A Rare Cause of Acute Kidney Injury, Forefoot Gangrene and Lower Extremity Weakness

LINC 2018

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Disclosure

WL Gore – Consultant
Bolton Medical – SAB
Cook IDE for Fenestrated and Branch Grafts – no Financial benefit
Spectranetics – SAB
Abbott Medical – Speakers Panel
MT

• **CC/HPI:** 81M known to vascular following 2014 EVAR (Gore Excluder) and previous lumbar fusion. Airlifted from Aruba following sudden onset abd pain, nausea followed by BLE paresthesia and paralysis. Symptoms began approximately 12 hours after arrival in Aruba. No trauma. +Anuria. Concern for acute aortic thrombosis, CTA in Aruba showed no acute pathology. Could not get MRI desired so airlifted to MSW.

• No c/o CP, SOB, f/c.

• **PMHx/SurgHx:** HTN, HLD, CAD s/p CABG, AAA s/p EVAR, Lumbar Fusion

• **Meds:** Inc. ASA, statin, B-blocker

• **Soc:** 40 pack-year smoker, quit 25y ago

• **Fam:** History of AAA, brother had open repair
MT

- Afebrile, AVSS UOP 500cc/6h
- CN II-XII intact
- CTA BL
- RRR
- Abd now soft, nt, nd
- Ext: palpable DP/PT bilaterally. Cyanosis/mottling of toes and distal forefoot
- Neuro: Weakness of hip extension, hip flexion, knee extension, knee flexion, ankle dorsiflexion and ankle plantar flexion on the right.
- Weakness of hip extension, hip flexion, knee extension, knee flexion, ankle dorsiflexion and ankle plantar flexion on the left.
- Flaccid muscle tone below the hips. There is diminished sensation of light touch: left foot. The plantar response is absent bilaterally.
• Relevant labs:
• CBC: WBC 8.6, H/H 11.2/35, Plt 78K
• INR/PTT: 1/35
• BMP: K+ 5.6, Lactate 0.9 BUN/Cr 95/7.2
• CPK 49K
• CRP 285
• CTA A/P
  • Impression: No evidence thrombosis from aorta->tibials. Endograft in place below renals without endoleak.

• MRI Thoracic/Lumbar spine
  • Impression: No cord compression or signs of anterior spinal artery syndrome
• **HD0: Admitted to SICU. Hematologic/vascular, neurologic, renal components**

• **Potential etiologies:**
  3. Neurologic: Embolic with spinal cord ischemia or atypical large fiber demyelination? (eg Guillain Barre syndrome)
  4. Renal: AKI 2’ to TTP?. Strict Is/Os.
  5. Infectious: Afebrile, nl WBC. Sending blood, urine cx.
  6. Rheum: Eval for vasculitis or inflammatory dx. ANCA, ANA, CRP and ESR, C3 C4
MT: Hematological Workup

- LDH trending down, haptoglobin normal, Bilis normal
- HIT panel negative

- RBC - some RBC are microcytic, hypochromic; mild polychromasia; 0-2 schistocytes/HPF, occasional bite cell occasional blister cell, occasional teardrop, occasional basophilic stippling
- WBC - no immature precursors, no toxic granules
- Plts - appear normal, no clumps

- Initial ADAMTS-13 2% consistent with TTP – Started plasmapheresis, steroids.
- Repeat ADAMTS-13 57% ~1w later - rules out TTP. Plasmapheresis stopped. Prednisone tapered.
MT: Neurologic Workup

- Lumbar Puncture – normal
- MRI T/L – normal
- EMG - diminished muscle amplitudes, consistent with myositis or muscle necrosis but ultimately myopathic origin
• Urine output always robust, Cr downtrending
• Renal U/S – no acute pathology, bilateral renal arteries patent and kidneys of nl size
• AKI- ATN sec to rhabdo + ?TTP.
• CPK peaked 70,000.
• c3,c4 normal. Hep BsAg negative, HCV Ab negative, HIV negative.
• Dialysis initiated on HD3 due to fluid overload, uremia.
• Last dialysis session was HD5
• CPK down to 100s from peak CK was 50k
MT: Infectious Workup

