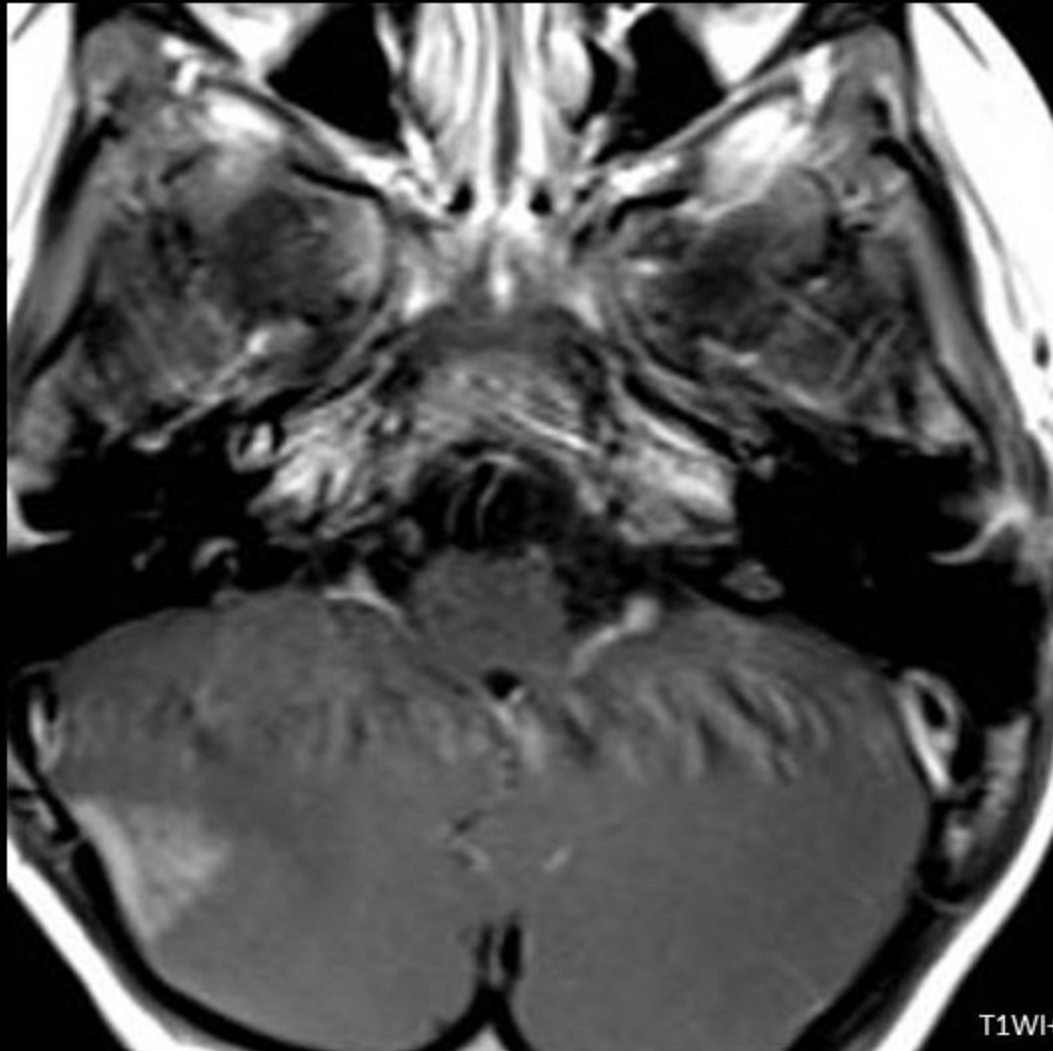


CASE 1



4-year-old boy with Tuberous Sclerosis and an enlarging, newly enhancing cerebellar lesion



