteritis, and, this interesting case. You scan the chief complaint: "Weakness and constipation - difficulty walking." Hmm…

You walk into the room and find a well-appearing two year old child. He has Paw Patrol on his parent's iPad, but is laying down and does not appear to be too interested in the show. Since he does not look ill, you have some time to sit down and speak to the parents. They tell you an interesting story. Apparently their son has had five days of fever to 103 F. The patents note the child, prior this, was diagnosed with "Fifth's Disease" one month ago. He seemed to recover uneventfully from this illness and was in his usual state of health until he developed fever again. He was seen 48 hours into this febrile illness by his pediatrician. At that time, the child was noted to have upper airway congestion and mild cough. He had rapid influenza and rapid strep testing, both of which were negative. Since he was otherwise well-appearing, no further testing was performed and he was diagnosed with a viral infection. He was treated with supportive care (ie oral fluids and anti-pytretics). The next day, he continued to have fever but began to indicate to his parents that he was having pain with ambulation; he would also state "ouch" when standing. He seemed to improve with Tylenol and Motrin, although the fever continued. On the fourth day of fever, which reached 105, the child was brought to the Pediatric Emergency Department for evaluation. At that visit, he was noted to have a temperature of 104; he did have mild upper airway congestion, but an otherwise non-focal examination, including no signs of Kawasaki's Disease. He was ambulatory without apparent pain. He had a chest xray performed, which did not show any findings consistent with pneumonia. A discussion was held with the parents regarding obtaining bloodwork; they were comfortable with some basic laboratory studies (CBC and blood culture); the CBC was not concerning. He was discharged with a diagnosis of a viral infection and followup was arranged with his pediatrician. The next day, then child was noted to have decreased activity and appeared to have difficulty ambulating. He was sent back to the Pediatric Emergency Department for re-evaluation.

His vital signs: Temp - 98.3 HR - 143 RR - 28 BP - 109/59 Sats - 99% (RA). He is not febrile, but he is certainly tachycardic, which is interesting since he is now sleeping. He is exam is unrevealing until you perform a neurologic exam. The patient is not ataxic but can't ambulate because when you stand him up, he appears to buckle at the knees and, while his tone seems equal throughout, he appears to have diminished strength in his lower extremities. He has diminished reflexes throughout. His coordination is grossly normal, as is his sensation. His cranial nerves are grossly intact. After you finish examining him, he goes back to sleep.

You think about the differential. The child does not have overt mental status changes but is fatigued, does not have pain with attempted standing or ambulation, but clearly is weak and not at his baseline, and not meningitic. You consider the differential diagnosis and wonder if imaging is indicated for this child.

Testing for Group A Strep (GAS) Pharyngitis: Guidelines

The Infectious Disease Society of America has issued guidelines for the testing and treatment of GAS infection in pediatric patients:

1. The diagnosis should be established by throat rapid antigen test (RADT) and/or culture. Negative tests should be backed up by culture.
2. Testing is NOT recommended for children with illness features that suggest a viral etiology (cough, rhinorrhea, oral ulcers, for example).
3. Testing is NOT recommended for children < 3 years old (acute rheumatic fever is rare in this age group); if there is an older sibling with GAS, then testing in this age group can be considered.
4. Diagnostic testing or empiric treatment of asymptomatic household contacts is NOT recommended.
5. Followup post treatment cultures are NOT recommended, in general.

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The origins of the celebration are a combination of Aztec and Roman Catholic traditions (All Saints Day on Nov 1 and All Souls Day on Nov 2). Calacas (skeletons) and calaveras (skulls) are commonly seen during the festivities honoring deceased ancestors.

The Differential Diagnosis for this Patient: Weakness

There is a broad differential for this child. History and physical examination are vital to guide further workup.

History: Is the weakness progressive; is bowel/bladder function normal; are there cognitive changes; is there pain; associated symptoms (double vision, changes in voice, trouble chewing/swallowing, shortness of breath); are there sensory changes?
Physical exam: Cranial nerve findings (ptosis, facial weakness, eye movements). Symmetric or asymmetric weakness of the face, eye muscles, or eyelids commonly occurs in disorders of the muscle unit, neuromuscular junction, or motor nerves. Unilateral facial weakness affecting only the lower division would indicate an upper motor neuron lesion in the cortex or basal ganglia. Assess muscle tone and strength. Gait evaluation is important. Assess reflexes (anterior horn cell, neuropathies, and usually muscle disease cause absence of reflexes while in upper motor neuron lesions the reflexes are increased on the affected side). Check rectal tone, as this affected in spinal cord lesions.

