AAN CHALLENGING HEADACHE CASES

Kathleen B. Digre MD, FAHS, FAAN
Salt Lake City, Utah

Deborah I. Friedman MD, MPH, FAHS, FAAN,
Dallas, Texas

Stephanie J. Nahas MD, MSEd, FAHS, FAAN
Philadelphia, Pennsylavina

We frequently learn the basics and guidelines on many headache diagnoses and treatments, but how do we apply this knowledge to real-life cases that we are challenged by in every day practice where the basics are not enough and even guidelines do not help us? This course is designed to challenge the attendee and the presenters as we work through complex headache cases—highlighting challenges to diagnosis, evaluation, interpretation, and treatment of complex cases. We will highlight guidelines (where they might exist), and best practice. In some cases the true "answer" might not be known, but discussion will focus on how best to proceed when the diagnosis and treatment options are not clear. This course assumes the attendee has had some basic headache medicine background and that the attendee is willing to grapple with complex issues in headache through discussion, sharing, and even debate.

IMPORTANT: We will post the cases ahead of time for the participant. **After the session we will post answers with references and key points made about the cases.**

Upon Completion
Participants will be able to diagnose a variety of headache disorders often with confusing presentations, formulate treatment plans that can incorporate multi-disciplinary regimens, analyze drug treatment side effects as well as interactions in therapeutic options, recognize the importance of co-morbidities that contribute to complexity in headache diagnosis and treatment, and integrate these factors into a therapeutic plan.

PLEASE REVIEW CASES BEFORE ATTENDING THIS SESSION

CASE 1: Treatment for persistent Aura (KBD)
Case of the continuous "aura"

A 17 year old male senior in high school presents with migraine and continuous aura
He has a family history of a twin brother with headache and anxiety. He was car sick as a child. His first headache occurred in 8th grade with severe pain behind his eyes, light and sound sensitivity and severe nausea and vomiting and lasted 6 hours. The next headache occurred that same year when he was in science lecture and looked at phosphorus which was very bright and he developed a blind spot in the center of his vision and then flashing lights; twenty minutes later he had a severe headache with light sensitivity and nausea and this lasted 9 hours. He has had a headache like this about once each month since. He was diagnosed with migraine with aura and he took Sumatriptan but developed chest tightness. He also tried rizatriptan once with success.

In 2014 he started to get silvery lines, a sense of a very tiny camera flash. They are present at least 12-20 times each day. He has trouble concentrating when he sees these things. He has no headache or nausea. They are most prominent when he looks at the sky, snow, or a bright surface. They do not obscure what he is looking at. He also has grainy vision.

PMH: mild trauma as a child—playing soccer took a header without LOC; surgery for deviated septum
Medications: prn acetaminophen; multiple vitamin

SH: good student

ROS: sleep is sometimes difficult;

Examination: VA 20/25 in both eyes; discs c/d 0.2 and normal. Normal neurological examination
MR normal

PHQ9: 4

GAD 7: 3

He now gets a headache about every 6 months. He is sent for prolonged aura. Is this aura? What treatments should we offer?

CASE 2: Migraine, Medication Overuse and Multiple Comorbidities (DF)

Chief Complaint: "I am having really bad headaches."

History of Present Illness: A 33-year-old lab technician (when first evaluated in September 2014) developed headaches when she was 13 years old. They progressed over the years and markedly worsened after having a subarachnoid hemorrhage (SAH) from a right supraclinoid internal carotid blister-like (fragile, thin-walled) aneurysm rupture in June 2014. However, that spring, before the aneurysm ruptured, she was having daily headaches and could not recall her last headache free day.

Her headaches occur each morning, preceded by "floaters or flashes" in her right eye which last for about a minute. The floaters started after the SAH. The headaches are either bifrontal or right-sided, described as pressure or "like someone stuck a knife in my forehead". There is photophobia and phonophobia but osmophobia, nausea or vomiting. They are aggravated with movement or activity and she has to lie down in bed to get relief. She has no dizziness, ataxia or diplopia. She occasionally has trouble getting words out when the headaches are severe but no dysarthria. There is no associated tearing, conjunctival injection, nasal congestion, ptosis, eyelid edema or rhinorrhea. She rates the pain a 7-8 out of 10 in intensity and they last for 30 minutes to 6 hours at the maximum.

She took hydrocodone/APAP 5/325 1-2 tabs for 3-4 times a week since 2012 for her headaches and she started taking it 6 times daily for almost 1-2 months, only recently stopping it after she was told that it might cause "rebound" headaches. She currently takes acetaminophen 6 times a day for past 2 weeks and naproxen twice daily for her headaches. She has been taking daily acetaminophen for years.

Weather changes and menstrual cycle trigger her headaches.

Her headaches affect her social, family life, and also her work. She has difficulty functioning at home. She has been out of work since June and is on FMLA.

