Finding a Foal Amongst the Zebras: An Uncommon Presentation of Lemierre's Syndrome

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Published In/Presented At

Mangla, S., Villalobos, T., Busse, C., Larussa, L. (2015, March 29). *Finding a Foal Amongst the Zebras: An Uncommon Presentation of Lemierre's Syndrome.* Poster presented at: Vignettes category at the Society of Hospital Medicine's Annual Meeting in National Harbor, MD.

Finding a Foal Amongst the Zebras: An Uncommon Presentation of Lemierre's Syndrome

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Objectives

- To recognize the value of tissue biopsies in the step wise work up of fever of unknown origin (FUO).
- To consider Lemierre's syndrome in the differential diagnosis of FUO, even when all of the classic findings are not present.
- To understand the potential complications of Lemierre's syndrome and importance of appropriate treatment.

Patient Presentation

8 year old previously healthy hispanic boy

- History of daily fever spikes 102-105F in the afternoons for >14d with rigors
- Only complaint was vague right upper quadrant abdominal pain
- Three visits to ambulatory setting failed to reveal possible source

Initial Evaluation

- Benign physical exam on presentation. No oral lesions, facial tenderness, pharyngeal exudates, or erythema.
- CT abdomen: Normal.
- Infectious workup:
  - Bacterial culture on day 1 positive for strep mitis and oralis treated with IV vancomycin and ceftriaxone for 5 days until follow up cultures sterile.
  - Bacterial culture on day 15 positive for strep constellatus treated w/ vancomycin and ceftriaxone x 7 days
- Elevated ESR 123 (nml 0-20) and CRP 255 (nml < 7.0)
- Rheumatologic workup negative
- Oncology evaluation including bone marrow aspiration ruled out hematologic malignancy.
- Leukocyte tagged bone scan normal
- US Neck: Normal
- Venous doppler Right Neck: Normal. No DVT.
- CXR: normal
- MRI chest: no evidence of blood vessel abnormality but did reveal a small cystic lesion posteriorly in the right lower lobe adjacent to the pleura.

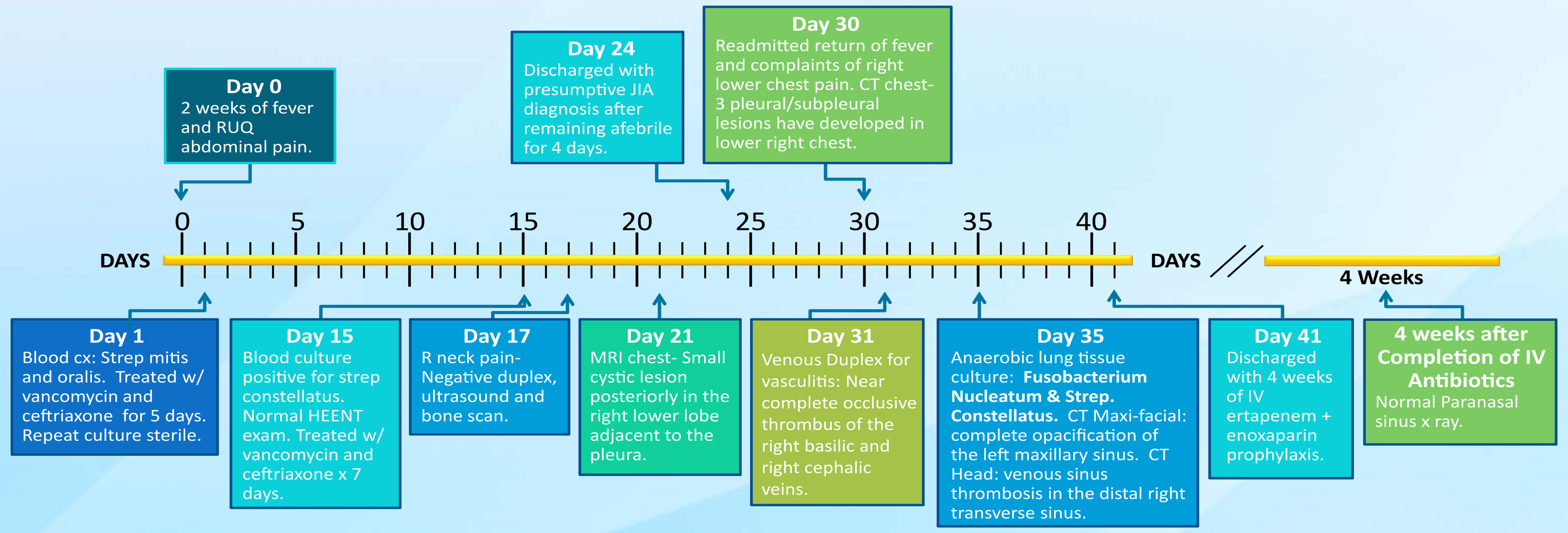
Outcome: After continued daily bimodal fevers for 2.5 weeks, he was afebrile for 4 days on prednisolone for presumed juvenile idiopathic arthritis. He was discharged home in stable condition with follow up outpatient with imaging to monitor cystic lesions.

