APPROACH TO ACUTE ATAXIA IN CHILDREN

PINTING CHEN
UCLA FAMILY MEDICINE, PGY-2
11/10/21

A CASE OF ACUTE ATAXIA:
THE HPI

- 18 month old F with no PMH presents to the ED with a 2 day history of left leg "shaking". Parents note an increase in clumsiness while walking along with a wobbly gait associated with this left leg shaking. Patient appears to rely heavily on right leg for stability, but is able to put full weight on her left leg.

- The left leg shaking is described as low frequency, and is most noticeable when patient is standing up from a squatting/sitting position. However, symptoms can occur at random regardless of whether patient is walking, at rest, or sleeping.

- There have been no recent trauma, illnesses, fevers, sick contacts, rashes, vomiting, increased irritability or obvious pain for patient. No recent weight loss or change in appetite. No recent travel or camping trips. Patient has remained overall in good spirits throughout.

A CASE OF ACUTE ATAXIA:
THE HISTORY

PMH: none

Allergies: NKDA

Fhx: no fx of any genetic or neurological disorders. No sudden deaths in family.

Soc hx: lives with mom and dad

Immunizations: up to date

Developmental hx: meeting all milestones

A CASE OF ACUTE ATAXIA:
THE PHYSICAL EXAM

- VS: WNL
- HEENT: PERRL, EOMI, no nystagmus, TMs clear
- CV: RRR
- Resp: CTA b/l
- Abd: soft, NT, ND, no HSM or masses
- Skin: no rash
- MSK: Full ROM, no joint swelling, able to move all extremities spontaneously
- Neurologic: Age-appropriate, no obvious focal deficits, normal muscle tone/bulk, no obvious discomfort with ambulation or weight-bearing, some gait unsteadiness/ataxia with intermittent low frequency tremors of LLE, patellar reflexes 2+ bilaterally, no clonus of hyper-reflexia appreciated

A CASE OF ACUTE ATAXIA: THE WORK-UP

Labs: CBC, CMP, Mg, Phos, lipase, amylase, CRP, ESR, CK all normal

CT brain wo contrast: motion-degraded; no large acute intracranial hemorrhage appreciated

XR left hip and pelvis: No acute fracture or dislocation, bony alignment is anatomic; soft tissues are within normal limits; nondilated loops of bowel overlie the pelvis and sacrum

A CASE OF ACUTE ATAXIA: THE IMPRESSION

- What we know so far:
  - Ataxic gait
  - No appreciable focal deficits on exam
  - Patient at neurologic baseline throughout episodes
  - HDS throughout ED stay
  - All labs and imaging unremarkable
- Possible differentials:
  - MSK-related etiology vs neurological disorder
  - Focal motor seizure
  - Unlikely head trauma, brain bleed, tumor
  - Unlikely OM or other focal pathology of LLE
  - Unlikely RLS
  - Unlikely muscular dystrophy

A CASE OF ACUTE ATAXIA:
THE CONSULT

- NEURO CONSULT:
  - Unclear etiology and requires further work-up
  - No need for admission as patient HDS with no focal deficits or change in baseline behaviors.
  - Low suspicion for seizures
- PLAN:
  - Discharge home with close neurology follow-up
  - Strict return precautions given
  - Will directly admit if presents again

A CASE OF ACUTE ATAXIA:
THE RETURN

- One day later, the patient is brought to the ED again for persistent left leg tremors, though now with involvement of L arm
- Strength and sensation still intact, but with decreased coordination
- Patient is admitted

ACUTE ATAXIA

Disturbance in the smooth, accurate coordination of voluntary movements

Ataxia is usually the result of cerebellar dysfunction

Acute ataxia is defined as the presence of ataxia for < 7 days

CAUSES OF ACUTE ATAXIA

- Acute infections
- Post-infectious inflammatory conditions
- Toxins
- Tumors
- Trauma

LIFE-THREATENING CAUSES

Tumors

- Posterior Fossa:
  - Slowly progressive ataxia and symptoms of increased ICP
  - +/- N/V, headache
  - Papilledema, cranial neuropathies, focal neurologic abnormalities
- Neuroblastoma:
  - Acute ataxia + opsoclonus-myoclonus

Intracranial hemorrhage

- Severe trauma
- Vascular malformation

LIFE-THREATENING CAUSES

Stroke:

- Sickle cell disease
- Hypercoagulable states
- Homocystinuria

Infection:

