Acute Right Hemiplegia

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Case summary

- 27 y/o woman transferred from outside hospital with “acute right hemiplegia”
- Acute onset of right sided paralysis while riding in taxi → to outside hospital
  - Stroke code called, CT head negative
  - Patient transferred to UW
  - Seen by Neurology in ED – “not a stroke”
Recent history

- Noted increasing abdominal pains over past 1-2 weeks
  - Recent onset of menses
- Poor po intake for weeks d/t moving
- Genital herpes outbreak 2 d ago
- No new medications
- ROS: +photosensitivity and itching (which responds best to IV Benadryl); +dysuria
PMHx

- Acute Intermittent Porphyria (AIP) diagnosed in late childhood
  - Multiple admissions for severe symptoms including 2 admissions with paralysis
    - On ventilator d/t resp muscle paralysis
    - Given TPA once d/t misdx as CVA
  - Always treated with panhematin via portacath and usually gets better in 2-3 days
- No major flares since 2009, all prior care at St. Vincent’s Hospital in NYC
SHx, FHx

- Non smoker, no EToH use or IVDA
- Living in domestic violence shelters, on the run
  - Sought care at EDs/UCs along the way
  - Father is abuser & a police officer & has ‘found her’
- Mom & maternal GF both have AIP, mom is ‘psychotic’ and she doesn’t have contact with her
- Became a Jehovah Witness in 2009
  - Immediately upon admission requested JW liaison to be called to bedside
**PE**

- VS: afeb, BP 133/82, P 102, RR 16, O2 sat 99% RA, wt 140kg
- HEENT: PERRL, EOMI, ? Right facial droop
- Lungs: good effort, clear, dec BS bases
- Heart: tachy, RR, no MRG
- Abd: obese, diffuse moderate tenderness, no rebd, +/- guarding
- Ext: R femoral portacath
- Neuro: CN 2-12 intact except no right shoulder shrug & + v. mild slurred speech; 0/5 R UE/LE; no sensation R UE/LE, hyporeflexic throughout
Labs

- CBC: Hgb 9.9, Hct 32, WBC 14
- BNP normal
- UA c/w UTI
- Upreg neg
Porphyria – disorders of heme biosynthesis
- 8 varieties, AIP most common: 1 in 20,000

Autosomal dominant deficiency in enzyme porphobilinogen deaminase (PBGD)
- Adults; F>M; Most people remain asymptomatic

Activating factor(s) must also be present for symptoms to occur
- Certain meds: sulfa, barbiturates, progestins, sz med
- Low carb/calorie diet (fasting, dieting)
- Hormonal changes premenstrually
- Smoking; stress from illness
AIP – typical symptoms/signs*

- **Abdominal pain (85-95%)**
- **N/V, Constipation (50-80%)**
- **Back, arm, leg pain (50-70%); sensory loss (10-40%)**
- **Muscle weakness (40-70%)**
  - Respiratory paralysis (9-20%)
- **Confusion, hallucinations, depression (40-60%)**
- **Tachycardia (65-85%), elevated BP (35-55%)**

*d/t nerve toxicity from porphyrin build up*
AIP - Diagnosis

- Increased levels of delta-aminolevulinic acid (ALA) and porphobilinogen (PBG) in urine
  - Light sensitive – must collect in dark room and covered bottle
  - Dark/reddish colored urine
  - Trace PBG test – rapid semi-quant test
    - Send-out at UW, 1-4 day turnaround time
- DNA testing in families for genetic defect
Clinical suspicion of acute porphyria

Rapid test for PBG (semi-quantitative, spot urine)

PBG level normal

AIP, HCP, and VP excluded

Measure PBG and ALA (same sample)

Both normal

Acute porphyrias excluded

ALA level increased, PBG level normal

Differentiate causes of ALA dehydratase deficiency (see text)

ALA dehydratase porphyria confirmed: Begin specific treatment

Determine ALA dehydratase mutation

PBG level increased

Acute porphyria (AIP, HCP, or VP) confirmed

Measure PBG, ALA, and porphyrins (same sample)

Plasma and fecal porphyrins
Erythrocyte PBG deaminase (see text for interpretation)

