

症例提示

愛媛県立中央病院 放射線科

小川遼 三木均

Case: 76-year-old man

【Chief complaint】 Palatal tremor, cerebellum malfunction

【Present illness】 He was introduced to neurology due to symptom of the cerebellum malfunction and palatal tremor. He complicated the brainstem symptom (MLF syndrome).

【Past medical history】 After surgery for esophageal cancer

【Family history】 Nothing particular

【 Examination of blood】

WBC $6.06 \times 10^3/\mu\text{l}$, RBC $3.25 \times 10^6/\mu\text{l}$, HGB 9.8mg/dl, PLT $27.7 \times 10^4/\mu\text{l}$
Alb 3.5g/dl, T.bil 0.3mg/dl, BUN 18.6mg/dl, Cre 0.88mg/dl
Na 138mEq/l, K 4.8mEq/l, Cl 104mEq/l, CRP 0.16mg/dl

<画像供覧>

頭部単純MRI(神経内科紹介時)

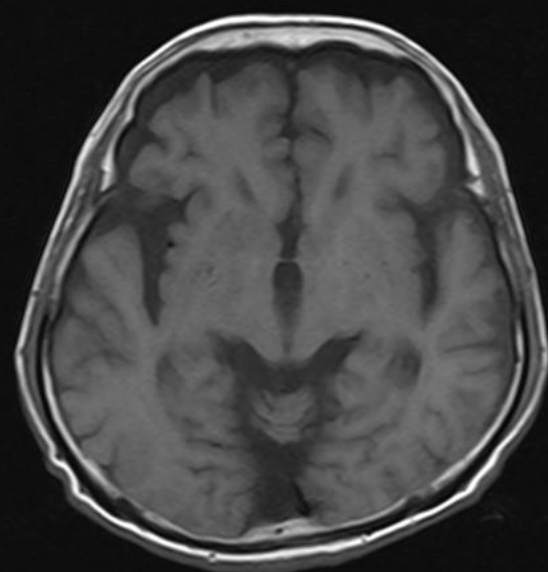
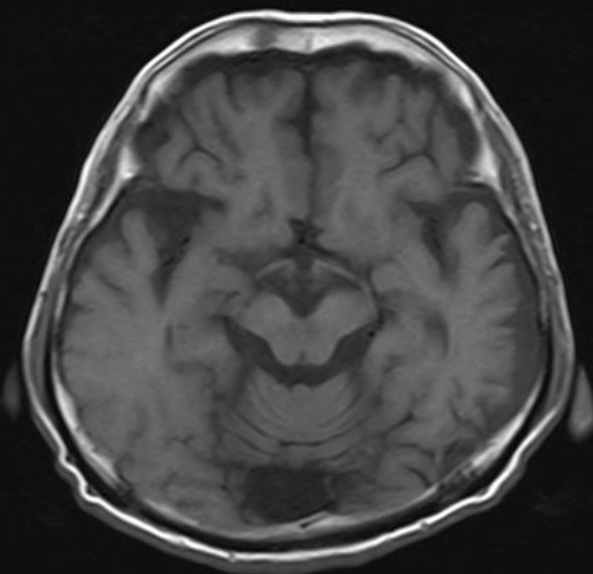
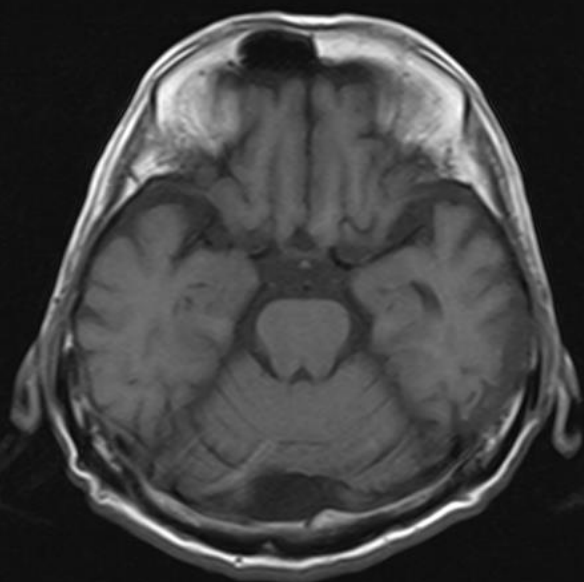
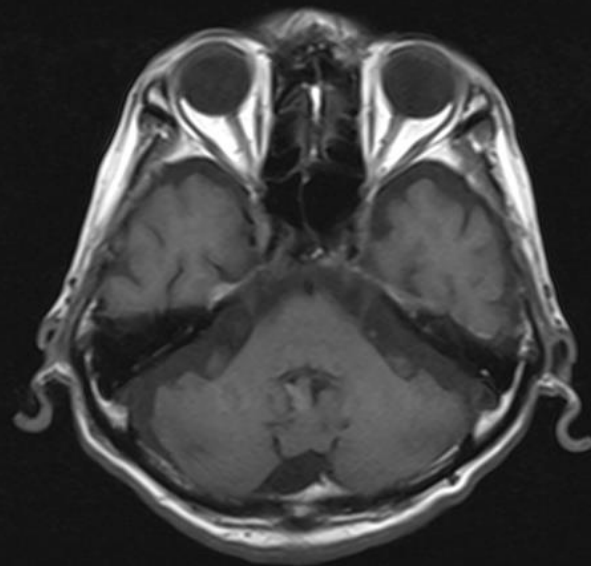
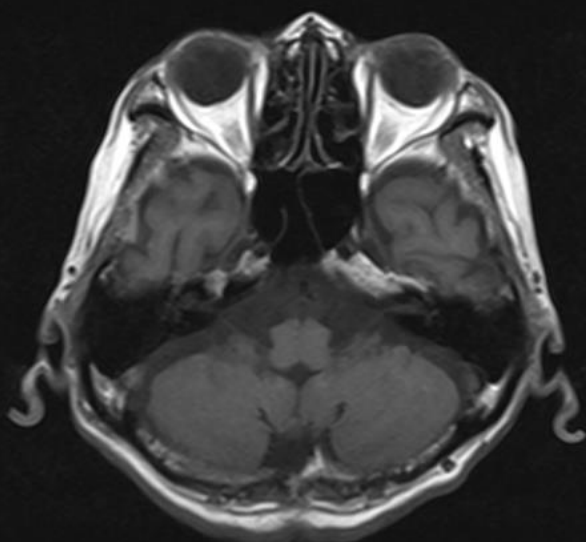
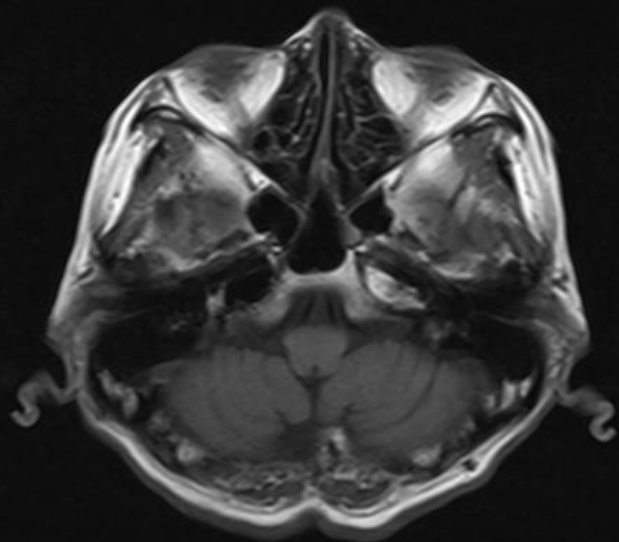
T1WI T2WI T2FLAIR

頭部単純MRI(紹介後約10ヶ月後)