V. Michelle Silvera MD
Boston Children's
Hospital, Boston, MA

History

Images

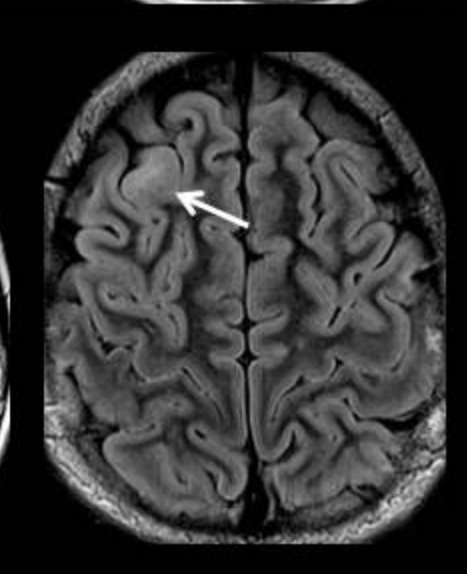
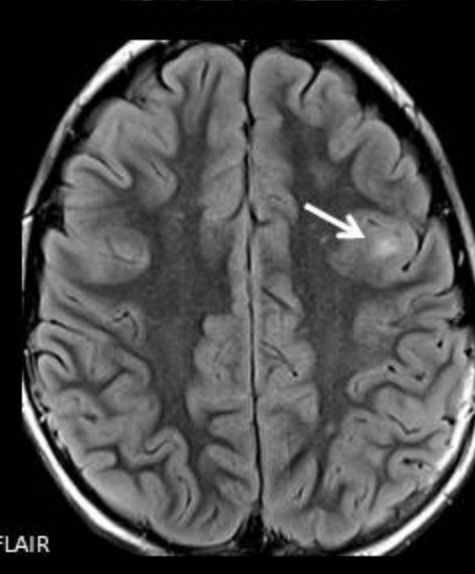
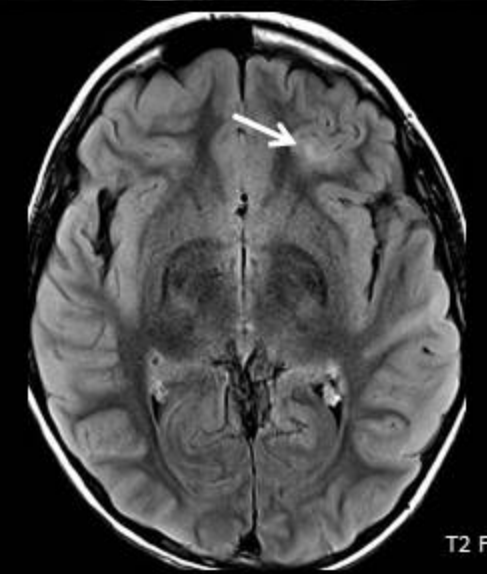
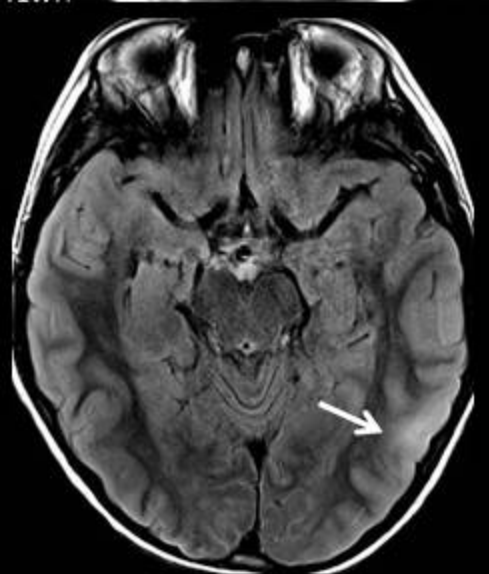
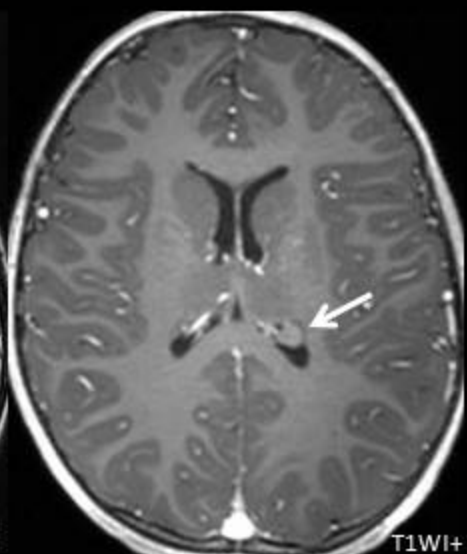
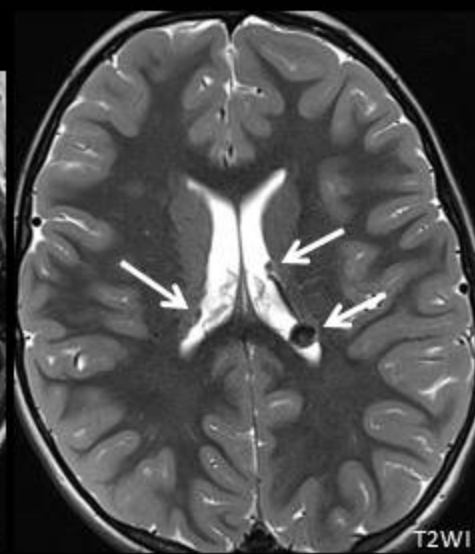
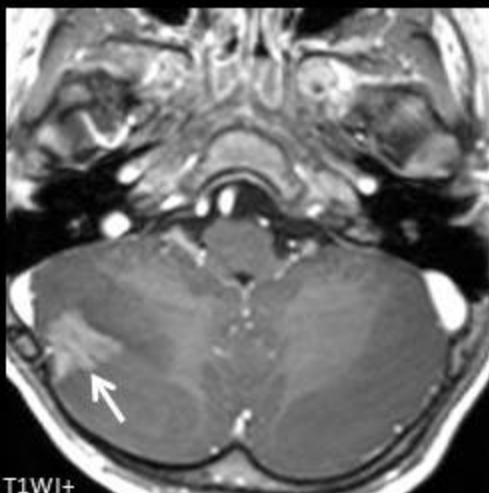
DDx