Differential Diagnosis:

Cerebral - Stroke, vasculitis, infection, tumor, degenerative disease, seizure, hemisyndrome migraine.

Spinal - Tumor (spinal or root pain, bowel/bladder impairment, sensory level, Brown-Sequard) transverse myelitis (sensory level, bowel/ bladder impairment, sudden onset), infection, trauma.

Anterior Horn Cell - Spinal muscular atrophy (absent DTR's, fasiculations, infants), polio /polio-type illness(asymmetric weakness, absent DTR's, pain, rapid onset, history of diarrhea, fever).

Guillan-Barre - Guillian-Barre (rapidly progressive, ascending or descending, absent DTR's, some mild sensory complaints).

Neuromuscular Junction - Myasthenia (bulbar symptoms, ptosis, worse with exercise, normal DTR's, respiratory insufficiency, autonomic system), botulism (baby, absent eye movements, bulbar symptoms, normal DTR's, hypoventilation), ingestions.

Muscle - muscular dystrophy, myotonic dystrophy, metabolic causes, rhabdomyolysis.

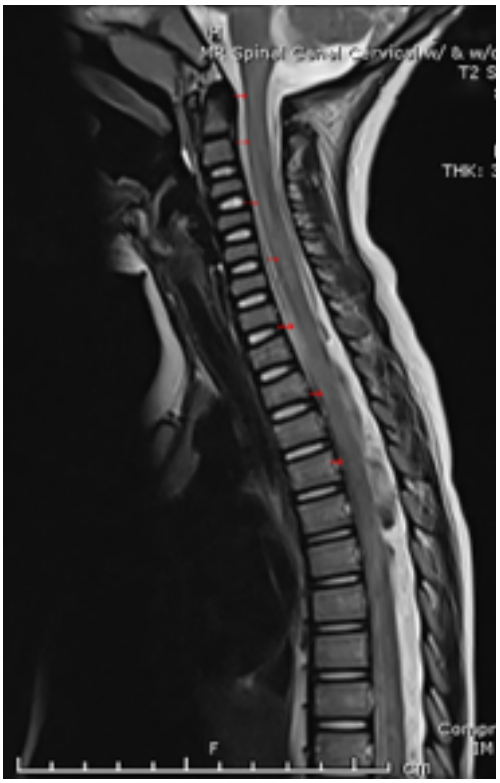
Other - conversion disorder, electrolyte abnormalities.

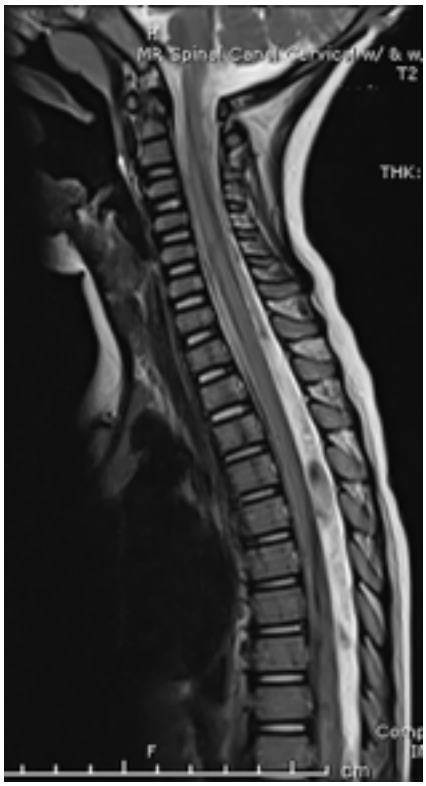
The child's symptoms were concerning for a neurologic process, possibly related to an underlying infectious etiology. In the emergency department, a complete metabolic profile, complete blood count, erythrocyte sedimentation rate, CK, and c-reactive protein were all obtained. The results of these tests were all unremarkable. Given his symptoms, the decision to obtain magnetic resonance imaging of the brain and spine was made. This was done with sedation by Pediatric Anesthesiology. Since the child was sedation, a lumbar puncture was performed as well.