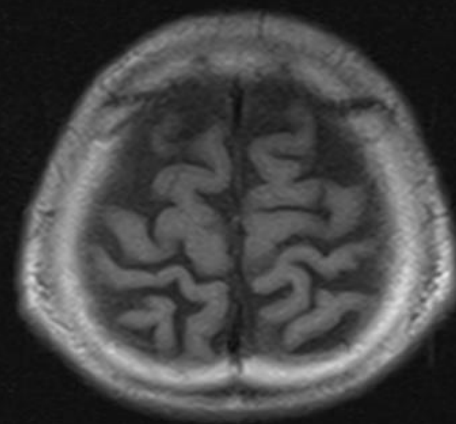
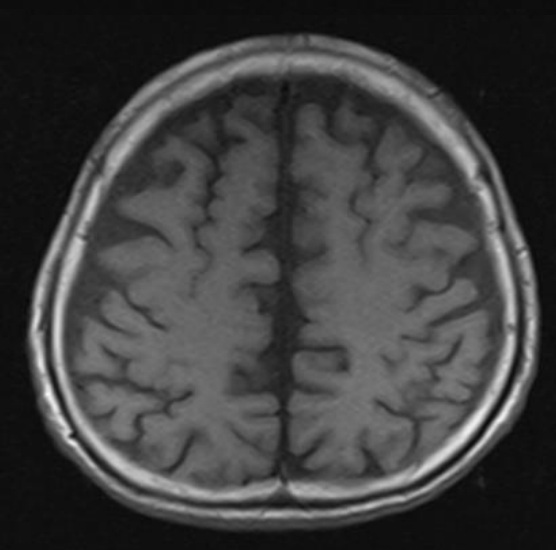
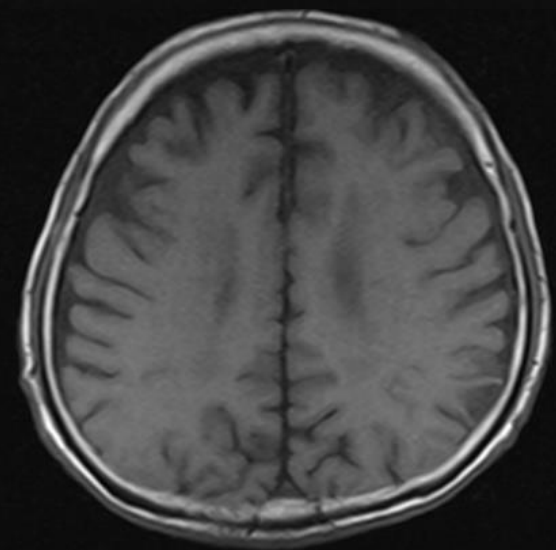
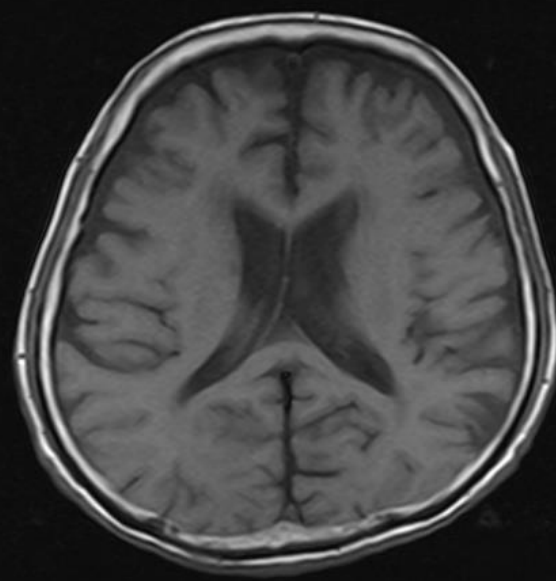
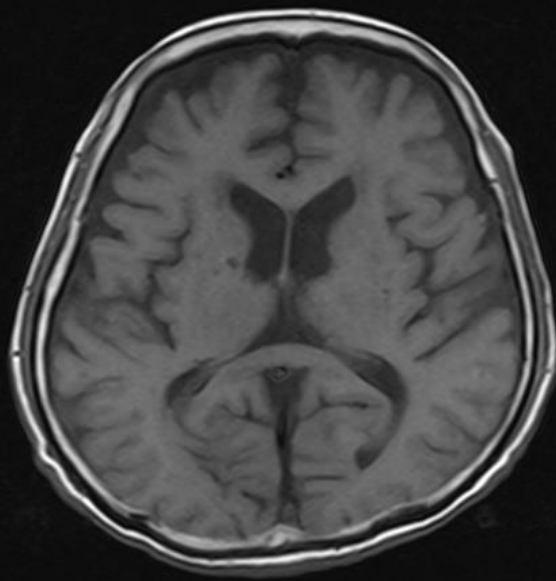
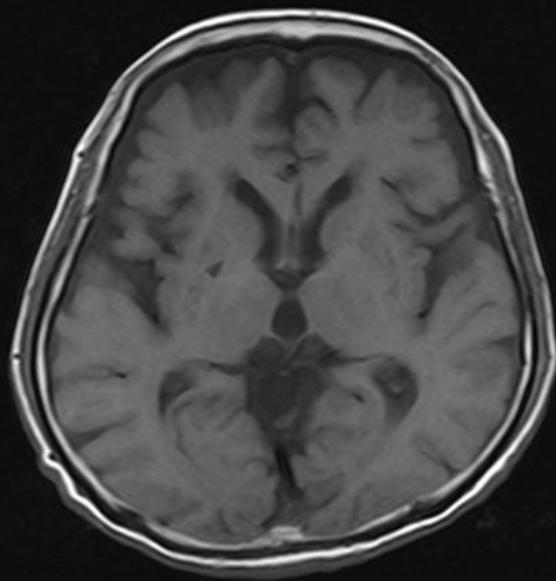
T1FLAIR T2WI T2FLAIR