Diagnostic work-up and results: CT head without contrast from 8/15/14 reportedly showed a vascular stent extending from the distal right internal carotid artery into the proximal right M1 segment and small area of encephalomalacia in the medial portion of the right temporal lobe.

Past Medical History: NMO spectrum disorder (diagnosed 05/2012, receives rituximab infusions every 6 months), hypertension since age 13 (previously treated with labetalol and Lisinopril, recently changed to verapamil) childhood, hypercholesterolemia and depression. Kidney stones, idiopathic thrombocytopenic purpura. There is no history of fibromyalgia, TMJ disorder, pelvic pain, painful bladder, irritable bowel syndrome.
Surgery: CSF flow diversion device placement on 7/9/14 after a SAH secondary to ruptured right supraclinoid internal carotid artery blister aneurysm (6/22/14), splenectomy for ITP, C-section, kidney stone removal on left side.
Family History: Her mother and sister has migraine headaches.

Current Medications: Sertraline 200 mg daily, verapamil SR 180 mg daily, hydrocodone/APAP 10/325 mg (once weekly), clopidogrel 75 mg daily, aspirin 325 mg daily, docusate sodium 100 mg daily, multivitamin daily, vitamin C 500 mg daily, B12 2500 mcg daily, acetaminophen 650 mg (6 times daily for past 2 weeks), vitamin D3 2,000 U daily, simvastatin 20 mg daily, naproxen 440 mg BID (for two weeks)
Previous Medications
Symptomatic: Hydrocodone/APAP, naproxen, acetaminophen. IV ketorolac, diphenhydramine, ondansetron and valproate along with an oral prednisone taper (no improvement)
Preventive: Gabapentin (tingling in feet and hands), amitriptyline, verapamil, methocarbamol
Procedures: None

Allergies: promethazine

Social History: She never smoked and does not drink. No drug abuse. She lives with her husband and 5-year-old daughter. She has 3 dogs. She does drink coffee now but used to drink a lot of Frappe with caffeine prior to her SAH. She does not use aspartame or any other artificial sweeteners. She has difficulty falling asleep as she is very anxious about her SAH and NMO. She sometimes wakes up in the night due to her headache. She snores lightly with no observed apnea. There is no history of abuse and she is not currently in an abusive relationship. The patient is currently out of work on FMLA because of the aneurysm and is supposed to return to work next week; she has used up her FMLA time.
Review of Systems: Weight gain about 30 pounds in past year. Tinnitus, more prominent in the right ear, which has been chronic for 2-3 years. Gait imbalance for the past 2 years which she attributes to NMO spectrum disorder.

Examination:
Vital Signs: BP 251/94, pulse 73, BMI 40.37 kg/m2, neck circumference 15.5 inches, Mallampati grade 1
Neurological exam showed decreased pinprick on the right face, not splitting the midline. Vibration sense spit the midline on the forehead and nose but not the chin. Pinprick and vibration sense were decreased 75% on the right side compared to the left side of the body. Pinprick loss did not split the midline but vibration sense split the sternum. There was tenderness of the neck or shoulders on the right side in the back. There was tenderness over the supraorbital nerves on the right side. The exam was otherwise normal.

Questionnaires:
Allodynia Score: 12 (severe allodynia)
MIDAS Score: 40 (severe disability) with 90 headache days in the past 3 months (per verbal history, daily headaches), rated an average of 8-10 out of 10 in severity.
General Anxiety Disorder (GAD-7) Score: 10 (consistent with moderate anxiety). The symptoms make it very difficult for her to function.
Sleep Apnea Risk Score: no risk

Patient Health Questionnaire (PHQ-9) Score: 9 (consistent with moderate depression). The symptoms make it somewhat difficult for her to do work, take care of things at home or get along with other people.
Post-Traumatic Stress Disorder Score: Does not support a diagnosis of PTSD

Case 3: Two kinds of pain (SJN)

A 68 year-old woman presented to our clinic in July 2016 with two forms of head pain.

Pain #1
In her 20s, she started experiencing attacks of headache characterized by moderate to severe stabbing pains in the right occipital, temporal, frontal, and eye regions lasting all day when untreated, aggravated by physical activity, and accompanied by neck pain, photophobia, and feelings of anxiety. She denies any nausea, vomiting, phonophobia, osmophobia, diarrhea, constipation, insomnia, increased urination, tinnitis, blurred vision, irritability, memory problems, confusion, increased appetite, decreased appetite, lacrimation, nasal congestion, eye redness, drooping eyelid, or change in pupils. The attacks happen a few times per month, and she treats them with ibuprofen and/or acetaminophen. This helps, but symptoms take several hours to resolve and sometimes her function remains impaired for the duration of the attack.

