March 2013 Critical Care Case of the Month: Beware the Escargot

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History of Present Illness

A 29 year old woman presented to the Phoenix VA Medical Center with complaints of headache and diffuse generalized weakness most pronounced in the lower extremities. She also noted recent fecal and urinary incontinence, abdominal pain, back pain, numbness in the feet and a non pruritic skin rash on the trunk. Onset of symptoms was about 2 weeks prior to her presentation. Since her symptoms began she had seen in multiple local emergency departments for these same complaints as they worsened and was discharged home in each case with suspected viral syndrome.

PMH, SH, FH

She had no allergies and her past medical history was only significant for post-traumatic stress disorder. She has had no major surgery in her life so far and her family history was not contributory to her current presentation. She smokes marijuana for recreational purposes and drinks alcohol socially. She was not taking any medications on regular basis. She had been in the military until six months prior to her presentation and her service included tours in Alaska and Hawaii. She had recently returned from Fiji. During her stay in Fiji, she reported eating snails and other uncooked food as well as drinking unpurified water.

Physical Exam

Vital signs on presentation- T 98.4°C, P 102 beats/min, R 18 breaths/min, BP150/78 mm Hg O2 sat 97% on room air

She was awake, alert, and oriented. She had mild nuchal rigidity and left ptosis. Lungs were clear and her cardiac exam was normal. Abdominal exam showed diffuse tenderness to palpation with hypoactive bowel sounds. Strength was 5/5 in the upper extremities, 4/5 on the right lower extremity, and 3/5 left lower extremity. Sensation was Intact throughout. Deep tendon reflexes were 1+. Exam was thought to be somewhat limited due to poor effort.

Laboratory findings

White blood cell count was 12,400 mm³ with 75% neutrophils and 8% eosinophils. Hemoglobin- 13.8 mg/dl; Hematocrit-41%; Platelet count was 317,000/mm³ Complete metabolic profile was normal. CPK was elevated at 696 IU/Liter.

Radiology

Chest x-ray showed some blunting of the left costophrenic angle with clear lung fields.

- Which of the following are <u>appropriate</u>?

 1. Observation. She probably has a viral syndrome.
 - 2. Head CT scan
 - 3. Cerebral angiogram
 - 4. Nerve conduction studies
 - 5. Liver ultrasound

Correct! 2. Head CT Scan

Clearly something is wrong. The presence of nuchal rigidity, ptosis, leg weakness and history of fecal and urinary incontinence in an athletic young woman all suggest a potentially serious neurological problem. It is unclear how a cerebral angiogram, nerve conduction studies or a liver ultrasound would be helpful.

CT of the head showed questionable grey-white matter hyperdensities otherwise it was normal. Another important test to rule out a spinal cord lesion is an MRI of the spine. Hers was normal.

CT chest of the chest (representative image - Figure 1) showed multiple small focal areas of consolidation.

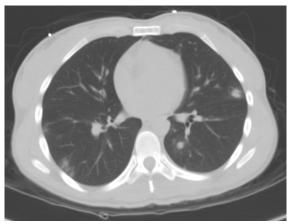


Figure 1. Representative image from the chest CT scan.

Which of the following is *indicated at this time*?

- 1. Begin Artesunic acid
- 2. Lumbar puncture
- 3. Bronchoscopy
- 4. CT guided needle biopsy of the lung lesion
- 5. Begin fluconazole

Correct! 2. Lumbar puncture

Artesunic acid is a medication obtained from the CDC for malaria. Malaria has not been reported in Fiji. Fluconazole is an anti-fungal and might be indicated should she prove to have coccidioidomycosis. However, at this time there is no evidence of coccoides infection. Bronchoscopy and needle biopsy might be indicated but our overriding concern was her neurological problem and for that reason a lumbar puncture was performed. It showed a white blood cell count of 553 cells with 73% eosinophils. Glucose level is CSF was 33 mg/dl. Protein level was 144 mg/dl. Gram stain was negative.

A diagnosis of eosinophilic meningitis was made.

Which of the following are <u>causes of eosinophilic meningitis</u>?

- 1. Angiostrongylus cantonensis
- 2. Coccidioides immitis
- 3. Hodgkin's disease
- 4. Ciprofloxacin
- 5. All of the above

Correct! 5. All of the above

Causes of eosinophilic meningitis fall into the broad categories of infectious, malignant tumors, drugs, foreign bodies and primary eosinophilic meningitis (Table 1).

Table 1. Causes of eosinophilic meningitis

Infectious	Noninfectious			
Angiostrongylus cantonensis	Malignant Tumors	Drugs	Foreign bodies	Primary eosinophilic meningitis
Gnathostoma spinigerum	Glioblastoma	Post myelography	Ventriculoperitoneal shunts	
Paragonimus spp	Hodgkin's disease	Ibuprofen		
Strongyloides stercoralis	Non-Hodgkin lymphoma	Ciprofloxacin		
Toxocara canis	Acute leukaemia	Vancomycin		
Loa Loa	Meningeal carcinomatosis	Gentamicin		
Toxoplasma gondii				
Taenia solium				
Coccidioides immitis				
Schistosoma japonicum				
Fasciola hepatica				
Trichinella spiralis				

Based on the clinical presentation and her travel history in association with eosinophilic meningitis, which of the following is the *most likely diagnosis*?