診断は？

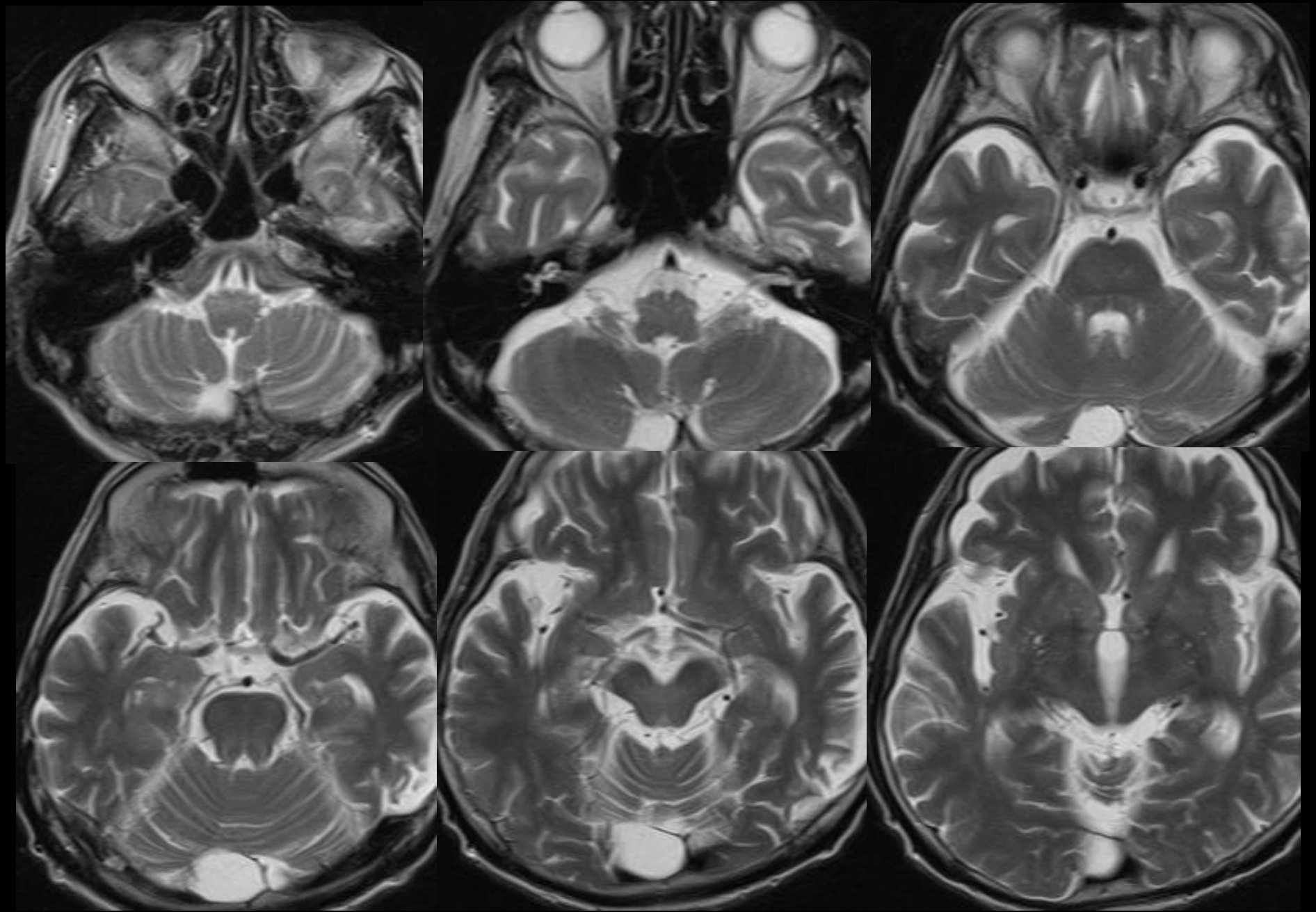
神経内科紹介時T1WI



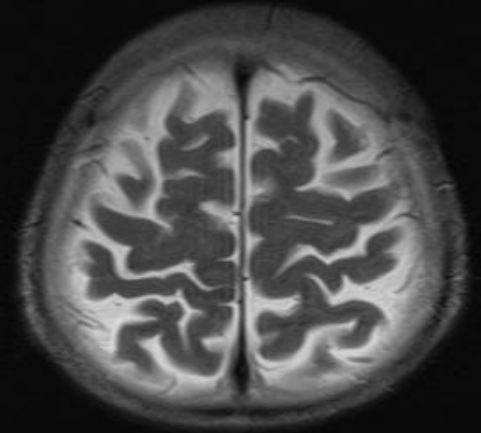
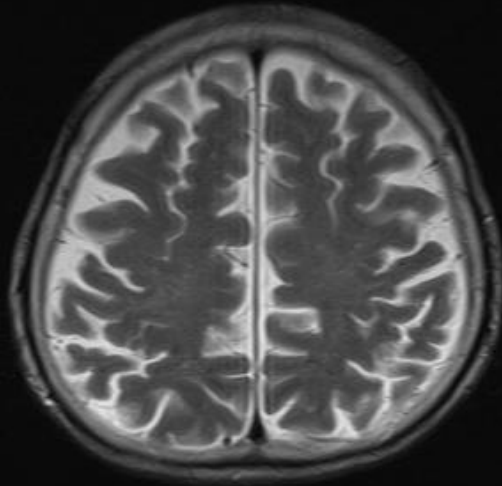
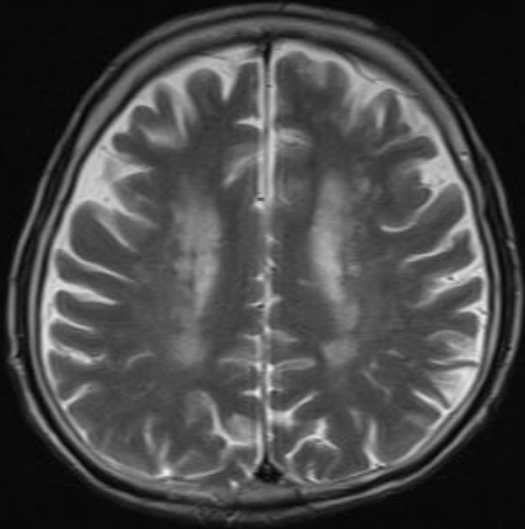
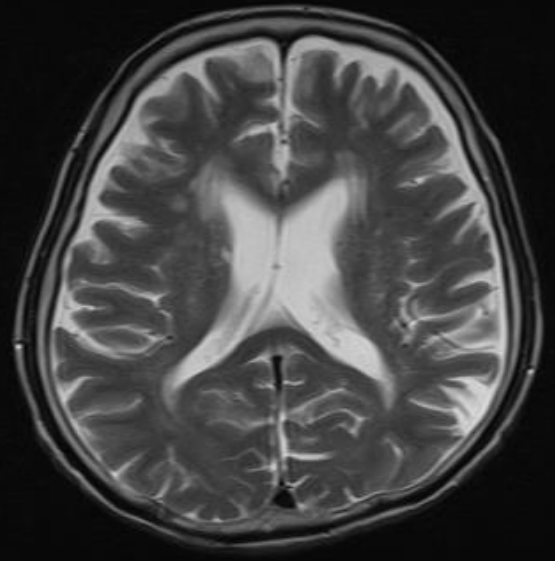
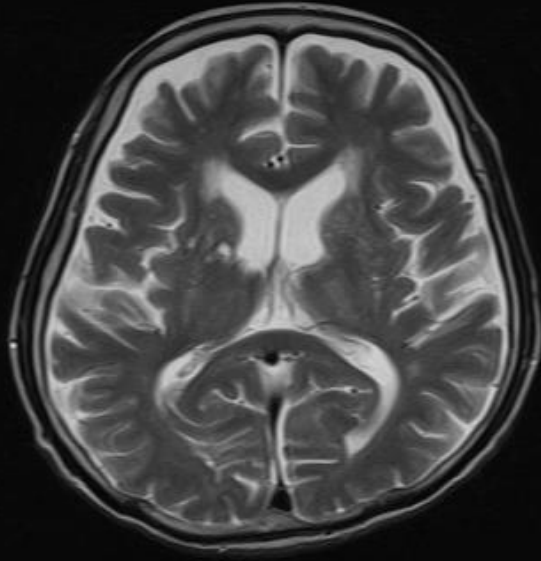
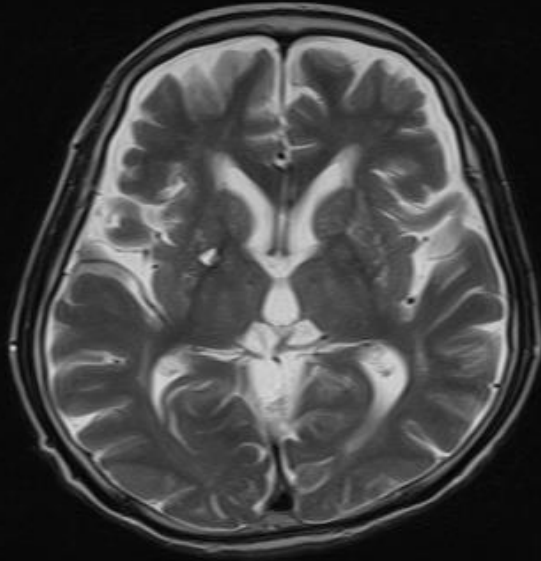
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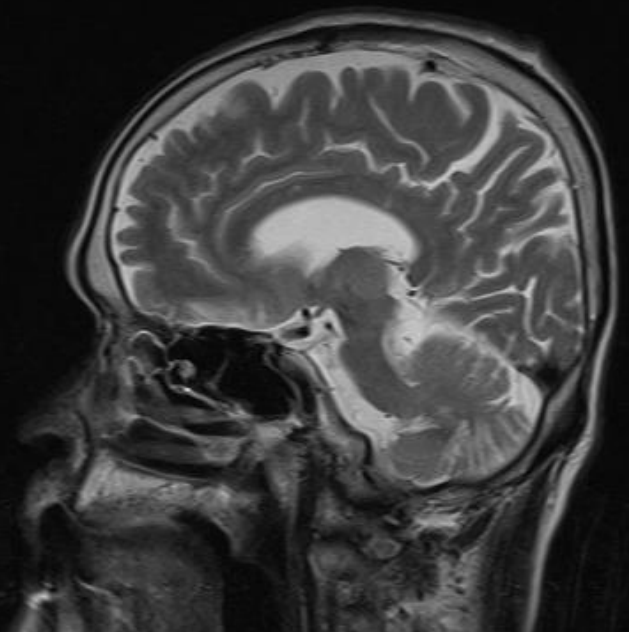