Pain #2
In 2009, a new kind of pain emerged. She describes severe stabbing pains in the right face and teeth, triggered by touching the skin of the right face, and not accompanied by any other symptoms or signs. There is constant mild right sided facial pain at baseline. Exercise can aggravate this pain. Prior to treatment, she experienced dozens to hundreds of stabbing pains per day, both spontaneous and triggered, with moderate baseline pain but now, attacks of stabbing pain only happen with touching the skin in a specific way, and the baseline pain is mild.

Diagnoses?

PMSHx: She had a surgical procedure for pain #2 December 1, 2015 with some benefit. Otherwise she is healthy.

SocHx: Married, retired school librarian, exercises 5 days/week, no toxic habits.

FamHx: no headaches and noncontributory

Meds:
Gabapentin 900 mg 5x/day
Nortriptyline 40 mg qhs
Multivitamin
Vitamin D3 1000 IU daily
Ascorbic acid 500 mg daily
Ginko biloba 120 mg daily
Ibuprofen 400 mg prn

Allergies/Intolerances:
Carbamazepine – rash all over body (tiny red dots, no blistering/peeling/itching, occurred several month after initiation)
Topiramate – GI distress

Previous Headache Medications

Preventives: Pamelor. Comments: Pamelor 50mg max, current 40, helps. Neurontin, Tegretol, Topamax. Comments: Neurontin 4500mg per day current, max 4800
Topamax- stomach issues
Tegretol- rash when titrating to higher dose, was on for several months before that, did help quite a bit. Klonopin, Comments: didn't help, made her "a zombie". Botox Injections. Comments: had Botox 3x, recalls 25 units each time, in 2013, no benefit. Other. Comments: baclofen helped but no longer needed since surgery.
Abortives: DHE-45, Migranal, Comments: for migrainous HA, effective, years ago. Advil/Motrin, Acetaminophen,

Head MRI: Abnormal **Test Date:** 09/29/2015 : Normal brain, vascular loop on right compressing the trigeminal root entry zone.

Exam is completely normal (VS, general, neuro)

How do you approach each kind of pain now?

Case 4: "He's Not going to school—what can we do?"
15 year old sophomore in high school
Cc: vomiting
HPI: He has a family history of headaches in his mother, who had posttraumatic headaches and a father who gets headaches when he drinks red wine. He also has a maternal grandmother that gets occasional headaches, a maternal aunt who gets significant headaches. He was definitely carsick as a child, and still is with actual vomiting riding in a car.

He, however, has not been headache prone or vomiting prone until 1-2 years before when he began having nausea and vomiting about 2 days at a time.

He was seen by Gastroenterology treated with Zofran with some help and had a GI workup which apparently was normal. He was tried on amitriptyline 30 mg at bedtime, but he still was not sleeping and he continued to have nausea and vomiting.

About 3 months later he began having a bifrontal temporal throbbing headache which would start in the morning, get better in the afternoon and then it would return at night. March through May he missed a lot of school because of headaches and episodic vomiting and he was put on Topamax, which caused flu-like symptoms, Periactin which was not helpful. He had 1 week of vomiting in May and had another GI workup which found some duodenal erosions and it was not clear whether this was related to medication he was taking for headache or the vomiting and nausea. He was hospitalized 6 months later where he was treated with Depakote without help, acetaminophen/butalbital (Fioricet) as well as amitriptyline.

Suring his hospitalization, he had a lumbar puncture, where an opening pressure was 260. He had no change of his headaches whatsoever after the lumbar puncture. He was tried on Diamox 250 mg twice a day, but this made him feel sick and actually made his nausea and vomiting worse so this was discontinued.

At the time he was first seen, he reported bifrontal throbbing headaches associated with severe light sensitivity, minimal sound sensitivity, and nausea and vomiting. The headache gets worse with movement, worse with reading and using a computer and, it is severe in the morning when he wakes up and it will linger through the early morning and it is present at least 4 out of 7 days. It is excruciating once every 2 weeks at which time, he is in bed in a dark quiet room trying to rest. Acetominophen/butalbital does seem to help the headache a little bit but rapid movements, driving,

reading all make this headache worse.

His vomiting now is occurring only in the middle of the night he wakes up 1 or 2 times at night and has vomiting. It used to be all the time and is becoming less common maybe 2 to 3 times a week. He is not chronically light sensitive and is not chronically dizzy.

His past medical history is really noncontributory.
He is on Prilosec 30 mg twice a day, Elavil 25 mg at bedtime, Coenzyme mg twice a day, vitamin B12, multiple vitamins, melatonin 3 mg at bedtime, L'Carnatine 500 mg, Fioricet 1 or 2 as needed the last time he used it was 07/18/2011, Zofran 4 mg as needed he has had none for 2 months And Tylenol 1 or 2 tablets as needed.

