

# 與大師對談 2019.05.21

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# 規則介紹

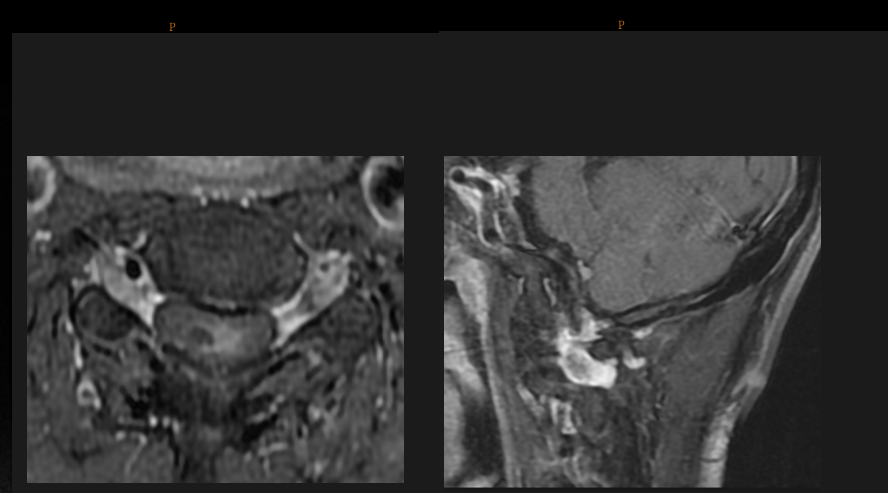
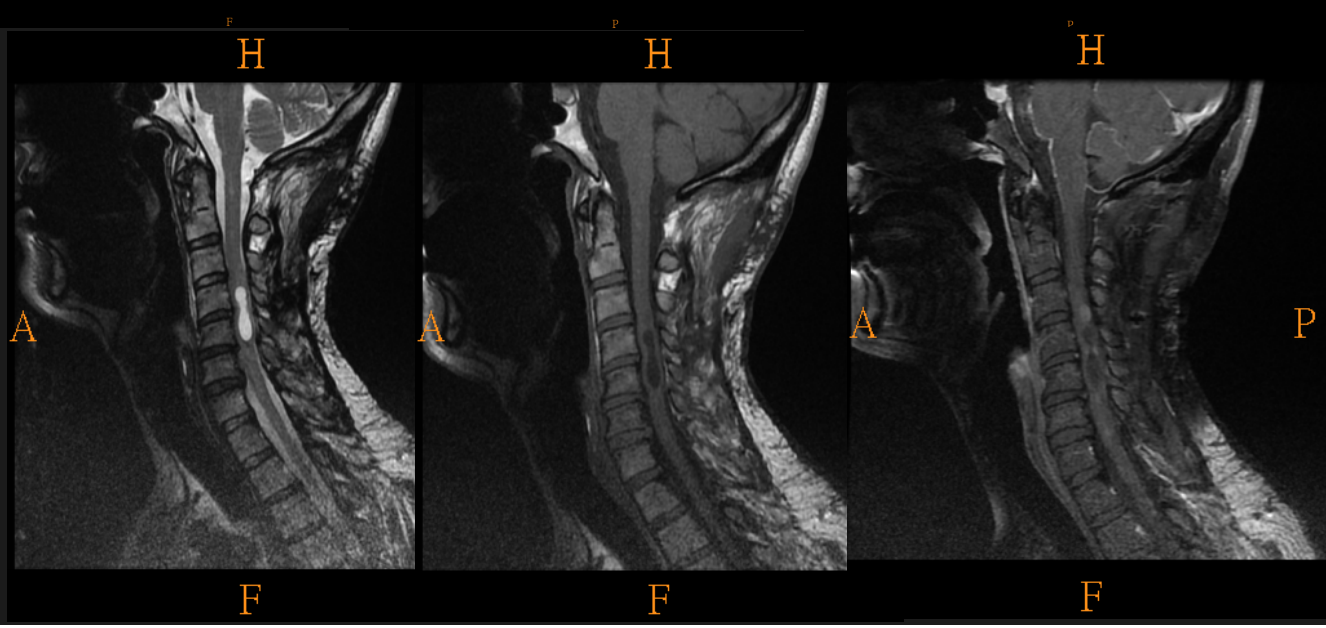