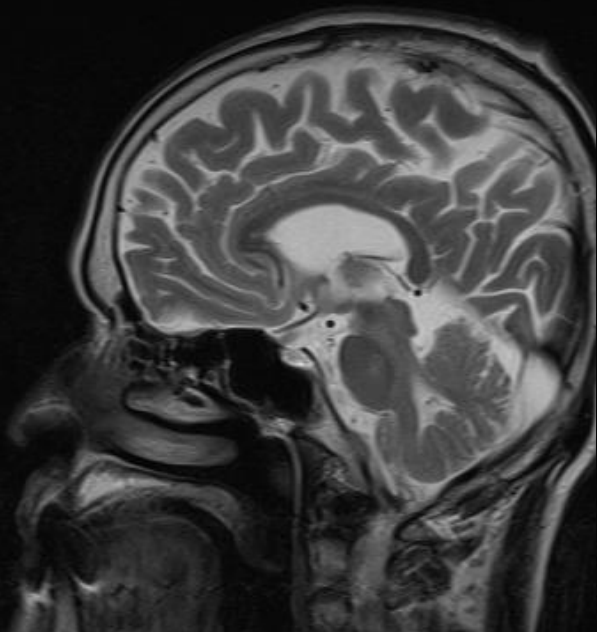
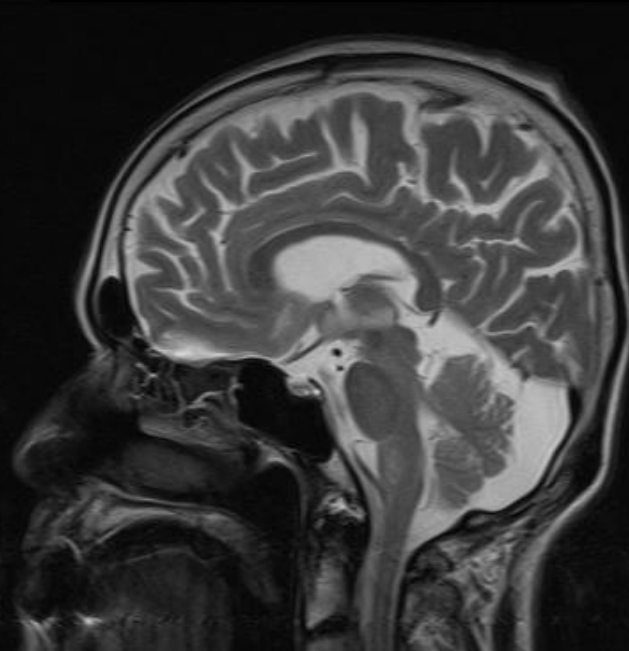
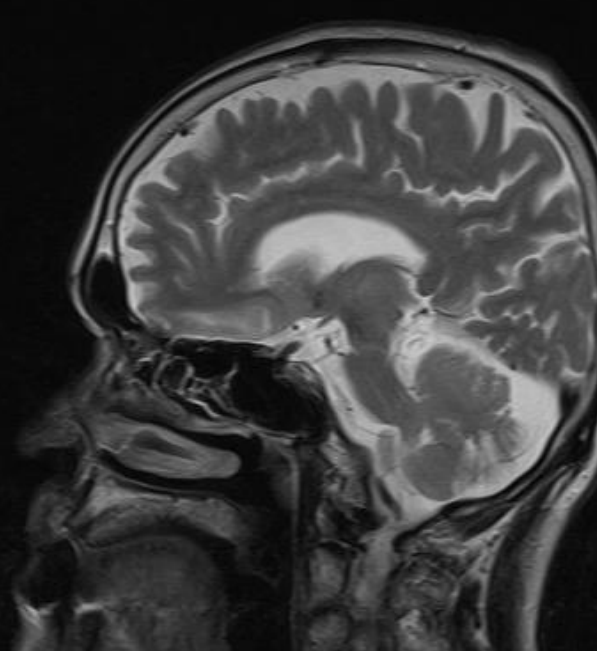
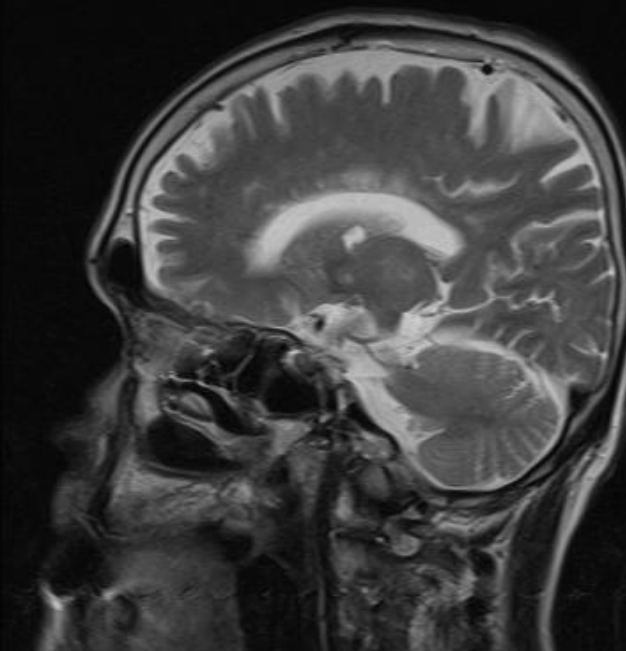
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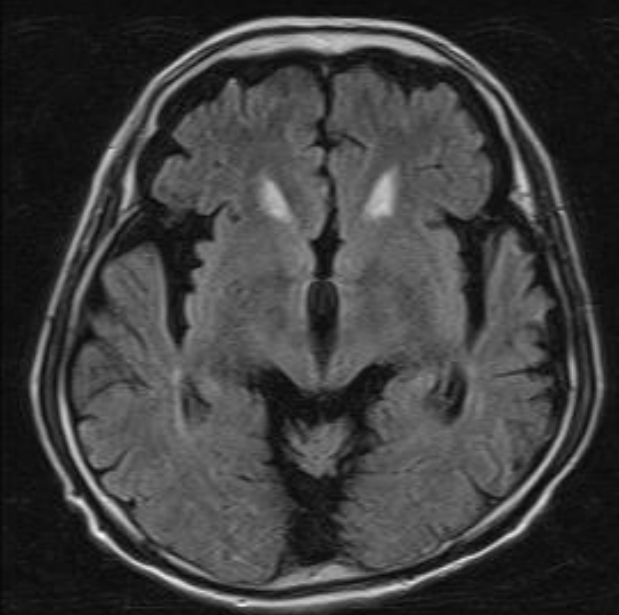
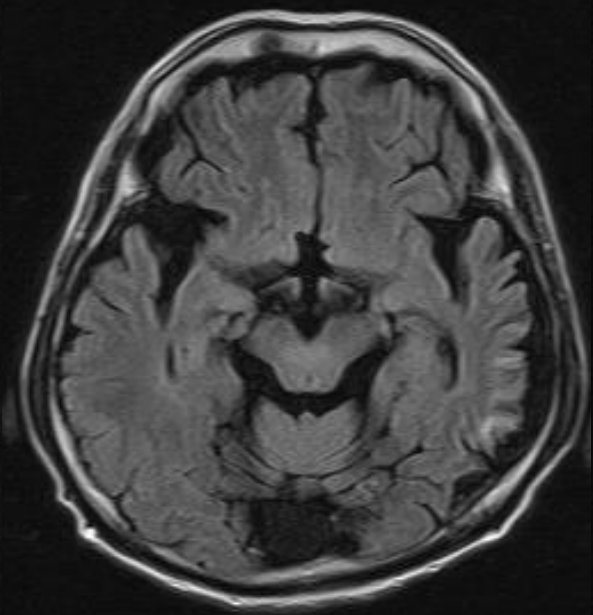
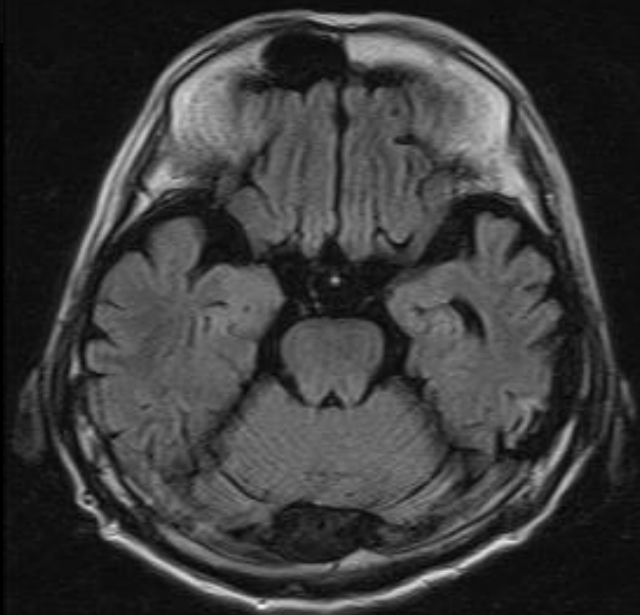
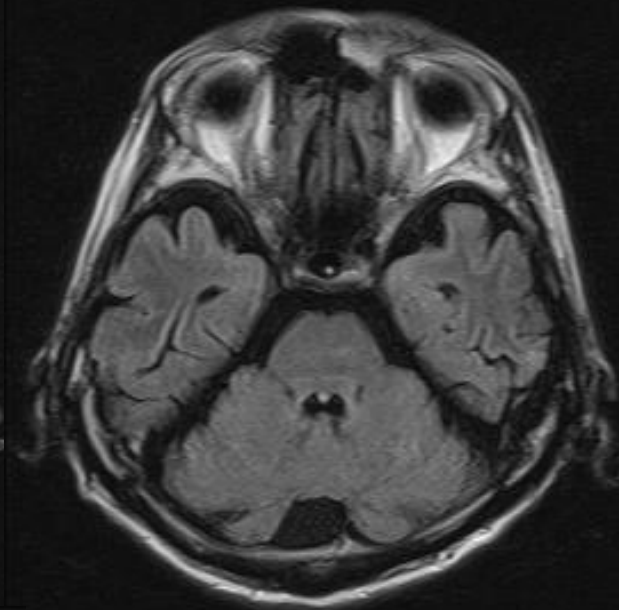
