Acute onset cerebellar ataxia

Taking a closer look into acute cerebellitis…

Clinical features:

- Ataxia + multiple symptoms: headache, vomiting, and altered consciousness.
- Most common etiologic agents: *Varicella Zoster* virus, *Epstein Barr virus, Herpes Simplex virus-1, Influenza, Respiratory Syncytial Virus, Rotavirus.*

Imaging features:

- Typically displays:
  - T1: cortical hypointensity
  - T2/FLAIR: cortical hyperintensity
  - DWI/ADC: may show restricted diffusion
  - T1 C+: may show cortical enhancement

Patients mostly have a benign, self-limiting condition. Steroids are typically used.

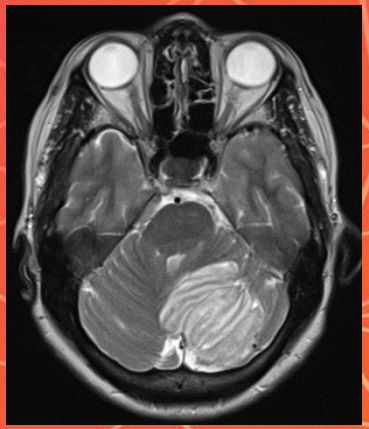
Attention on these differential diagnosis:

Postinfectious cerebellar ataxia:

- Autoantibodies target the Purkinje cells
- Isolated ataxia, typically

Lhermitte Duclos disease:

- Acute cerebellitis may present in a pseudotumoral form that mimics Lhermitte-Duclos disease
- It is a grade I WHO tumour, also known as dysplastic cerebellar gangliocytoma

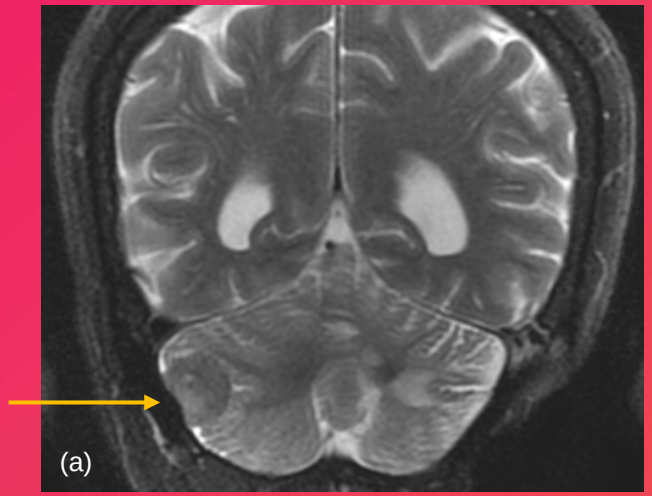


Typical presentation of Lhermitte-Duclos affecting the left cerebellar hemisphere with its laminated appearance

Subacute onset cerebellar ataxia

Case 3:

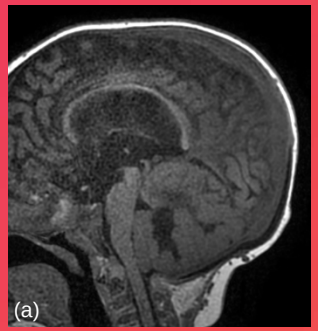
43-year-old man with slurred speech and headache

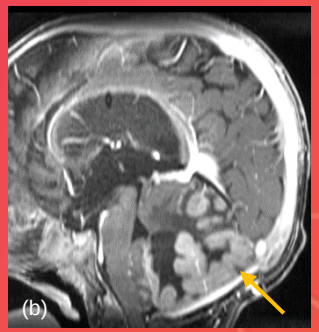


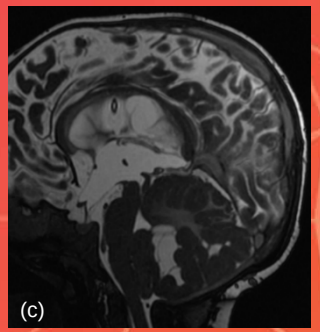
T2W image (a) demonstrates a well defined extra-axial mass at the right infratentorial region with a broad dural base and a dural tail, features in keeping with a **posterior fossa *meningioma.***

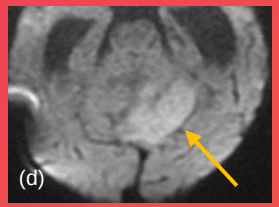
Case 4:

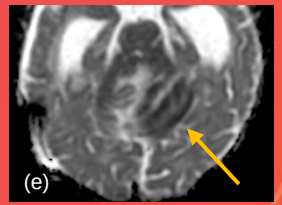
7-year-old male with headache and gait imbalance











(a) and (b) demonstrate avid enhancement of a midline mass in the roof of the 4th ventricle in T1W sagittal images.

FISP sequence (c) demonstrates its cystic component. There is also restricted diffusion on DWI and ADC map (d and e).

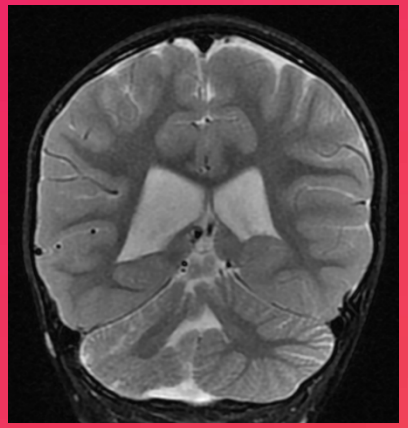
These features are typical of ***medulloblastoma***, the second most common brain tumour of childhood.