Review of systems reveals some weight gain when he was on Depakote, motion sickness, the chronic nausea and vomiting, heartburn, difficulty sleeping. He is unable to sleep fall asleep or stay asleep. He has not had any sleep study. He also has some type of contact dermatitis that is new when he jumped into the field of poison oak.

Family history is positive for arthritis, cancer, and diabetes, depression in the mother after a motor vehicle accident, headaches, heart disease, obesity, stroke, and thyroid problems.

He is a product of a normal pregnancy, normal delivery and has had normal development. He is a good student.

His acuity today was 20/15 OU with contact lenses. He is J1 plus at near. His color vision is 9/9, stereovision is 9/9. His pupils were equal. There was no afferent defect. Visual fields were full to confrontation. Extraocular movements were full. He had a 2 diopter exophoria and he had decreased convergence and divergence amplitudes of 4 to 6 diopters. Slit lamp examination was normal. Dilated examination revealed a small, slightly tilted myopic appearing disk. He had venous pulsations present.

His blood pressure was 116/62, heart rate was 87. His weight was 190 pounds at 5 feet 11 inches. He was well oriented, and gave a very good history and his complete neurologic examination was normal.

His formal visual fields were completely normal. There was no enlargement of the blind spot. A flicker fusion, which is a mini VEP was 38 OU, indicating normal optic nerve function.

We reviewed his MRI scan which showed no empty sella, no increased fluid behind his optic nerves and was basically normal. There was some cerebellar tonsillar ectopia.
The parents are fit to be tied because he has missed so much school—they have tried home school but are concerned about his academic progress. He has been a good student.
What are the diagnoses? What would you suggest for treatment? How will you answer the parents' question?

CASE 5: Headaches with Nocturnal Awakening (DF)

Chief complaint: "I want my life back."

History of Present Illness: A 56-year-old home builder (initial visit January 2017) started having migraines at age 21 which were treated with aspirin/acetaminophen/caffeine. When sumatriptan came on the market, she used the injections with good relief. Then she became "immune" to them and started using sumatriptan/naproxen. Her headaches became more frequent over the years.

She has typical migraine without aura that occurs 4-5 times a week. The migraines are most often right-sided but are occasionally left-sided. Sumatriptan 100 mg taken at the onset of her headaches is very effective; if the tablet does not work, she uses the injection. They last 3-4 days without treatment.

She developed a new type of headache 5-6 years ago that always occurs between 0200-0500. It is bilateral and awakens her from sleep. The pain starts at the back of her head and moves to the front of the head. It feels "like a sinus infection" and is pressure-like and "heavy". The pain is not as severe as her migraines headaches. It initially occurred 1-2 times weekly and now occurs 4-5 times a week. She believes that this headache converts into a migraine. She takes sumatriptan 100 mg for it, with relief. It may be triggered by sleeping on her side and usually does not occur if she sleeps on her back. Most of her current headaches are this type.

Diagnostic work-up: MRI 10 years ago was normal per the patient

Past medical history: Scoliosis, spinal degeneration, hyperlipidemia. Cervical spondylosis, getting physical therapy. Possible fibromyalgia.
Surgery: Breast implants 1990, liposuction
Family history: Her father, paternal aunts and a cousin have migraine. Her nephew has cluster headaches.

Current Medications: Aspirin/acetaminophen/caffeine (2 tablets weekly), calcium with vitamin D, sumatriptan 100 mg (4-5 times weekly), sumatriptan 6 mg SC (1-2 times monthy)

Previous medications:
Symptomatic treatment: Treximet (lost efficacy), Excedrin (somewhat helpful), Imitrex (helpful), Advil (not effective), butalbital/acetaminophen/caffeine, ondasetron 4 mg
Preventive treatment: Amitriptyline-chlordiazepoxide for about 3 years (initially effective but never went up on the dose), verapamil (unsure if effective). Propranolol was prescribed but never started.
Procedural treatment: Botox once (done by plastic surgeon and not the full migraine protocol)

Allergies: None

Social History: She does not smoke or drink. She lives with her husband, mother, son, and one dog. She drinks two cups of coffee per day. She does not use artificial sweeteners. She sleeps 7-8 hours per night. She falls asleep without difficulty but may awaken during the night and her husband snores. There is no history of abuse and she is not currently in an abusive relationship.