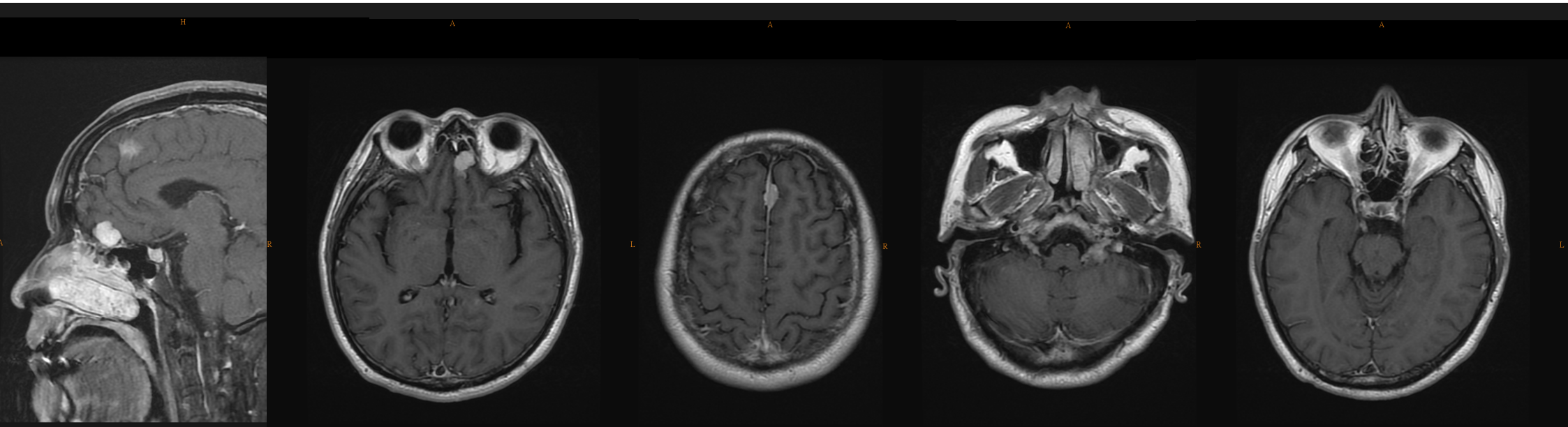
- 依照臨床時序，請大師模擬一線放射科醫師；於未知診斷，或者有限度臨床線索之情形下，進行閱片及解讀。
- 鑑別診斷為主要，確定診斷為次要。
- 目的在於學習大師之影像判讀邏輯思考。
- 針對神經影像判讀。
- 大師評論本院影像品質建議及改進。