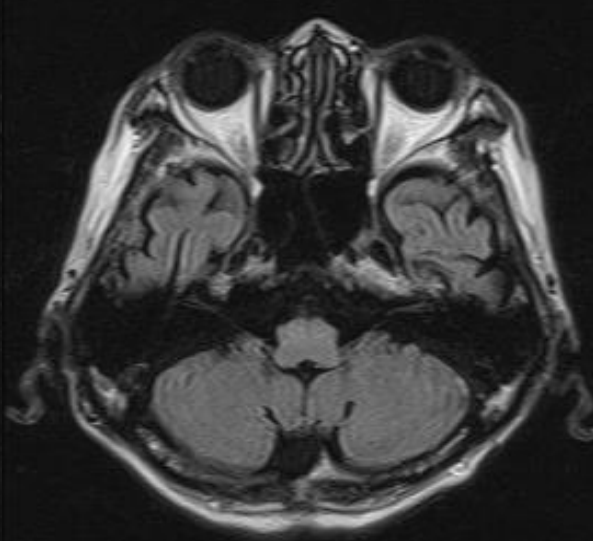
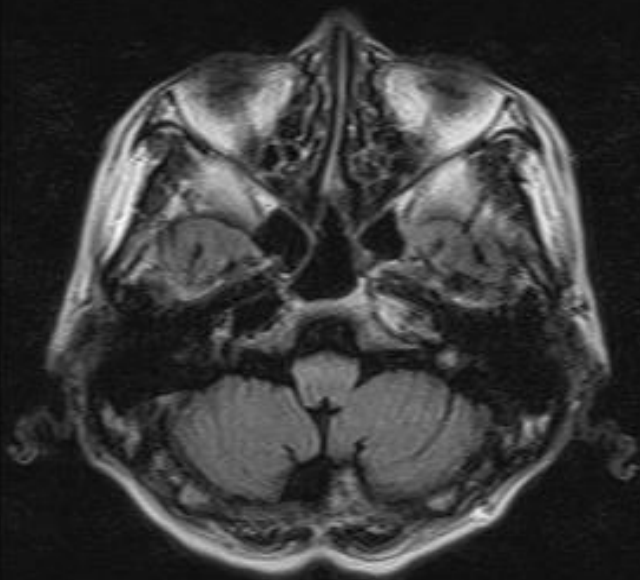
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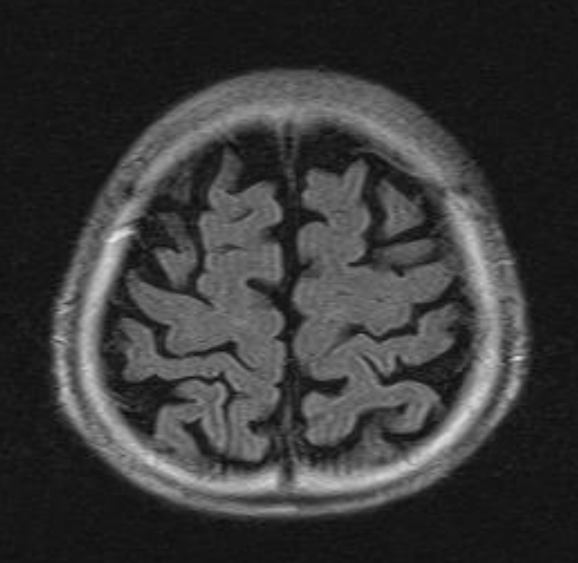
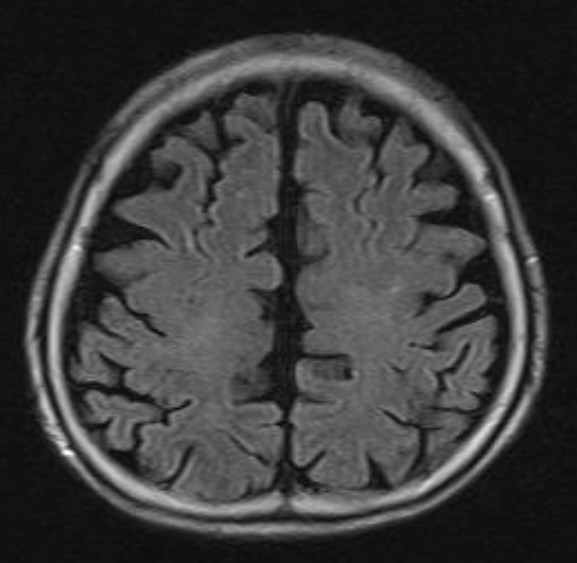
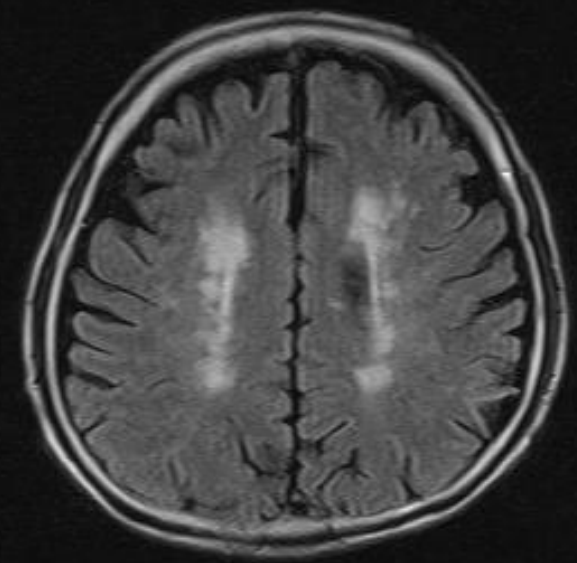
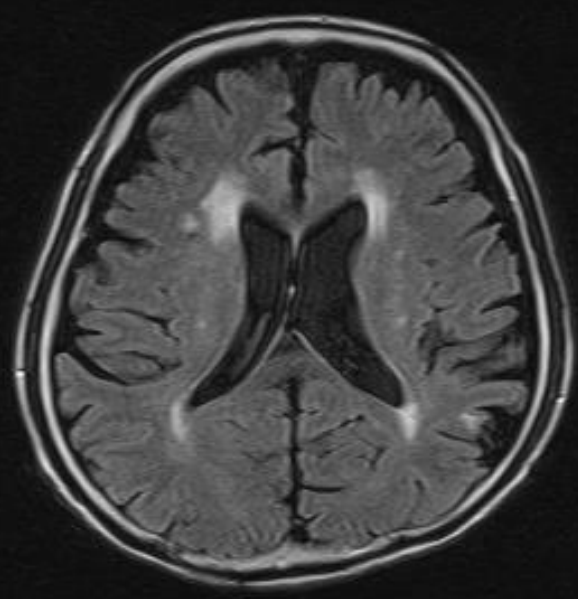
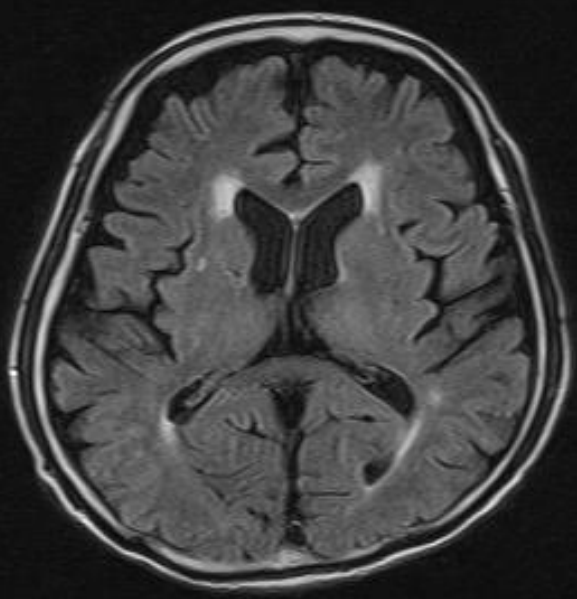
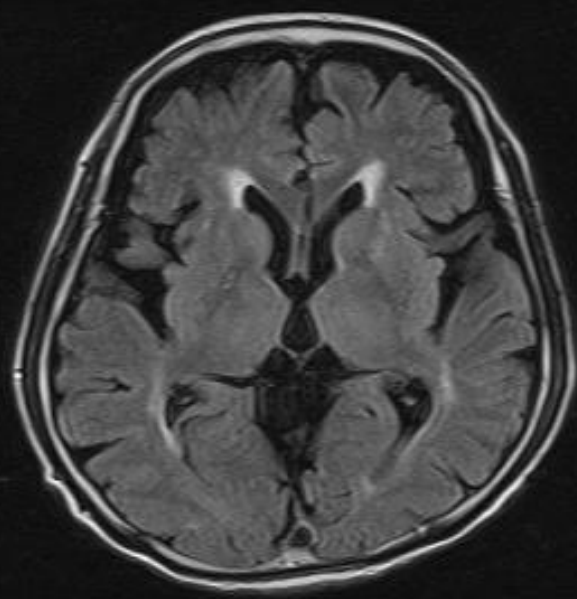
神経内科紹介時T2WI 矢状断