Examination:
BP 105/71, pulse 70, BMI 22.6 mg/m2, Mallampati grade 1
Neurological exam was normal with no pericranial nerve, neck or shoulder tenderness

Questionnaires:
Allodynia Score: 4 (mild allodynia)

MIDAS Score: 207 (severe disability) with 60 headache days in the past 3 months, rated an average of 7 out of 10 in severity.
General Anxiety Disorder (GAD-7) Score: 14 (consistent with moderate anxiety) The symptoms make it extremely difficult for her to function.
Sleep Apnea Risk Score: Low risk
Patient Health Questionnaire (PHQ-9) Score: 13 (consistent with moderate depression). The symptoms make it extremely difficult for her to do work, take care of things at home or get along with other people.
Post-Traumatic Stress Disorder Score: Does not support a diagnosis of PTSD

Case # 6: Sudden headache with surges (SJN)

A 38 year-old man presented in August 2016 with new onset headaches. He had no prior headaches until June 21, 2016, when he developed severe frontal throbbing headache accompanied by photophobia and phonophobia as well as fever. On June 24, he also developed back pain, and since his other symptoms persisted, he went to a local ER. After lumbar puncture showed elevated WBCs (lymphocytes) and protein, he was diagnosed with viral meningitis, treated conservatively, and sent home after 2 days with no relief (was told relief would simply take time). Given the persistent and severe nature of symptoms, he returned to the same hospital on June 28, was again admitted for 2 days, and again sent home without relief. He then presented to our hospital's ER July 3, 2016, and received an "IV cocktail," and then that didn't didn't work, came back July 5 and was admitted for more intensive treatment after neurology evaluation. He was inpatient for over 3 weeks getting a variety of IV medications, and ultimately he was sent home on topiramate 75 mg bid and oxycodone 30 mg prn. There has been some improvement but no resolution of the pain.

Previous Testing Results
Head MRI: Normal **Test Date:** 07/18/2016 (no contrast)
Head MRV: Normal **Test Date:** 07/08/2016
Head CT: Normal **Test Date:** 06/25/2016
Lumbar Puncture: Abnormal **Test Date:** 06/29/2016 +WBC, +protein

At the time of first office consultation, he described constant baseline pain 4/10 with surges up to 8/10 from face up to the bridge of his nose lasting about 45 seconds and occurring every 1-2 hours (previously, he experienced several of these per hour and also associated with lacrimation and nasal drainage as well as allodynia on top of his head).

PMSHx: HTN, HLD, hypothyroidism

SocHx: Married, 2 daughters, retail pharmacist, no toxic habits, does not exercise

FamHx: no headaches, non-contributory

Meds:
topiramate 75 mg po bid
oxycodone 30 mg q6 hrs prn
meloxicam 15 mg daily
levothyroxine 175 mcg daily
pravastatin 20mg daily
fish oil 2000 mg daily
cardizem CD 300 mg daily
HCTZ 25 mg daily
ASA 81 mg daily
zolpidem 10 mg po qhs prn

Allergies/Intolerances:
Bee sting (anaphylaxis)

Exam is normal (general and neuro); BP 130/90, HR 74, BMI 26.68
What do you do?

Case 7: What to do—how can I get back to my life? (KBD)
The patient is a 20 year old student who was supposed to be on a mission, but it was discontinued due to headaches.

He has a family history of a mother with 4 migraines in her life. He was never car sick as a child. He would get "dehydration headaches" in high school.

In June 20, 2014 while vacationing in Hawaii with his family. He awakened with a sharp pain on the right side of his head with mild light, definite sound sensitivity. It lasted 2 weeks; imaging was normal at home. The headache resolved.

He received immunizations from May thru July for an upcoming mission to Cambodia. He developed diarrhea, vomiting, fatigue and a rapid weight loss of 25 pounds. He also developed a sinus infection and this was treated with Flonase. The sinus infection didn't improve so he had a nasal scope and was treated with antibiotics and prednisone. He had a headache that started October 9 and it never went away. His diarrhea was evaluated and he was diagnosed with celiac disease. He was placed on a gluten free diet which helped the diarrhea, but his headache did not resolve.

Current headache is a sharp pain in both temples with pounding. He has no light or sound sensitivity but it worsens with activity. He prefers to lie in a dark quiet room. The pain worsens 3 times each week. Things that improve the headache include: diet gluten free, lying down and drinking water. Things that worsen include exercise.

<u>Evaluations</u>
Dentist - received mouth guard, no improvement
MRI Brain - 11/14/2015 Non specific T2 HI, otherwise normal
CT Neck - normal
Optometrist - visual field test, ?optic nerve abnormality (abnormal visual field), Normal IOP
ENT - x2 nasal endoscopy showed remnants of infection
Allergy testing -
Dietician - gluten avoidance helped
GI - endoscopy - celiac disease type 3 to type 1 lesion on follow up. Inflammation improved, but still present.
Celiac antibodies positive
Allergy testing normal
Neurologists- x 2
Echo with bubble (Davis hospital) – normal
Physical therapy, chiropractor
Medications tried:
Acute: Sumatriptan 50 mg (neck tight), zomitriptan 5 mg; toradol 3 days of 30 mg; prednisone, ondansetron, Excedrin, fioricet
Preventive: propranolol 120 mg no help; amitryptiline 50 mg; nortriptyline 25 mg (all preventives 1-2 months)
PMH: diagnosed Celiac disease; irritable bowel syndrome; tonsillectomy
Family history: thyroid disorder mother