- Blood cultures, urine cultures normal
- CXR nl, CTA without any evidence collections/infectious processes
- WBC nl (prior to starting prednisone)
- Afebrile throughout
• CPK, LDH downtrending
• Thigh muscle bx - consistent with rhabdo. unlikely polymyositis.
• No spread of cyanosis, demarcation of right toes
MT: Hospital course

• After ~4w inpt, pt slowly improving. Ambulating with assistance but with bilateral LE weakness, mostly w dorsi/plantar-flexion. Sensation mostly returned.
• Right toes demarcating, mid and fore-foot viable
• Upon discharge:
  – Plt 227 (from 74 on presentation)
  – Cr 1.09 (from 7.2)
  – CPK 600 (from 49k)

• Sent to SAR for continued progress
• Recent 4 toe amputation
Ciguatera Fish Poisoning

- Illness caused by ingestion of reef fish (barracuda, amberjack, moray eel, certain grouper, snapper, or parrotfish) contaminated with toxins from *Gambierdiscus toxicus*, a single-celled organism that grows on coral reefs

- Estimated 20-50k/yr with disease, ~16k in USA, 300 needing hospitalization, 5 deaths/yr

- Ciguatoxin is odorless, tasteless, and not inactivated by normal cooking methods
  - Lowers threshold for opening voltage-gated Na+ channels in nervous system synapses
Ciguatera Fish Poisoning

• Symptoms typically:
  – GI: vomiting, diarrhea, and abdominal cramping, 3-6/h (or as many as 30h) after eating contaminated fish
  – Neuro: 2-72h post-ingestion. peri-oral paresthesias, pruritus, painful urination, blurred vision, temperature-related dysesthesia. Paresis in ~10%
  – Cardiac: within hours, bradycardia, heart block, and hypotension can occur

• Time course:
  – GI: usually resolves within 24-48 hours, rarely 4days+
  – Cardiac: usually resolves within 24-48 hours
  – Neuro: typically few days to several weeks.
    – 20% have sx for months
    – 2% have sx for years
Ciguatera Fish Poisoning: Epidemiology

• **Estimated incidence 20-50k/yr w disease**
  – ~16k cases in USA
  – ~300 pts requiring hospitalization
  – ~5 deaths/yr (usually 2’ to cardiovascular collapse or resp failure)

• **Most cases from tropics or subtropics b/w latitudes 35’N and 35’ S**
  – Pts can be exposed to toxin without traveling after eating fish from these regions
  – But cases reported in Paris, Germany and NYC
Ciguatera Fish Poisoning: Pathogenesis

- Ciguatera fish poisoning is caused by several distinct toxins
  - All formed by dinoflagellates of the genus Gambierdiscus (single-celled algae-like organisms grow on and around coral reefs)
  - Dinoflagellates consumed by large, predatory fish (e.g., barracuda, amberjack, moray eel, snapper, and certain types of grouper) concentrate the toxin in their organs and flesh but are not affected by it

- Ciguatera toxin-containing fish do not taste, smell, or appear unusual
Clinical manifestations often vary based on regions:

• **Pacific and Indian Ocean:**
  - Patients often display early neurologic, gastrointestinal, and cardiovascular findings with neurologic findings predominating.
  - Coma and respiratory distress have also been described, infrequently

• **Caribbean:**
  - Usually presents with gastroenteritis followed by a neurologic illness without mental status changes and is usually not life-threatening
Ciguatera Fish Poisoning

Aftercare

• Consumption of fish, alcohol, caffeine, and nuts within six months of poisoning may trigger a recurrence of symptoms.

• Overexertion/dehydration can cause relapse of ciguatera sx and should be avoided until all initial sx are resolved

• Ciguatera is not an infectious disease: pts do not develop immunity to the toxin
Thank You
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