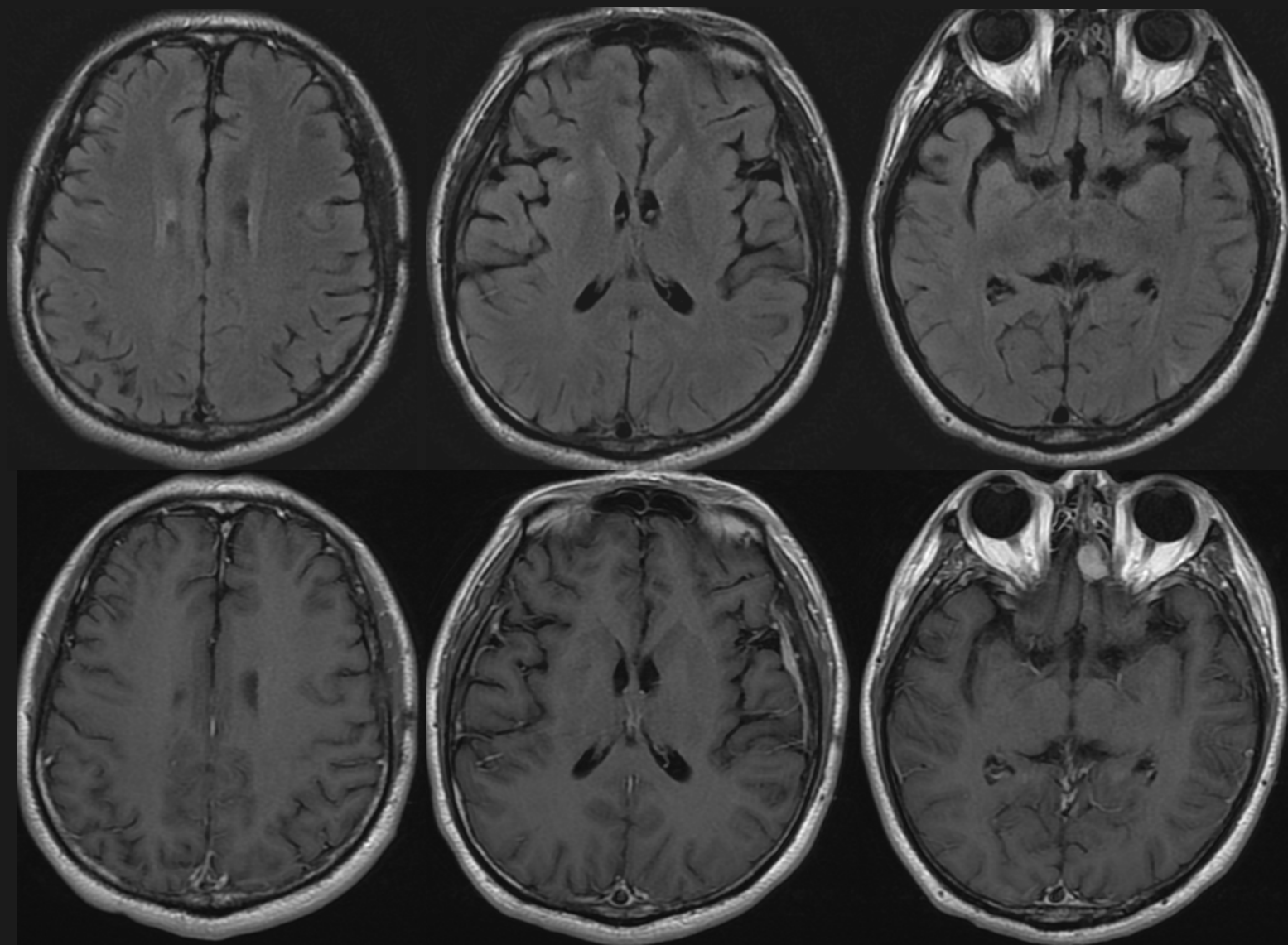
# CASE 1

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# 09508855

- 33 y/o, man
- Hearing impairment on the right ear
- Mild ptosis on the left eye
- Numbness and clumsiness of bilateral hands and intermittent twisting of the left hand