- Cerebellar Abscesses
- Brainstem encephalitis
- Acute disseminated encephalomyelitis (ADEM)
- Cerebellitis

COMMON CAUSES

- Acute cerebellar ataxia (most common cause): self-limited; usually post-infectious; dx of exclusion
- Guillain-Barre syndrome: post-infectious, immune-mediated process
- Toxic exposure: Associated sxs include lethargy, confusion, inappropriate speech or behavior
- Labyrinthitis
- Migraine syndromes or BPV
- Trauma: mild TBI

OTHER CAUSES

Hypoglycemia	Seizure disorder	Opsoclonus myoclonus syndrome	Inborn errors of metabolism
Tick paralysis	Congenital anomalies	Degenerative/genetic conditions	Conversion disorder

EVALUATION: OBTAINING THE HISTORY

- Onset of symptoms
- Associated symptoms
- Access to medications or toxins
- Head trauma
- Recent infection or vaccination
- Previous episodes
- Family history

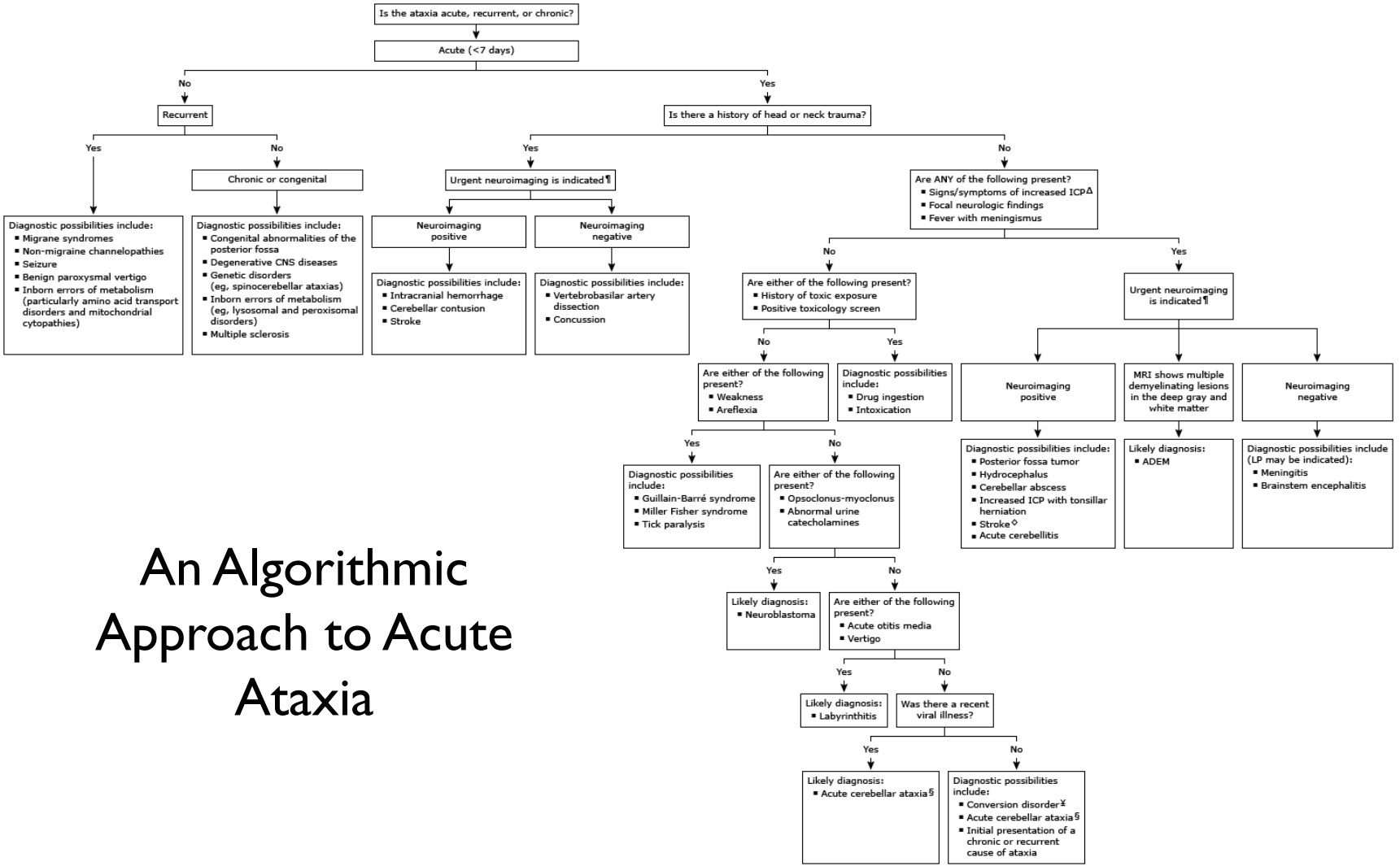
EVALUATION: PHYSICAL EXAM

- Vital signs
- Bulging fontanelle, head tilt
- Fundoscopic exam
- Nystagmus
- Otitis media, hearing loss
- Meningismus
- Healing rash
- Neuro exam