Current medications: Amitriptyline 50 mg q hs, hyoscyamine, multible vitamins, probiotic, propranolol 120 mg;
Allergy: sulfa antibiotics (rash)
ROS: insomnia, fatigue, abdominal pain, diarrhea, neck pain, memory loss, nausea, aching pain in eyes
Vitals: BP 110/70 mmHg | Pulse 64 | Resp 14 | Ht 185.4 cm (6' 1") | Wt 90.901 kg (200 lb 6.4 oz) | BMI 26.45 kg/m2 | SpO2 98%

A complete neurological exam was performed. Pertinent findings are below:
Neuro Physical Exam
General: Well-appearing, no acute distress
A complete neurological exam was performed. Pertinent findings are below.
Mental status: Alert and oriented x3
CN: II-XII intact, clear disc margins on funduscopic exam
Motor: Normal tone, 5/5 strength throughout
Reflexes: 2+ and symmetric, Babinski sign absent
Sensation: Intact light touch, intact vibration
Coordination: Normal finger-to-nose, normal heel-to-shin
Gait: Absent Romberg, normal tandem gait
Large pupils
Prominent venous pulsations
Patient's visual acuity for the right eye is 20/30. For the left eye it is 20/40.
MR imaging
MIDAS 0 but 90 days of headache; pain level 7
PHQ 9: 5 (mild depression)
GAD 7: 9 (mild-moderate anxiety)
What is the diagnosis? What to do?

8. **Consult from the ED: Possible Cavernous Sinus Thrombosis**

Chief complaint: Headache and blurred vision

History of present illness: A 23-year-old woman developed bilateral blurred vision the evening prior to evaluation. The blurred vision was worse at distance, to the point where she was navigating in her car based on lights only. She was seen in another ED that evening with a normal examination. She later developed a severe, bitemporal headache with photophobia, pain with eye movement and nausea. The pain was throbbing, rated an 8 out of 10 in severity and she only slept for 3 hours the previous night because of fit. There was no diplopia or ptosis. She had mild headaches associated with menses in the past that were relieved with ibuprofen and has never had severe headache in the past and there was no history of prior visual problems.

Past Medical History: Obese, no other medical problems
Surgery: None
Family History: No history of headaches

Medications: Promethazine 25 as needed, albuterol

Examination:
Vital signs: BP 116/76, P 95, BMI 34.26 mg/m^2

The visual acuity with correction at near was 20/400 OU. As she was reading the card, she held it progressively closer to her and was able to read the 20/20 line with the card about 4-5 inches from her eye OU. Amsler grid testing was blurred but otherwise normal. Visual fields were full to confrontation

OU. Pupils were ~5 mm, sluggishly reactive, with no RAPD. EOM were full with normal pursuits and saccades and no nystagmus. The orbital repositus was slightly increased with pain upon retropulsion of the globes. There was no ptosis and minimal conjunctival injection OU. The undilated fundus exam was normal.

The neurological exam was otherwise normal.

9. My head is on fire and I have no life! (SJN)

A 40 year-old woman presented to our clinic in July 2015 with the following history. In late February 2015, she began experiencing jolts and burning around the right eye and right eyebrow after spending 10 hours on her computer looking for post-doctoral jobs; she had just finished her dissertation. These continued, sometimes at random, but typically with computer work, before improving spontaneously sometime in March. In April she began experiencing scalp pain after 6 hits to the head within a 2 week period (all hits were during the process of moving her place of residence, none were from assault, motor vehicle collision, etc.). The pain recurred, similar to the prior jolts and burning, at the frontal and temporal hairline bilaterally but worse on the right. This evolved to continuous pain with allodynia (unable to wash and comb hair) within a week. Pain is typically worse as the day wears on and has been worsening over time. She attests to some throbbing quality as well as burning sensation in the right eye with some tearing and occasional injection. Brushing her teeth may trigger increased pain in the head, but there is no pain at all in the mouth or lower face.

PMSHx: Notable for sinus polyps for which surgery was done June 2014; she had an endoscopic exam the day prior to initial presentation which was unremarkable.

Social History: Recently completed PhD in Psychology but unable to work because of pain. Married March 2015, no children. No toxic habits. Used to exercise 5 days/week, but unable to since pain began.

Family History: Parents with hypertension, mother with tension-type headaches, osteoarthritis, and thyroid disease, grandmother with rheumatoid arthritis and thyroid disease.