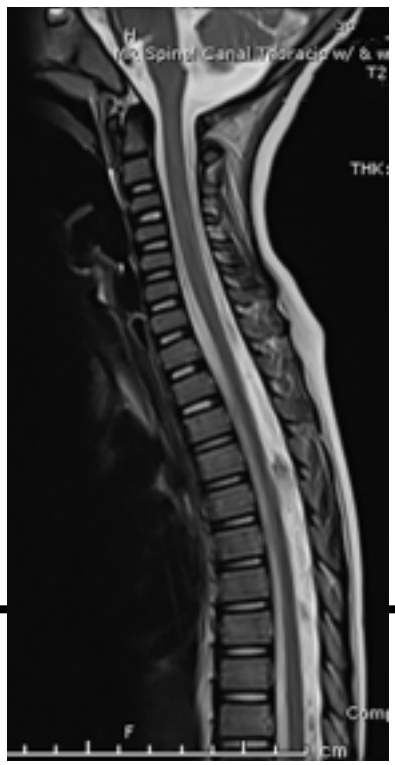
Selected MRI images are provided here. The immediate images to the right demonstrate abnormal T2 hyperintense signal of the central grey matter of basically the entire spinal cord, but worse in the cervical spine.

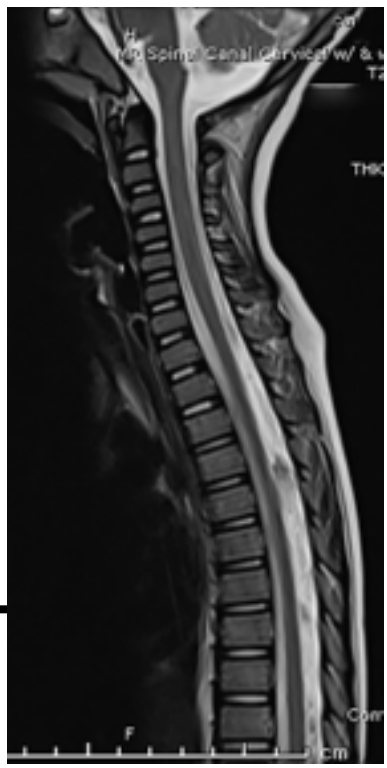
The child's lumbar puncture revealed 55 WBC, 10 RBC, Protein of 16, and Glucose of 56. The Gram stain was negative. The differential was 12 Segs, 8 Lymphs, 3 Monos.

Acute Flaccid Myelitis (AFM) was diagnosed and treatment was initiated. Followup MRI was obtained and selected images are provided (bottom right). These images are about one week after treatment initiation. Here we see significant improvement in the previous abnormal cord signal with minimal residual T2 hyperintense signal, remaining most prominent in the upper cervical cord.









La calavera Catrina is a common symbols associated with this holiday. Originally a zinc etching by Jose Posada, it was meant as satire against adopting European customs.

Calaveras de azucar (sugar skulls) are treats that are eaten during the holiday and they are sometimes used as offerings to the dead.

In 1953 a Swanson employee ordered too many turkeys (260 tons). To put them to use, he hatched the idea of putting the turkey (along with fixings) in aluminum trays. Thus, the first TV Dinner was born.

Acute Flaccid Myelitis

Acute flaccid myelitis (AFM) came to prominence after the emergence of cohorts of children with acute, sudden, unexplained paralysis. Initially, clusters of cases were reported in California in 2012, followed by Colorado and Utah in 2014. By the end of 2014, 120 children from 34 states met the Centers for Disease Control and Prevention (CDC) criteria for what was initially termed "poliomyelitis" or "polio-like syndrome"; ultimately, the term "Acute Flaccid Myelitis" was coined to avoid confusion with illness caused by poliovirus.

The exact cause of AFM remains unknown. Interestingly, there is a noted possible association with various viruses, including rhinovirus, respiratory syncytial virus, parainfluenza virus, and adenovirus, all of which have been isolated from patients with AFM. However, Enterovirus D68 has become an agent of interest in many cases, due to the presence of the virus in children with AFM and in many cases was the only virus isolated. Enterovirus D68 has been shown to cause paralysis in animal models. Additionally, AFM follows a seasonal pattern, with greater incidence in the late summer and early fall and increased identification of AFM occurred when this virus was high in circulation. In 2015, when there were few cases of AFM (only 21 worldwide and no US cases), Enterovirus D 68 was not high in circulation. There is compelling evidence that Enterovirus D68 is the causative agent of AFM, but this is not definitive and more study is needed. There does appear to be enough evidence to state that this virus certainly is associated with AFM.