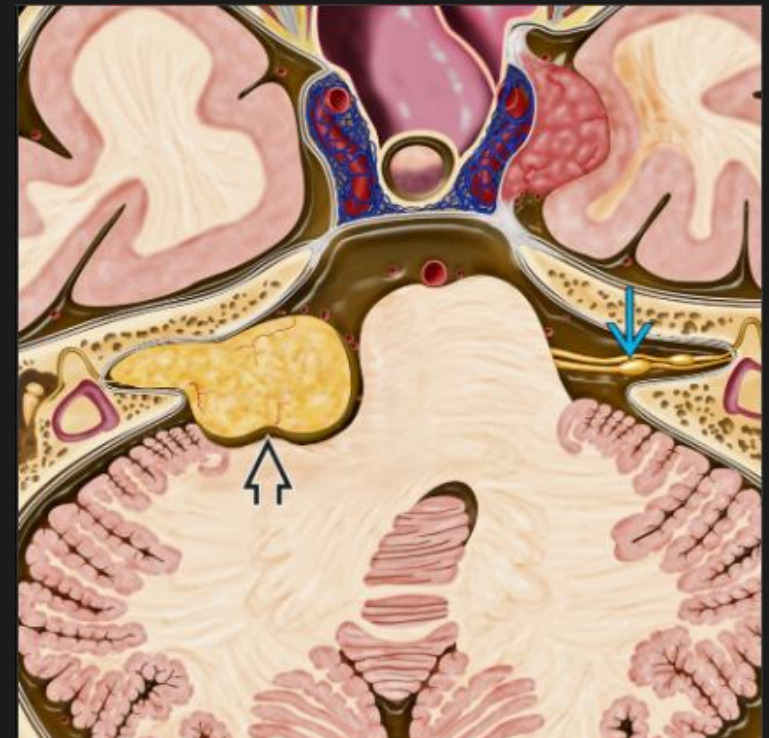


# Diagnosis

- Multiple intracranial meningiomas, schwannomas and ependymoma with multiple glial micro-hamartomas = Neurofibromatosis type 2

# Neurofibromatosis, type 2

- Terminology
  - Familial cancer syndrome (22q12, NF2 gene)
  - Acoustic neurofibromatosis, central neurofibromatosis
  - Multiple intracranial schwannomas, meningiomas, and ependymomas (**MISME**)
- Imaging
  - Multiple extra-axial tumors
    - Schwannomas of CNs and spinal nerve roots: **Bilateral vestibular schwannomas**
    - **Meningiomas** on dural surfaces (up to 50%)
  - Intra-axial tumors
    - **Ependymomas** in spinal cord and brainstem (6%)
- Symptoms
  - Usually presents between 2nd and 4th decades with **hearing loss**,  $\pm$  vertigo

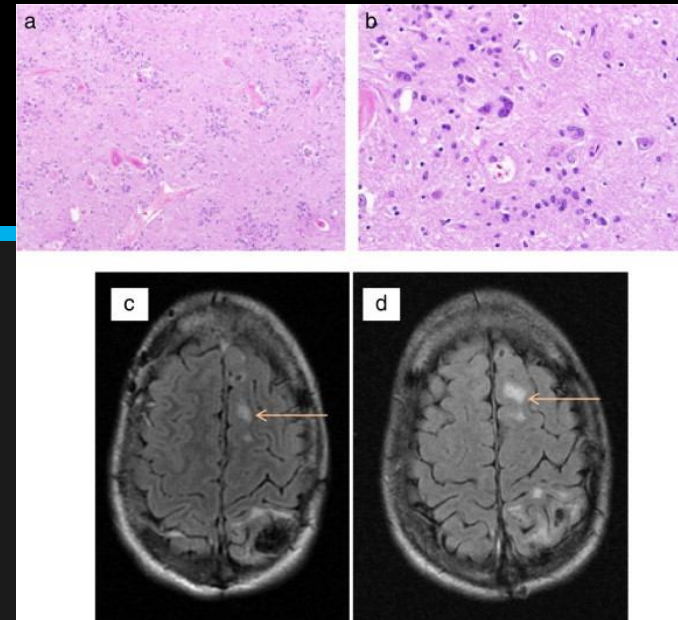




# Glial micro-hamartomas

## Pathology

- the tightly regulated expression of “merlin” during migration of cells to the cortical plate might predict cortical abnormalities in patients with NF-2 gene mutations
- foci of dysplastic, immature neuroectodermal cells within the cerebral cortex and deep gray matter



Patient demographics.	
Number of patients in the study (N)	34
Median age at time of first incidental MRI finding (years)	20
Female, no. (%)	11 (32)
Male, no. (%)	23 (68)
Received cranial irradiation, no. (%)	3 (9)

Incidental brain MRI finding	Number of patients with finding, no. (%)
Any incidental finding	23 (68)
Nonspecific T2 hyperintensities	17 (74)
Wedge shaped cortical/subcortical T2 hyperintense lesions	15 (65)
Transmantle sign	9 (39)
Cortical T2 hypointensities	8 (34)
Well circumscribed cortical T2 hypointensity with associated T1 hypointensity	6 (26)
Migrational cerebellar anomalies	3 (13)
T2 hyperintense lesions associated with an enlarged Virchow–Robin space	7 (30)
More than one of the above abnormalities	15 (65)