Diagnosis

Discussion

References

Imaging



T2 FLAIR

History

Images

DDx

Diagnosis

Discussion

References

Imaging



T1WI+



T2WI



TRACE



ADC

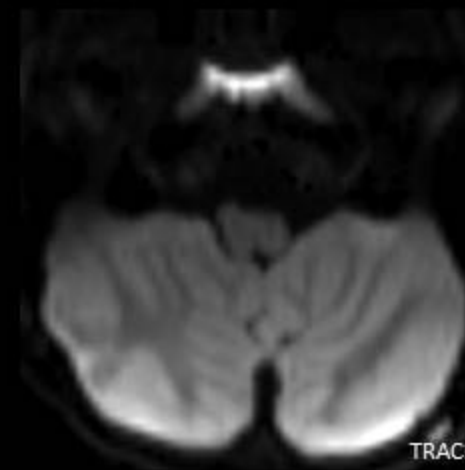
At 20 months of age



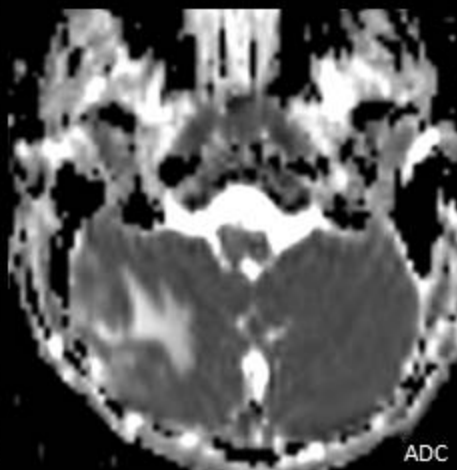
T1WI+



T2WI



TRACE



ADC

At 4 years of age

History

Images

DDx

Diagnosis

Discussion

References

Findings

- Scattered cortical and subcortical foci of T2 signal prolongation within both cerebral hemispheres consistent with tubers
- Nodular, circumscribed lesions, dark on T2WIs along the margin of the lateral ventricles, consistent with calcified subependymal glial nodules
- Right cerebellar wedge-shaped lesion, increased in size and showing new striated enhancement

DDX

- Cerebellar Tuber
- Lhermitte Duclos (dysplastic cerebellar gangliocytoma)
- Juvenile Pilocytic Astrocytoma

Diagnosis

Cerebellar Tuber in a Patient with Tuberous Sclerosis

History

Images

DDx

Diagnosis

Discussion

References

Discussion

- Tuberous Sclerosis (TS) is a neurocutaneous disorder characterized by seizures, mental retardation and adenoma sebaceum
- 2/3 of TS cases are sporadic and 1/3 are inherited as an autosomal dominant condition
- TS is caused by mutations in the TSC1 or TSC2 tumor-suppressor genes that encode for hamartin and tuberin, respectively
- CNS imaging abnormalities include: cortical/subcortical tubers, white matter abnormalities (e.g. radial bands), subependymal glial nodules, subependymal giant cell astrocytomas, parenchymal cysts, retinal phakomas and hyperostosis of the skull

Discussion

- 1/3 of patients with TS may have a cerebellar tuber
- Cerebellar tubers are typically wedge-shaped in configuration
- Cerebellar tubers may show enhancement, typically in a “zebra-like” pattern, can calcify, show folia distortion and retraction abnormalities and may increase in size
- Cerebellar tubers should not occur in isolation or hemorrhage
- Why these cerebellar tubers enhance has not been elucidated
- It is important to recognize typical-appearing cerebellar tubers to avoid unnecessary surgery

Discussion



At 20 months of age



At 4 years of age
The tuber is increased in size



At 10 years of age
The tuber is decreased in size with increased
folia retraction/atrophy

History

Images

DDx

Diagnosis

Discussion

References

References

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- Vaughn J, Hagiwara M, Katz J, Roth J, Devinsky O, Weiner H, Milla S. MRI characterization and longitudinal study of focal cerebellar lesions in a young tuberous sclerosis cohort. Am J Neuroradiol 2013 Mar;34(3):655-9. Mar;34(3):655-9.
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