Subsequent Course

- Readmitted 5 days later for return of fever and complaints of right lower chest pain.
- Chest CT: 3 pleural/subpleural lesions in lower right chest.
- Venus duplex UE for vasculitis: Near occlusive thrombus of right basillic and cephalic veins.
- Lung biopsy via open thoracotomy and wedge resection. Anaerobic lung tissue culture: **Fusobacterium nucleatum**
- Diagnosis of Lemierre's Syndrome.
- CT Head:
  - Non-occlusive venous sinus thrombosis in the distal right transverse sinus and right sigmoid sinus with minimal extension into the right internal jugular vein.
- CT Maxi-facial
  - Paranasal sinus disease with complete opacification of left maxillary sinus.

Outcome: Defervesced within 48h of IV meropenem and completed 5 weeks of IV ertapenem as outpatient with complete resolution of his symptoms. Prophylactic enoxaparin was given while the PICC line was in place.

Timeline



Imaging

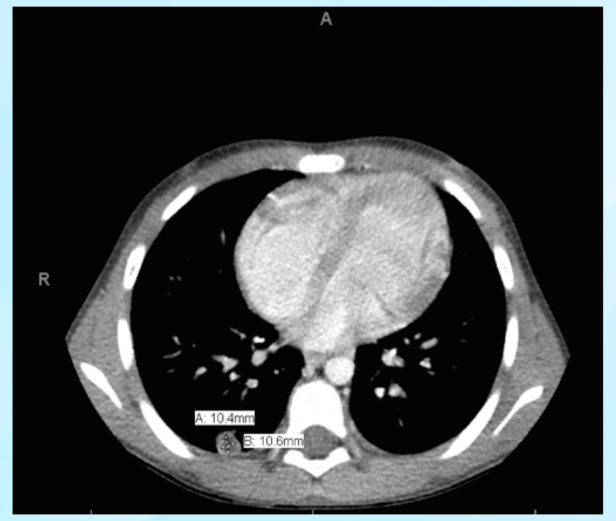


Figure 1.
CT Chest: Cystic Lesion to R lung.

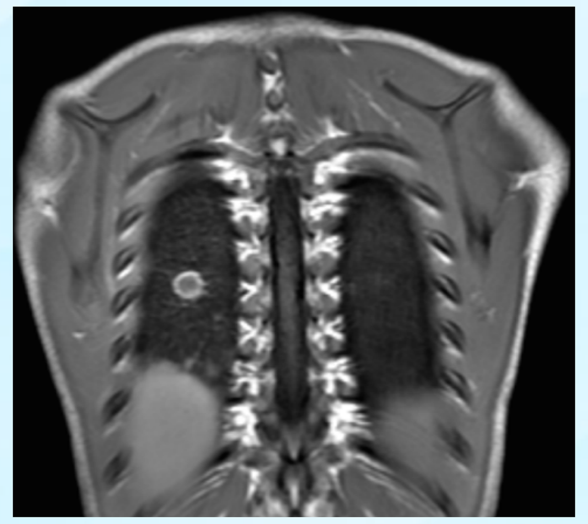


Figure 2.
MRI Chest: Cystic lesion in R lung adjacent to pleura.

