Introduction:

Welcome to CUGH’s bi-weekly clinical case-series, “Reasoning without Resources,” by Prof. Gerald Paccione of the Albert Einstein College of Medicine. These teaching cases are based on Prof. Paccione’s decades of teaching experience on the medical wards of Kisoro District Hospital in Uganda. They are designed for those practicing in low resource settings, Medicine and Family Medicine residents, and senior medical students interested in clinical global health. Each case is presented in two parts. First comes a case vignette (presenting symptoms, history, basic lab and physical exam findings) along with 6-10 discussion questions that direct clinical reasoning and/or highlight diagnostic issues. Two weeks later CUGH will post detailed instructors notes for the case along with a new case vignette. For a more detailed overview to this case-series and the teaching philosophy behind it, see Introduction to “Reasoning without Resources”. Comments or question may be sent to Prof. Paccione at: gpaccion@montefiore.org

Note: If you would like to be notified when a new case is posted (along with instructor notes for the previous one), send your e-mail to Jillian Morgan at jmorgan@CUGH.org.

About the Author:

Dr. Gerald Paccione is a Professor of Clinical Medicine at the Albert Einstein College of Medicine in the Bronx, New York. His career has centered on medical education for the past 35 years – as a residency Program Director in Primary Care and Social Internal Medicine at Montefiore Hospital, and director of the Global Health Education Alliance at the school. He has served on the Boards of Directors of Doctors for Global Health, Doctors of the World USA, and the Global Health Education Consortium. Dr. Paccione spends about 3 months a year in Uganda working on the Medicine wards of Kisoro District Hospital where he draws examples for the case studies.

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Case 38 – Stiff Stride

A 60 year old farmer was in his usual fully-functional state of health until about a week ago when he began to experience chest pain and “failure to breath” when climbing hills 1-2 times/day which forced him to stop for a few minutes before continuing. Two days later his two shoulders felt “burned” whenever he’d move, without notable abnormal movements or tightening he was aware of, and two days after that his jaw felt “tight” and he couldn’t open his mouth all the way. A day later, the day prior to admission, he began to feel pain in his thighs and hips, and was unable to walk normally. He’s had trouble sleeping for the past week. He had no significant past medical history: no adult hospitalizations, other episodes of chest discomfort, history of hypertension, smoking or diabetes. He’s generally stayed away from health facilities, and didn’t remember taking medications or receiving injections in over 20-30 years. He hasn’t used rodenticides nor ingested any unusual substances. He’s had no fever, weight loss or cough, although this past week his throat felt sore without congestion or rhinorrhea. He’s had no ear pain or discharge, headache, mouth or tooth pain when chewing. He has noted episodes of double vision for the past 2 days that reverted after minutes, and problems initiating a swallow. He recalls no significant trauma or wounds in the past year.

Physical Exam: Anxious appearing older male, sitting straight in bed, flanked by two male friends who walked him to the bed, one bracing each arm.

- BP 126/90 (range first day, 115-128); HR 105 (range first day, 90-112); RR 16; T: 37.2 (range first day, 37.2-38.0)

- Skin: no evidence of trauma, infection, burns or abrasions;

- Head: held slightly extended; face: taut with wide smile, retracted eyes, wrinkled forehead, sweating profusely; can open mouth 2 cm, tongue protrudes with slight deviation to left, masseters tense, no pain while tapping teeth with throat-stick but reflex biting of stick with slow release; (-) Chvostek; Mouth moist, no thrush;

- Eyes: PERRLA, with dilated pupils; EOM full, without obvious strabismus or diplopia elicited; conjunctiva normal, no pallor nor icterus;

- Fundi: benign, without arteriolar narrowing, A/V nicking, hemorrhages or papilledema;

- ENT: ears normal, no evidence of otitis externa or media;

- Neck: mild-moderate rigidity in all directions; no JVP/HJR; no nodes; thyroid normal;

- Lungs: clear to auscultation and percussion

- Heart: PMI normal, non-displaced; normal S1, S2; no murmurs or gallops;

- Abdomen: bowel sounds slightly decreased; tense musculature, no hepato-splenomegaly to palpation or percussion; no tenderness or guarding; Rectal: no masses, stool brown, guaiac negative;

- Extremities: no edema, pulses +2 diffusely; no cyanosis or clubbing; joints/hips normal, without heat, effusion or pain on movement;

- Neurologic: Mental Status: normal orientation x 3; memory intact; anxious but lucid
  - Cranial Nerves: normal vision and eye movements as above; V sensation normal, and VII facial movement normal, with increased resting tone; hearing normal; testing “Gag reflex” instead elicits a strong bite on the throat stick; XI intact; XII: as above, tongue deviates to left;
  - Motor: diffuse muscle tone/rigidity all muscle groups, legs>arms; strength 5/5 diffusely; occasional twitching of muscle fibers evident over chest wall and thighs bilaterally;
  - Sensation: intact to vibration, position, pin prick; Reflexes: +2 diffusely except knees and ankles: +3-4 with myoclonus;
  - Cerebellar: difficult exam due to increased tone, grossly intact; no tremor outstretched hands;
  - Gait: stiff, extended posture, halting, robotic, slow.
1. What is the frame of this case (i.e. the key clinical features from the history and physical that the final diagnosis must be consistent with)?

2. a. What is the significance of the Physical Exam findings?
   b. What exam observation noted here has been published as a highly accurate diagnostic maneuver for the disease this patient has?

3. a. What’s the diagnosis in this case? How is the diagnosis made?
   
   b. What is the epidemiology and pathophysiology of the disease?

   c. What is the spectrum of clinical presentation?
      What’s atypical about this patient’s presentation?

   d. What is the relevant differential diagnosis of this disease?

4. a) Why are prognostic estimates for this disease particularly relevant in rural Africa?
   b) What is the prognosis for this patient?

5. What is the treatment for this disease?