50-year-old female who presents with new-onset headaches, blurry vision, and neurosensory hearing loss.

> Erica Shen, MD PhD Abner Gershon, MD Leo Wolansky, MD





# T2 FLAIR Axial



RADIOLOGY









# Gd-T1 Sagittal



### Gd-T1 Coronal













## T2 Sagittal





#### T2 Axial













# Neurofibromatosis 2





## T2 FLAIR Axial

- Isointense mass involving the sagittal sinus.
- The superior sagittal sinus is invaded and likely occluded.





- Hyperintense lesion with heterogeneous enhancement involving the sagittal sinus.
- The superior sagittal sinus is invaded and likely occluded.
- The mass bulges toward the left and displaces the left superior frontal gyrus toward the left.
- The mass has the features of a typical meningioma.





## Gd-T1 Sagittal

- Hyperintense lesion with heterogeneous enhancement involving the sagittal sinus.
- The superior sagittal sinus is invaded and likely occluded.





### **Gd-T1** Coronal

- Hyperintense lesion with heterogeneous enhancement involving the sagittal sinus.
- The superior sagittal sinus is invaded and likely occluded.





- Hyperintense lesion with avid enhancement on Gadolinium postcontrast T1.
- The lesion involves the posterior aspects of the superior sagittal sinus.





# T2 Sagittal

- Soft tissue mass extending exophytically from the inferior aspect of the left lobe of the thyroid gland.
- The mass is hyperintense on T2weighted mages.





### T2 Axial

- A soft tissue mass (orange arrow) extending exophytically from the inferior aspect of the left lobe of the thyroid gland.
- The mass is hyperintense on T2weighted mages.
- Another lesion (green arrow) with sharp margin adjacent to the left side of the thoracic spine centered at the level of the T1 to T2 intervertebral disc. The mass extends slightly into the left T1 to T2 neural foramen.



#### Gd-T • A lesion ( adjacent to spine cen interverte

- A lesion (green arrow) with sharp margin adjacent to the left side of the thoracic spine centered at the level of the T1 to T2 intervertebral disc. The mass extends slightly into the left T1 to T2 neural foramen.
- The mass enhances diffusely following administration of gadolinium-based contrast agent.



# Neurofibromatosis 2

• Hereditary tumor syndrome characterized predominantly by the development of schwannomas, along with meningiomas, ependymomas, and ocular abnormalities.

#### • Autosomal dominant pattern:

- Diverse phenotypic variability.
- 50% of cases resulted from sporadic mutations.
- 100% penetrance by age 60.
- Incidence: 1/25,000 births.
- Prevalence: 1/100,000-200,000 persons.
- Mutation in the NF2 gene on chromosome 22.
- Mutation in a protein, called merlin.



Kresak, J.L., Walsh, M. J., Pediatr Genet., 2016.

# **Clinical Diagnostic criteria**

MAJOR:

(1) Bilateral vestibular schwannomas.

(2) First-degree relative with Neurofibromatosis Type 2 PLUS

#### Unilateral vestibular schwannomas OR

 Any TWO of the following: meningioma, glioma, schwannoma, or ocular abnormalities.



# **Clinical Diagnostic criteria**

#### ADDITIONAL:

(1) Unilateral vestibular schwannoma PLUS

• Any TWO of the following: meningioma, glioma, schwannoma, ocular abnormalities.

(2) At least two meningiomas <u>PLUS</u>

- Unilateral vestibular schwannomas OR
- Any TWO of the following: meningioma, glioma, schwannoma, or ocular abnormalities.



# References

- Dubey, P., et al. Radiology, a core review. Philadelphia: Wolters Kluwer. 2018.
- Harbaugh, R.E., Shaffrey, C., Couldwell, W.T., Berger, M.S. Neurosurgery Knowledge Update, a comprehensive review. New York, Thieme, 2015.
- Louis, D.L., Ohgaki, J., Wistler, O.D., Cavenee WK. WHO Classification of Tumors of the Central Nervous System. Lyon, France: World Health Organization. International Agency for Research on Cancer; 2007.
- Kresak, J.L., Walsh, M. Neurofibromatosis: a review of NF1, NF2, and Schwannomatosis. J. Pediatr Genet 2016; 5: 98-104.
- https://radiopaedia.org
- Shen, E., Gershon, A., Wolansky, L. Neurofibromatosis 2. Radiology Online, 2021.