AIP confirmed: Determine PBG deaminase mutation

HCP confirmed: Determine CPO mutation

VP confirmed: Determine PPO mutation

Specific treatment
AIP – Treatment

- Remove any offending medications
- Narcotics for pain control
- Glucose – D10 infusion or oral intake (300-500 grams dextrose daily)
  - Suppresses hepatic ALA synthase
- IV Heme – Panhematin
  - Use if AIP proven by a marked increase in urine PBG or known history
  - Severe, prolonged attacked, esp paresis
Our plan on Saturday

- Dextrose administration IV
- Start Panhematin
  - 4 doses of drug in WI (at Froedtert)
- Treat UTI
- Look up every new drug - website
- Monitor in IMC for progression of neuro symptoms
- Confirm AIP flare with urine testing
- Obtain outside records
Taking a step back....

- Knows a lot about AIP
- Focused on IV dilaudid & benadryl admin
- Requests certain neuro testing not be done
- Cannot verify her history with outside medical records
- Neuro consult: “functional features”
  - + Hoover sign: extension of paretic leg when asked to raise strong leg
When in doubt, consult

- Hematology consult
  - Agreed with starting Panhematin and checking urine PBGs

- Psychiatry consult
  - Patient not overtly psychotic, has dmc
Follow-up

- No change in pt reported neuro sxs with 2 doses panhematin
  - Various clinicians see right hand movement

- Cannot find any outside records, hematologist does not exist
  - Pt stated she had changed her last name

- Urine PBGs – negative: confronted and left hospital

- Went to St. Mary’s, our records available
Factitious Acute Right Hemiplegia: Challenges of Treating Patients Without Universal Electronic Medical Records

Hector Díez-Caballero, M.D.
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Case Report

Factitious disorder is defined as the intentional induction of physical symptoms in the absence of a known physical disease. Patients with factitious disorder intentionally cause their own illness by using self-inflicted injuries, self-poisoning, and production of symptoms. This disorder is mostly diagnosed in young, female patients in their late teens or early adulthood, with right-sided paralysis, sensory loss, slurred speech, and abdominal pain. The patient seemed somewhat more concerned about being given intravenous pain medications for her abdominal pain and less concerned about her hemiplegia.

...history of...sexual abuse by her father. She refused to provide any contact information for family members, saying she was hiding from them.

...the work-up for porphyria returned negative results.

No collateral information was available, and the presence of a portacath gave credibility to her self-reported history.

The patient was transferred to the medical intensive care unit, where she became febrile. Blood cultures were positive for Enterobacter cloacae, raising the suspicion that the patient might have been receiving incorrect porphyria medication.

Once stable, Ms. S. was transferred to the general medical floors, where she became febrile. Blood cultures were positive for Enterobacter cloacae, raising the suspicion that the patient might have been receiving incorrect porphyria medication.

Case

"Ms. S." was a 23-year-old Caucasian, unemployed, single woman who presented to the emergency department with a complaint of right-sided paresthesia, slurred speech, and pain. The patient reported having recently developed these symptoms and was brought to the hospital by an acquaintance who had noticed changes in her behavior. She denied any medical history or exposure to medications but reported a history of sexual abuse by her father. She refused to provide any contact information for family members, saying she was hiding from them.

The patient seemed somewhat more concerned about being given intravenous pain medications for her abdominal pain and less concerned about her hemiplegia.

The work-up for porphyria was ordered, leading the patient to reveal that she had recently undergone an extensive work-up at a local hospital. She consented to disclosure of the hospital records, which showed that she was diagnosed with porphyria. A new porphyria work-up was ordered, leading the patient to reveal that she had recently undergone an extensive work-up at a local hospital. She consented to disclosure of the hospital records, which showed that she was diagnosed with porphyria.
Interesting tidbits...

...the brain attack team wanted to ensure that her presentation was not psychiatric in nature before giving her the tissue plasminogen activator, and thus they consulted psychiatry to assess her for decision-making capacity. Psychiatry determined that the patient had the capacity to accept tissue plasminogen activator...

...the patient was very emotive, discussing at length her history of sexual abuse and adding that she had been a victim of Münchausen syndrome by proxy, reporting that her mother would intentionally withhold her porphyria medication...