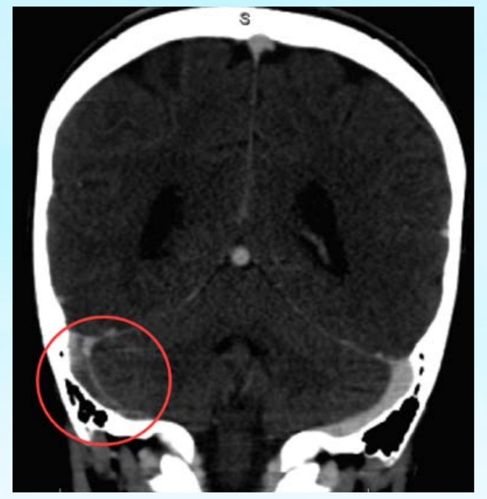


Figure 3.
CT Head outlining nonocclusive venous sinus thrombosis within the distal right transverse sinus.

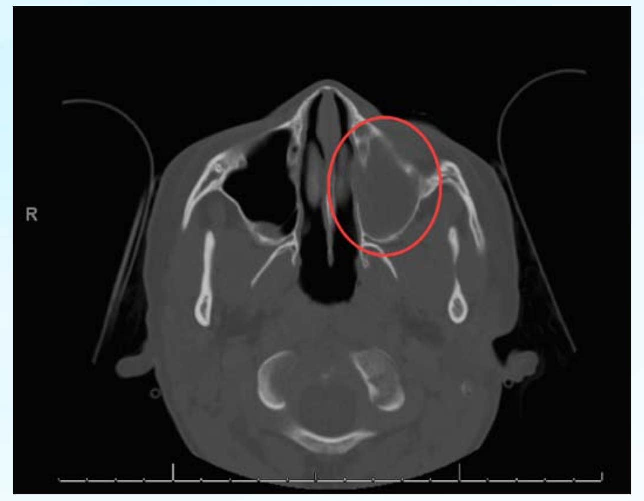


Figure 4.
CT maxofacial outlining complete opacification of the left maxillary sinus.

Clinical Features and Complications of Lemierre's Syndrome

- Classic Presentation: Septic thromboplebitis of the internal jugular vein, but thrombophlebitis of other veins described as well.
- Most common pathogen: **Fusobacteria,** normal flora of oropharynx, anaerobic Gram - bacilli
- Commonly follows primary oropharyngeal infection
- Symptoms: sore throat, neck pain and swelling, high fevers, rigors
- Timing: Primary Infection leading to local invasion of IVJ and septic thrombophlebitis in1-3 week period
- Complications: Pulmonary Infections, Osteoarticular Infections, CNS infections, Mortality Rate 6-18%

Diagnosis and Treatment

- Culture: Anerobic Gram - rod, can take 5-8 days to grow
- Imaging: CT neck best diagnostic modality
- Treatment: prolonged course of IV antibiotics for 4-6 weeks
  - Carbapenems
  - Metronidazole + macrolide
- Anticoagulation: Debatable

Take Home Points

- Lemierre's Syndrome must be considered in all patient with history of oropharyngeal infection who presents with neck pain.
- Clinical picture in children can be more atypical and original oropharyngeal infection may have resolved by the time patient presents with limited exam findings.
- Four to six weeks of antibiotics are indicated to avoid the potentially severe complications, including pulmonary emboli and sinus venous thrombosis.

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