AP featured a desmoplastic / nodular pattern medulloblastoma.

Chronic and congenital onset cerebellar ataxia

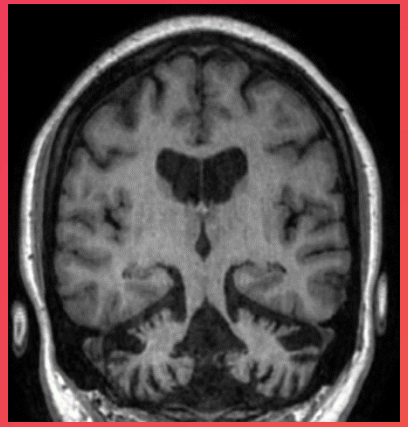
This is the subgroup in which carrying out the differential diagnosis is more troublesome

Describing cerebellar morphologies along with their associated conditions is a reasonable first step into differentiating the causes for chronic / congenital onset cerebellar ataxia



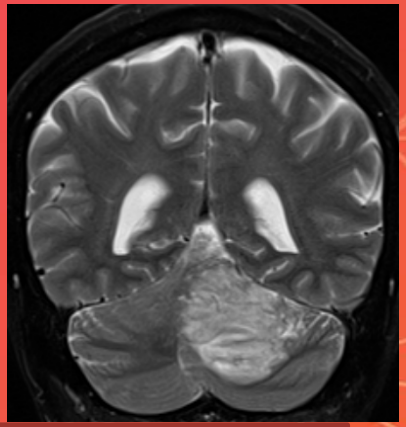
Cerebellar hypoplasia

- Malformed, irregular contours, asymmetric appearance
- Stable appearance
- Can be global, unilateral or affect the vermis



Cerebellar atrophy

- Enlarged *sulci*
- Progressive
- Usually genetic or postpartum acquired
- Mostly global



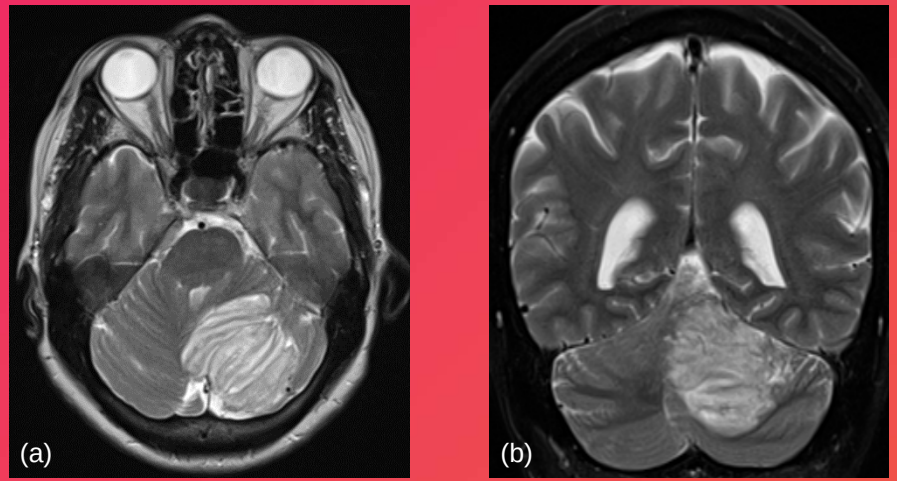
Cerebellar dysplasia

- Abnormal cerebellar foliation, white matter arborization and gray-white matter junction
- Global or unilateral
- Cortical / subcortical cysts may be present

Cerebellar dysplasia

Cerebellar dysplasia is the most uncommon cause for cerebellar ataxia

Very few posterior fossa malformations are reported (Chud-McCullough syndrome, a-dystroglycanopathies, GPR56-related polymicrogyria, and Poretti-Boltshauser disease)



Lhermitte Duclos disease:

- Rare and benign cerebellar tumour
- T2 hyperintense / contrast enhancement is rare
- Associated with Cowden syndrome

T2W axial and coronal images demonstrate widened cerebellar folia and the typical tigroid appearance of Lhermitte Duclos disease

Take home messages

Cerebellar ataxia onset

Acute onset

Ischemic and hemorrhagic strokes / cerebellitis / parainfectious syndromes / toxic disorders

Subacute onset

Mass lesions in the posterior fossa / Creutzfeldt-Jakob disease / vitamin deficiencies / alcohol abuse

Chronic / congenital onset

Check cerebellum morphology

Atrophic

Pure cerebellar atrophy

ataxia-telangiectasia, several spinocerebellar ataxias

With white matter abnormalities

peroxisomal disorders, Niemman-Pick type, Wilson disease, mitochondrial disorders

With dentate nucleus signal change

L-2-hydroxyglutaric aciduria; CTX; Wilson disease; Succinic semialdehyde dehydrogenase deficiency

With basal ganglia involvement

Kearns-Sayre syndrome and other mitochondrial disorders; Cockayne syndrome; Aicardi-Goutieres syndrome;

With cerebellar cortex T2-hyperintensity

Marinesco-Sjorgen syndrome; congenital disorders of glycosylation 1a; Christianson syndrome; coenzyme Q10 deficiency; mitochondrial disorders

Dysplastic

Rare causes (Lhermitte Duclos)

Hypoplastic

Hindbrain anomalies

Disruptive events