Medications:
Nortriptyline 50 mg qhs
Sertraline 50 mg daily
Pregabalin 150 mg bid
Magnesium oxide 400 mg bid
Melatonin 15 mg qhs
Clorazepate dipotassium 15 mg daily
Acetaminophen 1000 mg daily
Acetaminophen-diphenhydramine 1000-50 mg qhs
Montelukast 10 mg dailu
Levocetirizine 5 mg daily
Allergy shots every 3 weeks
Esomeprazole 40 mg daily
Sucralfate 1 gram daily
Wheat dextrin powder 6 tbsp daily
Bicasodyl 30 mg daily
Acidophilus probiotic cap daily
Lutein and zeaxanthin cap daily
Multivitamin daily
Ester-C vitamin mixture daily
B complex cap daily

Ergocalciferol 10,000 IU weekly

Past Medications:
Preventives: Has been on sertraline since prior to pain onset, paroxetine in past in college. Amitriptyline in past and nortriptyline started recently. Valproate 500mg bid - 5 day trial just finished, made her tired. Pregabalin current 300 mg - on for a month total building up to this dose, no definite side effects other than constipation for which she is also taking symptomatic medication as above. Oxcarbazepine 150 mg bid - nausea, dizzy, rash, stopped after a few days. Gababpentin 1200 mg - took for 4 weeks, didn't help, caused dry, painful skin on face. Sphenoid palatine and stellate blocks: slight benefit from the latter. Melatonin - current since February or March. Magnesium past 10 days.
Abortives: Dihydroergotamine no help, had in ER and infusion, single doses each time. Several opioids in past including hydromorphone, morphine, tramadol, and tapentadol. Butalbital-acetaminophen-caffeine no help. NSAIDs (aspirin, ibuprofen, naproxen) historically not tolerated (gastrointestinal effects, gastritis, breathing difficulty). Prochlorperazine intravenously caused severe restlessness.

Studies:
Brain MRI with contrast May 2015 disclosed normal brain and pituitary with incidental sinus inflammation and small right temporal arachnoid cyst.

Initial Exam is normal unless indicated otherwise:
Height: 65 inches **Weight:** 153.7 pounds **BMI:** 25.67 **Pulse rate:** 80 **Blood Pressure:** 98/72 mm Hg
General Appearance: seems anxious and uncomfortable
HEENT: Normal except for restricted jaw opening since causes pain in temples, also tender temporomandibular joints
Trigeminal allodynia: V1 R>L

How do you help?

10. Periodic, Debilitating Headaches in a 29-Year Old Man (DF)

Chief Complaint**:** "I get severe, debilitating headaches at the scale of 10 that happen every 6 months."

History of Present Illness: A 29-year-old right-handed consultant began having headaches at age 18, during his freshman year of college (evaluated January 2017). They started about a week after he got a DUI and he attributed them to the stress of the event. He still experiences the same type of headache that occurs in bouts.

The headache starts with throbbing pain in his temples, rated a 3-4 out of 10 in intensity and increases to 10 out of 10 in intensity over about 20 minutes. Along with the baseline throbbing pain, he also gets intense “lightning bolt” pain across the temples and stabbing pain in the eyes. It is more often in the right eye it can be bilateral; it never occurs solely OS. He also feels a pulling sensation behind the eye. He has associated photophobia and phonophobia but denies osmophobia, nausea or vomiting. There is no conjunctival injection rhinorrhea, nasal congestion, ptosis, eyelid edema, facial edema or aural fullness but he may have bilateral tearing. He cannot concentrate because of the severity of the pain.

The headaches sometimes wake him up in the middle of the night, usually around 0200-0400. He dreams about the pain and awakens when it is at maximum intensity. Initially, the headaches lasted about 3 hours at a time and he would have to lie down in a dark room. There is no restlessness or agitation. They used to last longer because of the anxiety he had when he first got them. After learning relaxation techniques, they no longer last as long anymore, and resolve in about an hour. However, he is very tired after the headache resolves and is unable to do any activities for the rest of the day. His headaches rarely occur more than once per day.

He gets a "cluster" of headaches every 6 months to 1 year (July-August and October-November are common times). His initial attack in college lasted 3 months. For the first week, it occurred every day, then every 3-4 days and gradually the period between headaches lengthened until it went away completely. His next cycle was about a year later.

He now knows when he is going to have a severe headache and takes oral sumatriptan which aborts it within 20 minutes. He knows it is going to be intense when the others in the cycle have been intense. He takes sumatriptan about once every other week and also has sumatriptan injections if needed. He also takes ibuprofen 200mg daily when he has the headaches though he is not sure if it really helps. He tries cold compresses or pushes on the trochlea when he has a headache. He has noticed that heat makes his headache worse.

He related one particularly bad cycle in July 2013 after attending a bachelor party for which he drove from Tennessee to Alabama. He smoked cigarettes more than he usually does during that party and when he came back from that trip he had a bout which lasted 1.5 months.

His last headache cycle was in October 2016, which was about a year after his last one. It lasted for about two weeks and he only got two headaches during that time period. He took his last sumatriptan tablets during this cycle.