- 1. Angiostrongylus cantonensis
- 2. Paragonimus spp
- 3. Strongyloides stercoralis
- 4. Toxoplasma gondii
- 5. Coccidioides immitis

Correct! 1. Angiostrongylus cantonensis

Angiostrongylus cantonensis (rat lung worm) is a neurotrophic nematode which is one of the most common causes of eosinophilic meningitis. Rats are the primary hosts for this parasite. Snail, slugs, fresh water prawns, land crabs and various other hosts act as intermediate hosts. Human infections are due to the accidental ingestion of the third stage larvae by eating slugs, other intermediate hosts, or unwashed plants contaminated with snail slime (Figure 2).

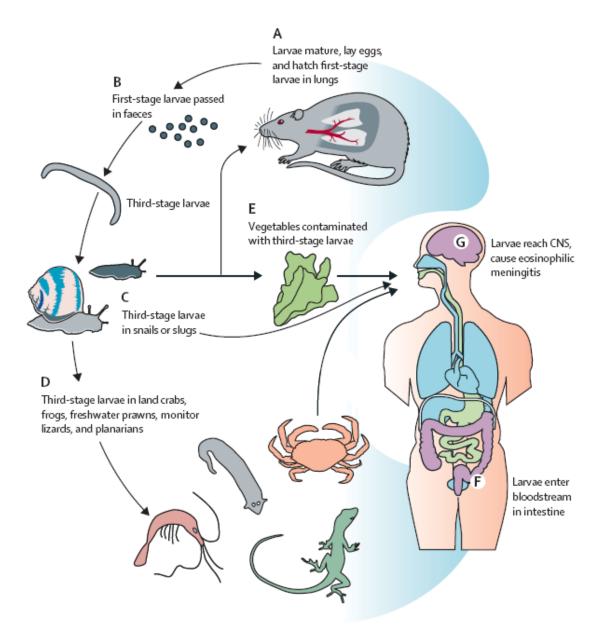


Figure 2. Life cycle of Angiostrongylus cantonensis (1).

Once Ingested the third stage larva migrates to the CNS and develops in to an adult from. The migration and development in the CNS results in intense symptomatic meningitis. Adult worms eventually die in CNS though on rare occasions may migrate to the vitreous humor of the eyes. Infected patients present with typical symptoms of meningitis with headache, fever and somnolence in children. Adults have headache, neck stiffness, parenthesis and quite rarely muscle weakness (Figure 3).

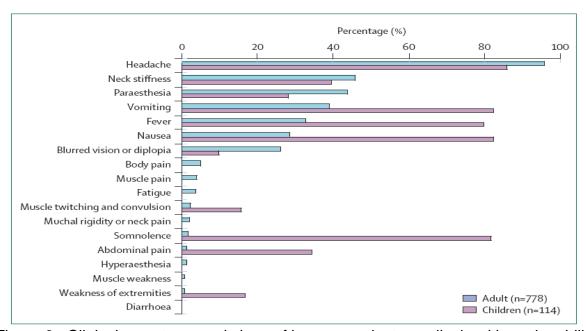


Figure 3: Clinical symptoms and signs of human angiostrongyliasis with eosinophilic meningitis (1).

Most cases of human angiostrongyliasis have been reported from China, Southeast Asia, and the Pacific Islands. However, widespread travel and shipping has resulted in the dissemination of the infection to other areas of the world and the infection is now thought to be endemic in the southern United States, the Caribbean, and the Amazon basin (1-8). Cases are increasingly being reported in the United States and angiostrongyliasis is now the most common infective cause of eosinophilic meningitis in the US.

Which of the following are *appropriate*?

- 1. She should begin physical therapy and rehabilitation
- 2. CSF should be sent to the CDC for *Angiostrongylus cantonensis*
- Albendazole should be started
- 4. Prednisone should be started
- 5. All of the above

Correct! 5. All of the above

The clinical course of angiostrongyliasis is usually improvement even without specific treatment. Mortality rate is low at around 0.5 % (4). People infected with rat lung worm have pulmonary infiltrates which improve with clinical improvement of the patient (1).

In our opinion, the neurologic symptoms in this patient were sufficiently severe to warrant treatment. Oral albendazole and prednisone were begun on day two of the admission (9,10). Both medications were continued for 2 weeks. During the second week of her hospitalization PCR results from the CDC were reported as positive for *Angiostrongylus*. The patient slowly improved clinically with resolution of her headache in 4-5 days. Her lower extremity weakness improved over the next 4 weeks but needed ongoing rehabilitation at the time of discharge. She spent one month in a rehab facility and was eventually discharged home. Follow up chest CT three weeks after admission showed near complete resolution of her lung infiltrates—which coincided with her clinical Improvement (Figure 4).

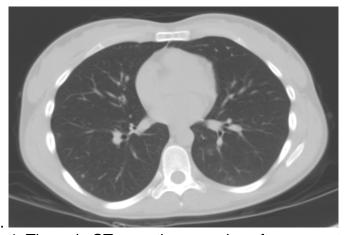


Figure 4. Thoracic CT scan three weeks after presentation.

This case illustrates the classic history, clinical presentation and lung imaging findings of *Angiostrogylus cantonensis* with confirmed diagnosis by PCR.

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