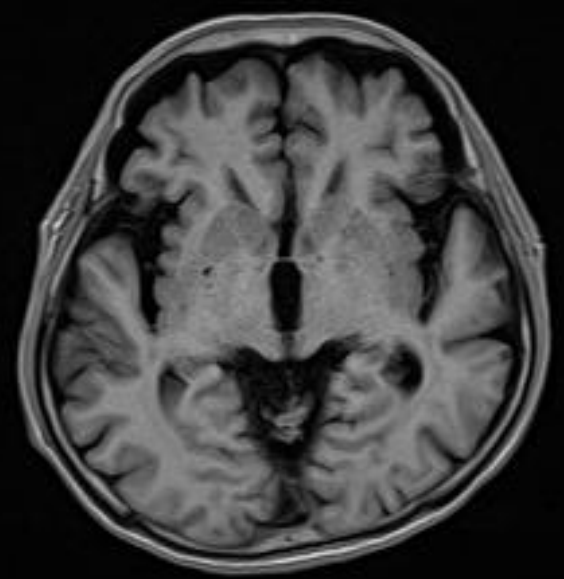
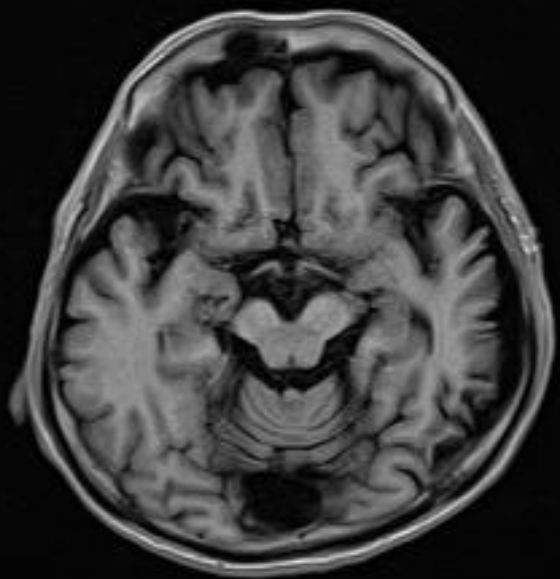
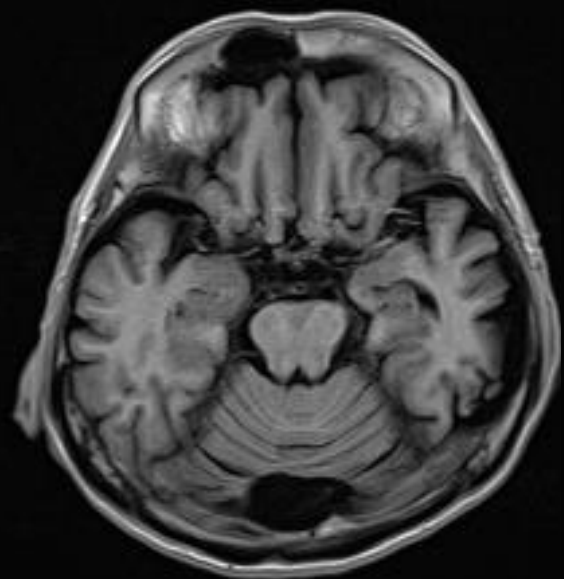
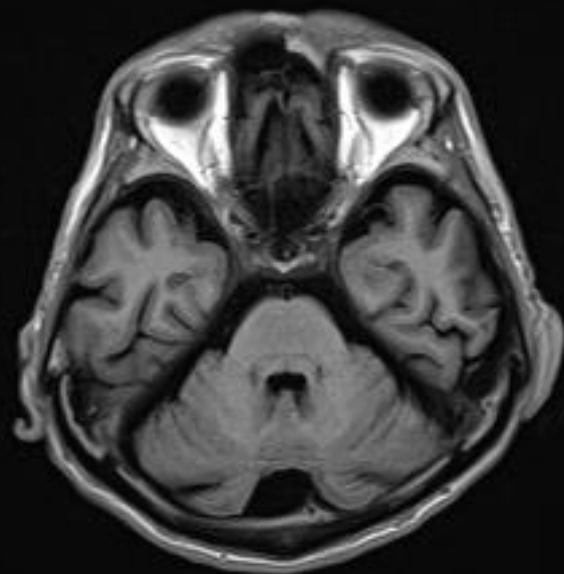
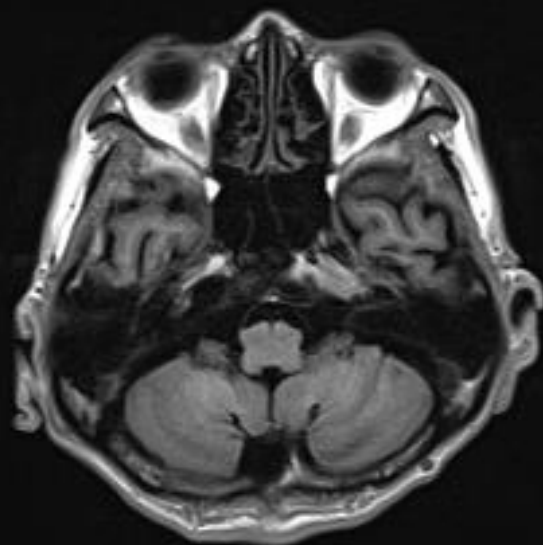
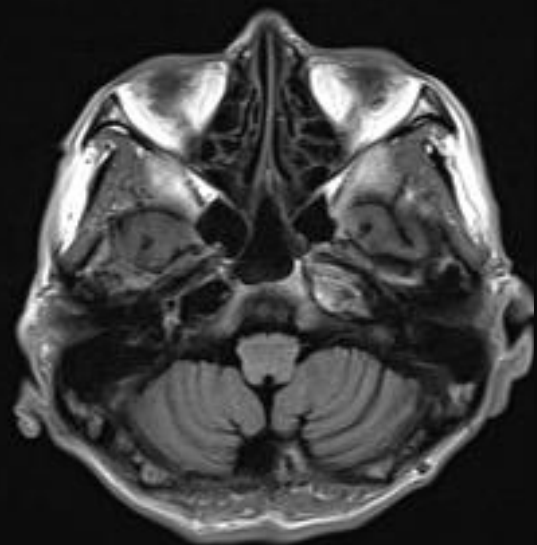
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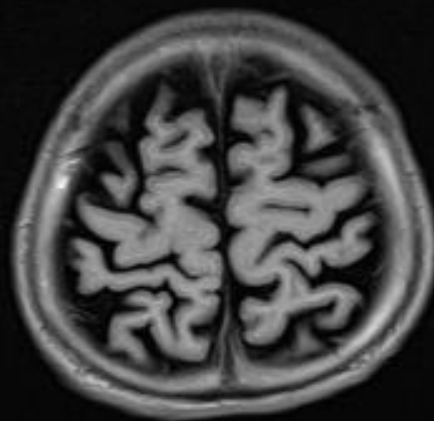