EVALUATION: LABS AND IMAGING

Utox	Blood glucose	Metabolic evaluation
CSF*	Neuroimaging	EEG*

An Algorithmic Approach to Acute Ataxia



BACK TO THE CASE…

Further work-up results

- vEEG: negative
- MRI brain, C-spine: wnl
- Utox negative
- LP with increased WBC, otherwise unremarkable (negative culture, VZV, HSV, MBP, enterovirus, meningitis/encephalitis panel)
- ASO, DNAse, bartonella, HHV-6 negative

Management

- ID, neuro consulted
- s/p acyclovir tx x 1

THE PATIENT'S COURSE

Gait abnormality progressed from patient's left side to her head and eventually to her right side →

In discussion with specialists, patient's clinical picture was most consistent with acute cerebellar ataxia →

By hospital day 5, patient had shown improvement in her ambulation, though still had ataxic gait →

Patient was discharged with plans to f/u with PCP, OT/PT for rehab

ACUTE CEREBELLAR ATAXIA

Epidemiology:

- Post-infectious disorder
- Accounts for 35-60% of pediatric ataxia
- Usually occurs in children under 6

Pathogenesis:

- Likely autoimmune
- Some studies have shown infection of brain tissue

ACUTE CEREBELLAR ATAXIA

- Presentation:
  - Rapid onset and progression of symptoms over days
  - Can occur within days to weeks of a prodromal illness
  - Gait disturbance is primary symptom
  - Associated symptoms:
    - Nystagmus, slurred or garbled speech, vomiting, irritability, dysarthria
    - Absent: fever, meningismus, seizures

ACUTE CEREBELLAR ATAXIA

- Diagnosis of exclusion
- Treatment:
  - Supportive
  - Antiviral medications do not appear to alter disease course or outcome
- Prognosis
  - Resolves without sequelae within 2-3 weeks of presentation in most cases

BUT WAIT, THERE'S MORE…

- Urine catecholamines came back with elevated dopamine
- Patient presented to RR 6 days after discharge from SMH with new symptoms:
  - L eyelid flickering, bilateral L to R horizontal movements
  - Increasing lethargy
  - Loss of truncal support
  - Slurred speech
  - Increased drooling
  - Refusal to walk or eat

MORE WORK-UP AND FINALLY A DIAGNOSIS!

- Now likely opsoclonus-myoclonus syndrome
- US abdomen
- MRI chest/abdomen/pelvis
- Urine catecholamine f/u studies: homovanillic acid and vanillylmandelic acid
- mIBG scan

The diagnosis?

NEUROBLASTOMA

PATIENT PROGRESS

- S/p resection and biopsy
- S/p rituximab
- S/p IV pulse dose steroids
- Currently receiving monthly IVIG
- Speech, vocabulary improving significantly
- Still with some motor issues but is walking steadily now

REFERENCES

- Agrawal D. Approach to the child with acute ataxia. UpToDate, Post TW (Ed), UpToDate, Waltham, MA.
- Gilbert DL. Acute cerebellar ataxia in children. UpToDate, Post TW (Ed), UpToDate, Waltham, MA.
- Caffarelli M, Kimia AA, Torres AR. Acute Ataxia in Children: A Review of the Differential Diagnosis and Evaluation in the Emergency Department. Pediatr Neurol 2016; 65:14
- Stumpf DA. Acute ataxia. Pediatr Rev 1987; 8:303
- Casselbrant ML, Mandel EM. Balance disorders in children. Neurol Clin 2005; 23:807
- Matthay KK, Blaes F, Hero B, et al. Opsoclonus myoclonus syndrome in neuroblastoma a report from a workshop on the dancing eyes syndrome at the advances in neuroblastoma meeting in Genoa, Italy, 2004. Cancer Lett 2005; 228:275

THANK YOU!