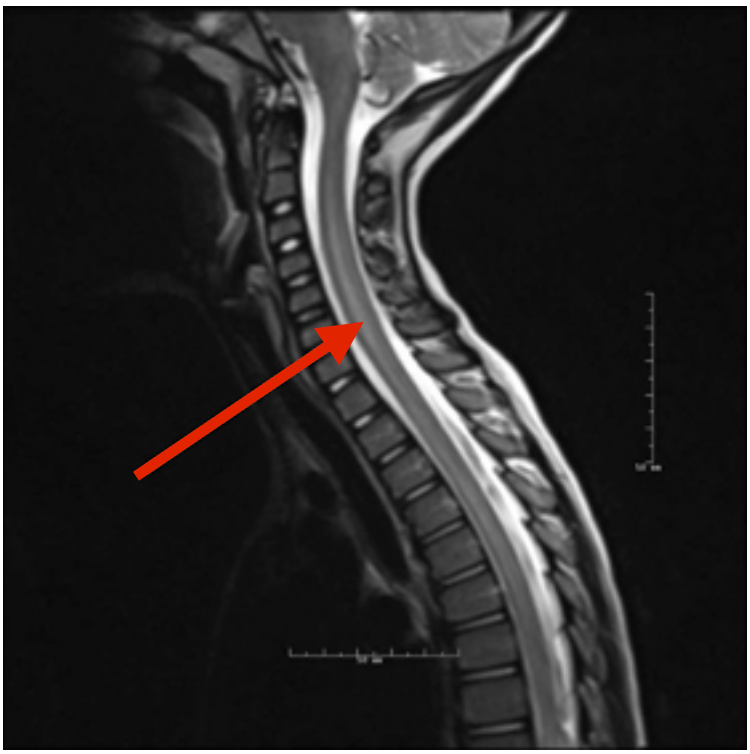
AFM is a disorder affecting the grey matter of the brainstem and/or spinal cord. While the average age of affected children is 4 years, the range is quite wide. Most affected children are previously healthy. The illness begins with symptoms that appear to be a viral illness, with upper respiratory infection symptoms (ie cough, congestion) or gastrointestinal symptoms (ie vomiting and diarrhea). Myalgias follow (most often involving the upper limbs). A rapid progression of commonly asymmetric limb weakness, which can be mild to complete paralysis, follows; this progression can evolve over hours to days. Symptoms can be variable and in the original groups this was the case. For diagnostic purposes, the CDC promulgates the following criteria:

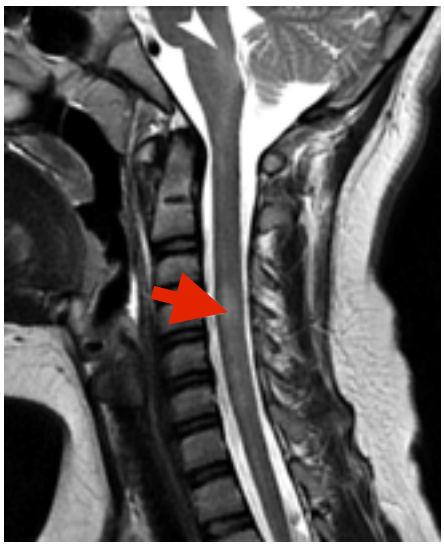
Criteria Diagnostic of AFM:

1. Acute onset of flaccid limb paralysis.
2. Spinal cord lesion on MRI largely restricted to grey matter and spanning one or more segments

Probable Case of AFM:

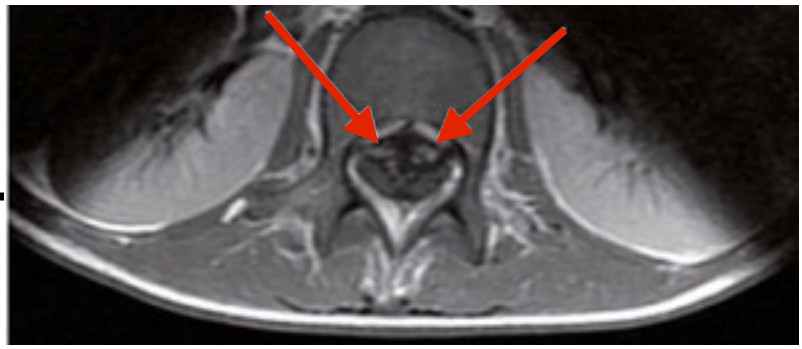
1. Acute onset of limb paralysis.
2. Cerebrospinal fluid pleocytosis.





Imaging AFM

MRI is the preferred imaging modality for AFM. There are characteristic findings: lesions do not enhance with gadolinium and are optimally seen on T2 series. These lesions appears as poorly defined signal abnormalities, involving the grey matter. With disease progression, anterior horn cell involvement can be seen. Multiple cord segments are usually involved and can extend the entire length of the cord. The brainstem (most often the pons) may be involved, and cranial nerve findings can be present clinically.



Cleaning of loved ones graves is an important part of the celebration. Adult graves have orange marigolds while children's have white orchids.

Sarah Josepha Hale convinced President Lincoln to officially declare Thanksgiving a national holiday. She also was a writer and editor and composed "Mary Had a Little Lamb".

The first Macy's Thanksgiving parade did not feature any balloons. It did feature animals from the Central Park Zoo.

Management Options for Acute Flaccid Myelitis

Treatment has been controversial and there are few agreed-upon options that are universally accepted as efficacious. Here are the current CDC Guidelines:

1. There is no specific therapy that is preferred or should be avoided.
2. Consultation with Pediatric Neurology and/or Pediatric Infectious Disease is recommended as quickly as possible.
3. Corticosteroid use has not clear efficacy in controlled trials; use is neither recommended nor discouraged.
4. Intravenous immunoglobulin (IVIG) has shown no clear efficacy and its use is neither recommended nor discouraged. Use of IVIG does not appear to be harmful.
5. Plasmapheresis has shown no clear efficacy in controlled trials and its use is neither recommended nor discouraged. Use of plasmapheresis is not likely to be harmful.
6. Fluoxetine has not been shown to be efficacious and use is not routinely encouraged.
7. Antiviral agents (Acyclovir, for example) should not be routinely used unless there is suspected HSV infection. It is recommended appropriate antiviral medications be initiated until HSV infection has been ruled out.
8. Interferon should not be used; efficacy is not proven and use may be harmful.
9. Biologic agents should not be used; efficacy is not proven and use may be harmful.

CDC Acute Flaccid Myelitis: Interim considerations for clinical management.

Case Resolution:

This child was admitted for further evaluation and treatment. Pediatric Infectious Disease and Pediatric Neurology were consulted. Corticosteroids, Intravenous Immunoglobulin (IVIG), and Acyclovir were begun. Initially he was placed on the inpatient unit, but his weakness progressed and he was briefly transferred to the Pediatric Intensive Care Unit. Gradually the child improved and he was discharged on hospital day #9. At that time, he had improved motor strength but still had deficits. With Pediatric Rehabilitation Medicine consultation, a physical therapy regimen also utilized. At one year post hospitalization, he made a full recovery.

Teaching Points

1. The exact etiology of acute flaccid myelitis in the pediatric population is not known. There is a strong association with a febrile or respiratory illness (up to 80% of patients) prior to the onset of neurologic symptoms. Enterovirus D68 has been implicated as a possible pathogen.
2. Acute onset of limb weakness and variable cranial nerve involvement are typical signs. The weakness is often asymmetric and favor the upper limbs but can occur anywhere.
3. Limb myalgia, mental status changes, headache, neurogenic bladder or bowel are other symptoms that may be seen.
4. Neurologic deficit progression can be rapid; respiratory failure is a potential complication.
5. Diagnostic criteria include: Acute flaccid paralysis and typical MRI lesions. Probable case criteria include CSF pleocytosis and acute onset of flaccid paralysis.
6. Few management options have proven efficacy, but include IVIG, corticosteroids, and antivirals if HSV infection is suspected. Supportive care is essential and multiple subspeciality involvement is needed for optimal management.

References

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