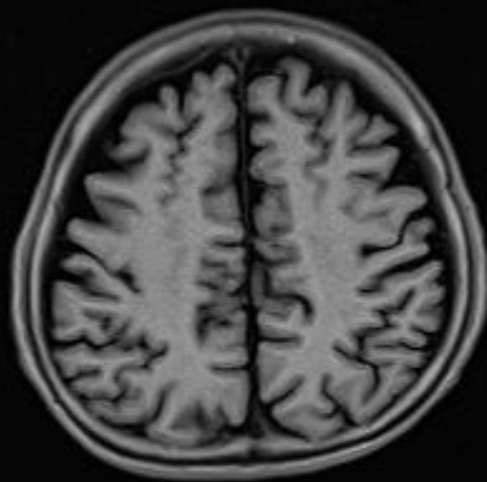
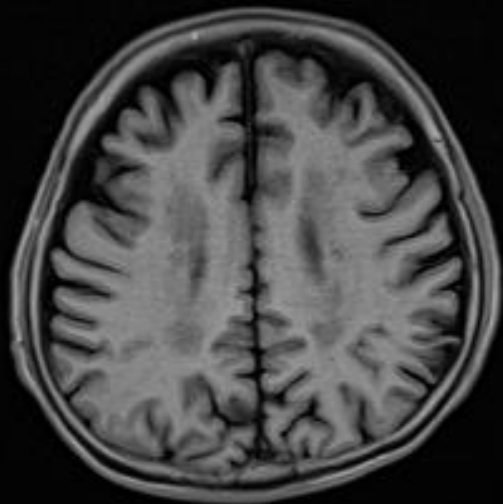
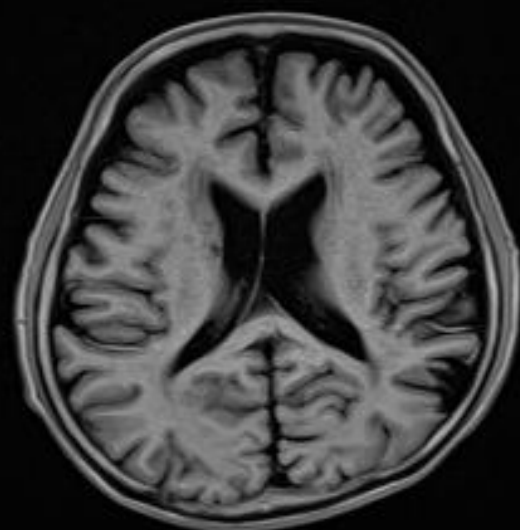
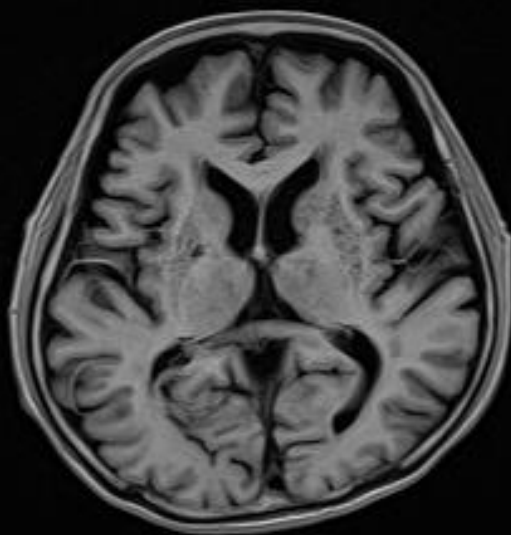
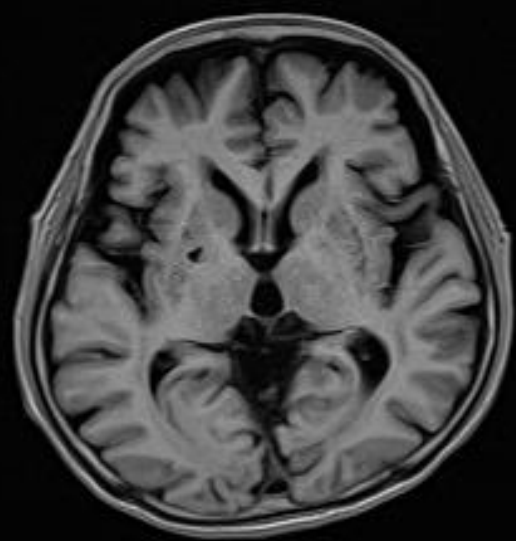
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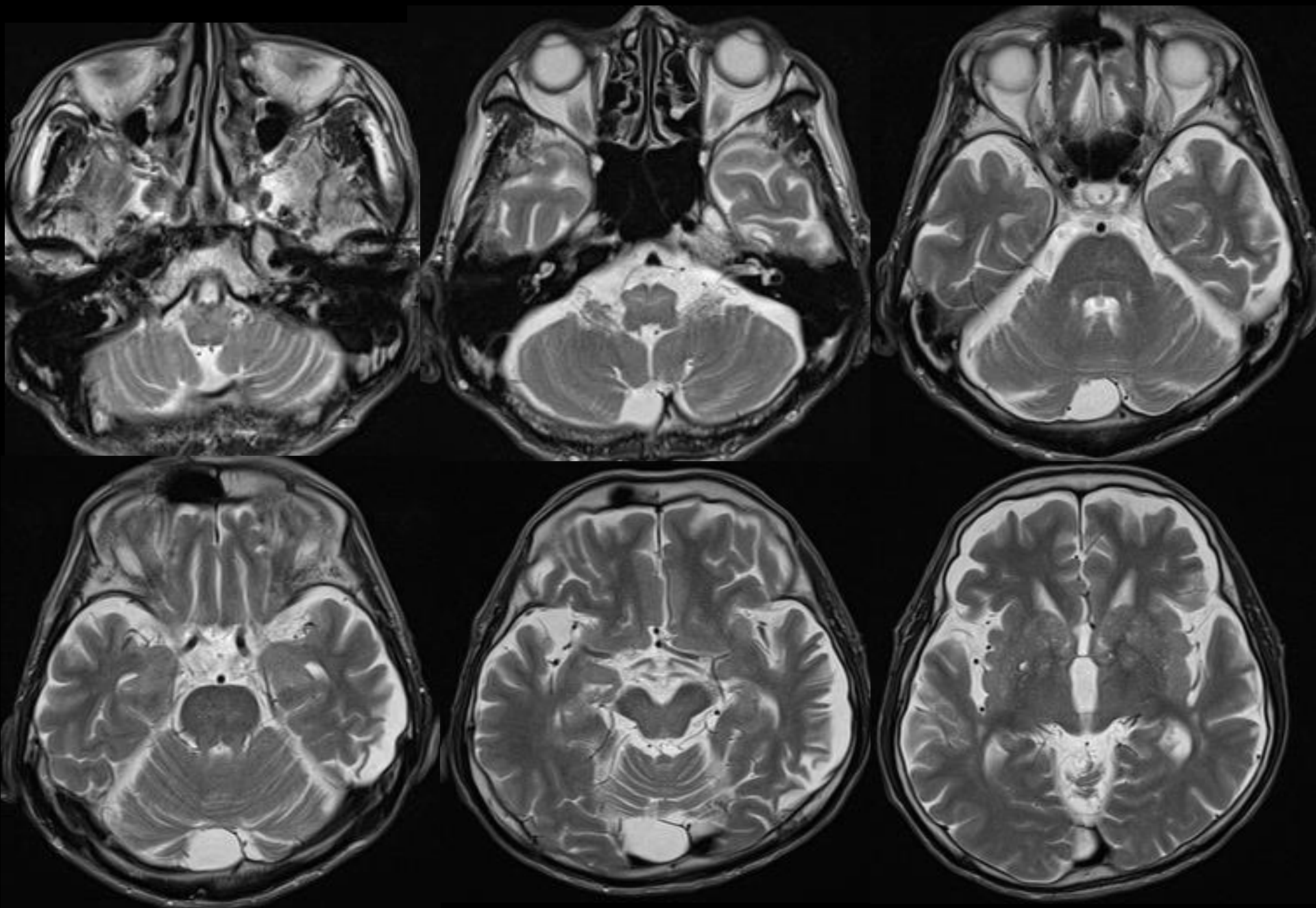
神経内科紹介約10ヵ月後T1FLAIR