He also gets mild, dull, achy headaches around the temples and in the middle of the forehead that are rated a 1-3 out of 10 in intensity. They occur 3 times per week without associated photophobia, phonophobia, nausea, or vomiting. He is somewhat irritable with them and that moving around or exercising takes his mind off of them. He treats them with ibuprofen.

Triggers include smoking (which he has done on and off for the last 8 years) and stress.

These headaches impact many aspects of his life including his job, relationships, and social life.

Diagnostic work-up and results: MRI Brain without contrast from August 20, 2013 was reportedly normal but the images were not available for review.

Past Medical History: GERD and Barret's esophagus by description, seasonal allergies. He had three head injuries as a child, between 6 and 12 years of age. He lost consciousness during two of them and required stitches for them. During the last one, he hit an artery on top of his head which bled but no loss of consciousness. He was in a motor vehicle accident on 1/7/17. He hit a patch of ice at 30 mph on an overpass, got T-boned and hit the barrier. The car was totaled but the airbags did not deploy. There is no history of fibromyalgia, TMJ disorder, pelvic pain, painful bladder, irritable bowel syndrome.

Surgery: Nissen fundoplication for severe reflux (2001)

Family History: There is no family history of headache. His brother suffers from alcohol and substance abuse as well as bipolar disorder.

Current Medications: Fexofenadine/pseudoephedrine as needed (during allergy season), ibuprofen (when having headache), sumatriptan 100 mg (when having headache), ranitidine (PRN reflux)

Previous medications:
Symptomatic treatment: sumatriptan oral (effective), sumatriptan injection (effective), ibuprofen (less effective), (not effective)
Preventive treatment: None
Procedural treatment: None

Allergies: None

Social History: He smokes occasionally but not since October 2016. He uses snuff 2-3 times per day. He drinks 1 glass of wine or beer per day on average. He uses marijuana occasionally which helps his headache. He lives with his wife and one dog. He drinks 12 oz of coffee per day and uses aspirin/acetaminophen/caffeine. He does not use aspartame. He sleeps well, about 7 hours on average. He wakes up once per night once per 2 weeks or per month, usually because he has work on his mind that he wants to get done. There is no history of abuse and he is not currently in an abusive relationship.

Examination:
BP 105/60, pulse 57, BMI 24.76 $kg.m^2$
The neurological exam was normal.

Questionnaires:

Allodynia Score: 4 (mild allodynia)
MIDAS Score: 39 (severe disability) with 30 headache days in the past 3 months, rated an average of 5 out of 10 in severity.
General Anxiety Disorder (GAD-7) Score: 9 (consistent with mild-moderate anxiety) The symptoms make it somewhat difficult for him to function.
Sleep Apnea Risk Score: low risk
Patient Health Questionnaire (PHQ-9) Score: 11 (consistent with moderate depression). The symptoms make it very difficult for him to do work, take care of things at home or get along with other people.
Post-Traumatic Stress Disorder Score: Supports a diagnosis of PTSD. Event: automobile accident on January 7, 2017.

11. The RED eye headache (KBD)

39 yo woman sent for headache and eye pain.
Patient has a family history of headaches in her sister. She was car sick as a child. She started having headaches in her teenage years. She developed jaw problems in her 20s and was diagnosed with tension type headache. She had jaw surgery and headaches resolved for 5 years. She developed a new headache in August 2014—stabbing pain behind the left eye. The pain is continuous about 5/10 with flares of stabbing. They flare about every 3 weeks when they become really severe: 9/10, conjunctival swelling, rhinorrhea, no ptosis, but eyelid swelling. She may see little bubbles in her vision. Most of the time the pain is on the left side but sometimes on the right side. It can last from 30 seconds to 3 hours. She had one week of continuous pain for 5 days. The longest she has gone without a flare is 3 days.

Headaches are improved with coffee, medication, yoga and oral contraceptives and worsen with alcohol and stress.

She has tried: indomethacin which made her dizzy, verapamil which had no effect, beta blockers, acupuncture, oxygen, steroid, magnesium. Sumatriptan mad the headache worse. She can have associated nausea without vomiting; she has anxiety with flares.

CT normal, MR, MRA all normal. Hypercoaguable work up was negative

PMH: miscarriage x 4, Ovarian cyst, Depression, Iron deficiency anemia
Wears glasses at computer

Jaw surgery 2009, sinus surgery 1994, d and c x2; gall bladder surgery
Current meds: b-complex vitamins, citalopram 20 mg , magnesium, ortho-novum OCP
SH: Married, no smoke or alcohol

Blood pressure 120/58 HR 85
VA 20/25 +3, 20/15
IOP 17. 19 mm Hg
Pupils equal—no dilation lag
Color vision normal
Sle: normal except bilateral chemosis and injection
Discs flat 0.2 c/d
Neurological examination-- normal
Is this cluster headache? How should we treat?