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 102-105°F in the afternoons for >14d with rигors
• 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 mips 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 (ml) (0-20) and CRP 255 (ml < 7.0)
– Rheumatologic workup negative
– Oncologic evaluation including bone marrow aspiration ruled out hematologic malignancy.
– Laskooy tagged bone scan normal
– US Neck: Normal
– Venous doppler Right Neck: Normal. No DVT.
– CRP: normal
– MRI chest: no evidence of blood vessel abnormally 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 prednisone for presumed juvenile idiopathic arthritis. He was discharged home in stable condition with follow up outpatient with imaging to monitor cystic lesions.

Subsequent Course

– Resettled 5 days later for return of fever and complaints of right lower chest pain.
– Chest CT, 3 pleural/subpleural lesions in lower right chest.
– Venous duplex US for varicose veins: Normal
– Lung biopsy via open thoracotomy and wedge resection: Anamnic 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:
– Pancreas sinus disease with complete occlusion of left maxillary sinus.

Outcome: Deterioration within 4th of IV meropenem and completed 5 weeks of IV etrapenem as outpatient with complete resolution of his symptoms. Prophylactic treatment was given while the PICC line was in place.

Clinical Features and Complications of Lemierre’s Syndrome

– Classic Presentation: Septic thrombophlebitis of the internal jugular vein, but thrombophlebitis of other veins described as well.
– Most common pathogen: Fusobacteria, normal flora of oropharynx, anamonic Gram - bacilli
– Commonly follows oropharyngeal infection
– Symptoms: sore throat, neck pain and swelling, high fevers, rigors
– Timing: Primary Infection leading to local invasion of IVJ and septic thrombophlebitis int 3-4 week period
– Complications: Pulmonary Infections, Osteoarthritis Infections, CNS infections, Mortality Rate 6-16%

Timeline

Figure 1. CT Chest: Cystic lesion in R lung.
Figure 2. MRI Chest: Cystic lesion in R lung adjacent to pleura.
Figure 3. CT Head outlining nonocclusive sinus thrombosis within the distal right transverse sinus.
Figure 4. CT maxilo facial outlining complete occlusion of the left maxillary sinus.

Diagnosis and Treatment

– Culture: Anaerobic Gram - rod, can take 5-6 days to grow
– Imaging: CT neck best diagnostic modality
– Treatment: prolonged course of IV antibiotics for 4-6 weeks
– Carben penem
– 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 embolism and sinus venous thrombosis.

REFERENCES:

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