Case Report

Fictitious Acute Right Hemiplegia: Challenges of Treating Patients Without Universal Electronic Medical Records

Hector Diez-Caballero, M.D.
Shirley Sostre-Oquendo, M.D.

Fictitious disorder is defined as the intentional fabrication of symptoms with the motivation of assuming a “sick role” (1). The disorder can be challenging and costly to diagnose. Many patients are mood quitters, and personality quitters are frequent comorbidities (3). Multiple etiological factors have been suggested, but the need for nurturance appears to be a critical component (4). Many patients with the disorder have histories of early childhood physical or sexual abuse, disturbed parental relationships, and emotional deprivation (5). Psychosomatic experience suggests that factitious phenomena may be understood as variants of posttraumatic stress disorder (PTSD) (6). We introduce a patient who presented to the emergency department with neurological symptoms and received treatment with tissue plasminogen activator before being diagnosed with fictitious disorder.

Case

“Miss S” was a 23-year-old Caucasian, unemployed, single woman who presented to the emergency department through emergency medical services with a chief complaint of right-sided paralysis, sensory loss, slurred speech, and abdominal pain. The patient reported having developed these symptoms while in an amusement park, prompting emergency ambulances. She reported a medical history of von Willebrand’s disease and was admitted for treatment of PTSD secondary to abuse by her father. She refused to provide any contact information for family members, stating that she was hiding them, and she also refused to provide information for her primary physician, stating that he was an “alcoholic.”

The patient visited the emergency department, physical examination revealed no tachycardia, and chest X-ray was normal. The location of a previous neurological examination revealed an intracranial hemorrhage and a small area of tissue plasminogen activator was administered within the 3-hour window period, and the patient was transferred to the medical intensive care unit.

The next day, an MRI ruled out stroke. During physical therapy, the therapist observed the patient using her right arm for support, but she did not confront the patient with this information. During psychiatric follow-up assessment in the medical intensive care unit, the patient was very emotive, discussing at length her history of sexual abuse and adding that she had been a victim of Münchausen syndrome by proxy, reporting that her mother would intentionally withhold her porphyria medication.

Once stable, Ms. S was transferred to the general medical floors, where she became febrile. Blood cultures were positive for Enterobacter cloacae, raising the suspicion that the patient might have been self-inoculating her infections through her portacath. Because additional signs of sepsis were present, the patient was admitted to the medical intensive care unit for further observation and treatment.

The patient was admitted for a second course of antibiotics, and the decision was made to remove the portacath. The patient was transferred to the medical intensive care unit for further observation and treatment.

During the patient’s hospitalization, she received almost daily psychiatric follow-up...
Factitious disorder

- Classified as “somatic symptoms and related disorders” in DSM 5
- Falsification of physical or psychological signs or symptoms
  - Self or imposed on others
- No obvious rewards to explain why individual is deceiving others
- Must r/o delusions & psychosis
FD

- **Epi:** Acute- F>M, 20-40y/o; Chronic- Men
- **Cause:** h/o abuse or neglect as a child, or a h/o frequent illnesses in themselves or family that require hospitalization
- **1% of hospitalizations!** (More common than AIP)
- **Treatment**
  - Modify behavior, reduce medical resource use, ensure safety
  - Psychotherapy
Warning signs of FD

- Dramatic, inconsistent medical hx
- Unclear symptoms; change once tx begun
  - Predictable relapses following imprvmt
- Extensive knowledge of hospitals and/or medical terminology, & textbook descriptions of illness
- Willingness to have tests, operations, or other procedures
- Lots surgical scars
- Treatment seeking hx diff hospitals/clinics, different cities
- Patient refuses HCP to meet with/talk to family members, friends, & prior HCP
Objectives

- Review presentation, evaluation and treatment of Acute Intermittent Porphyria
- Review factitious disorder and warning signs
References

- American Porphyria Foundation
  http://www.porphyriafoundation.com/about-porphyria/types-of-porphyria/AIP
- Panhematin drug site on AIP http://www.aiporphyria.com/
- Factitious disorder
  http://my.clevelandclinic.org/disorders/Factitious_Disorders/hic_An_Overview_of_Factitious_Disorders.aspx