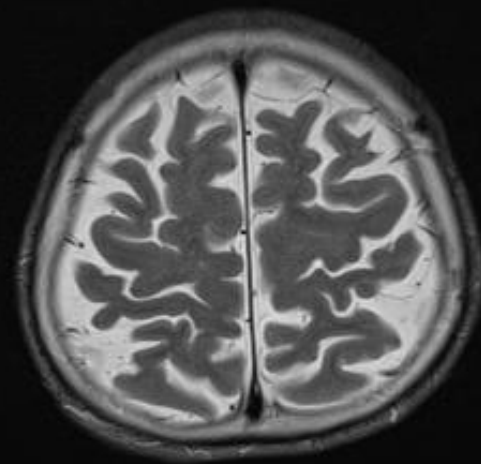
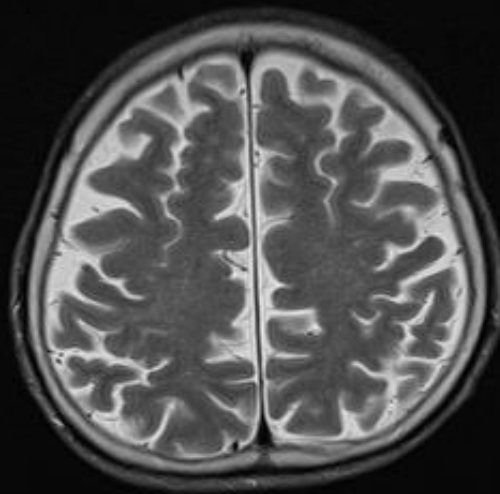
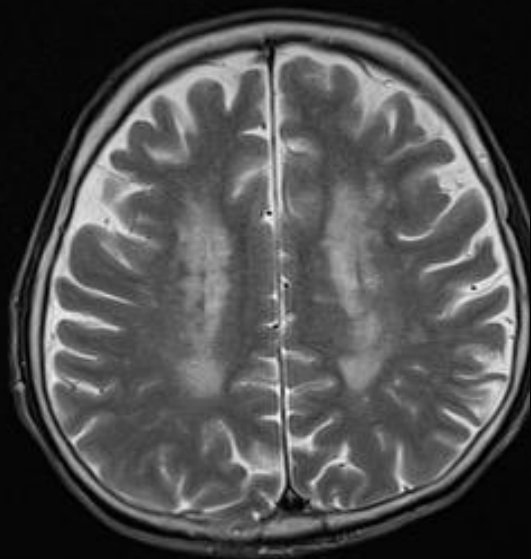
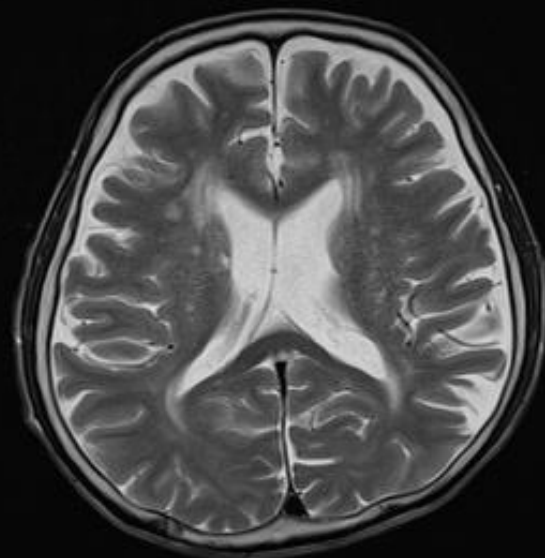
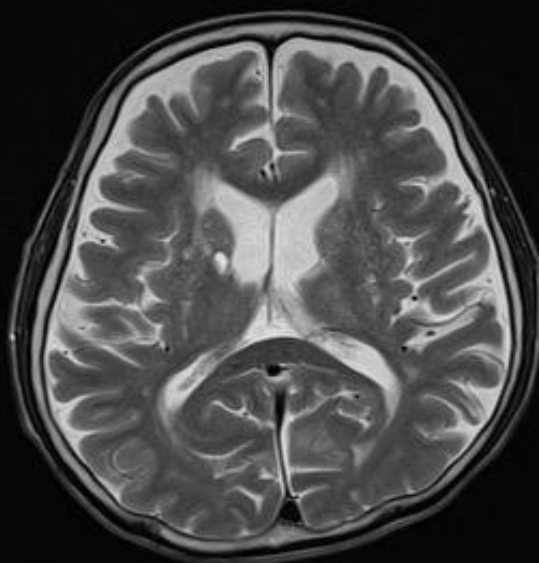
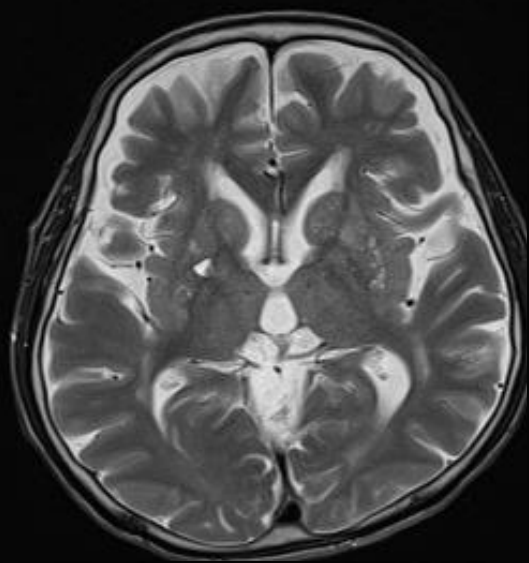
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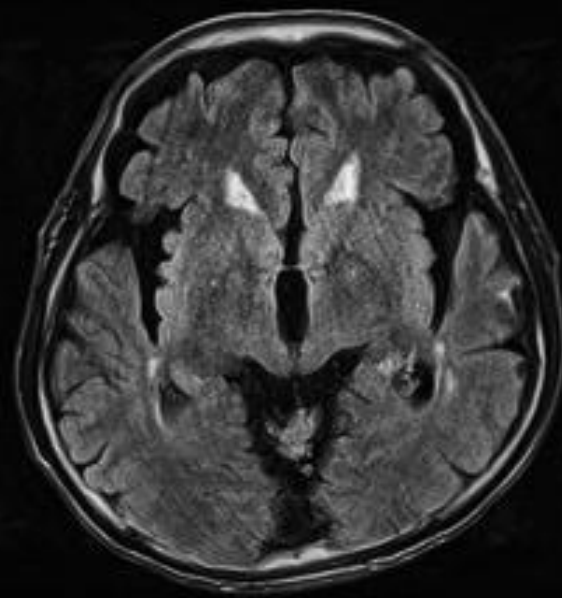
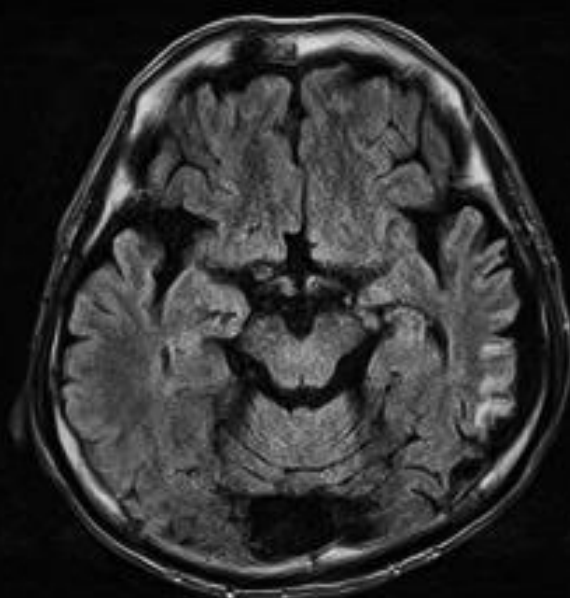
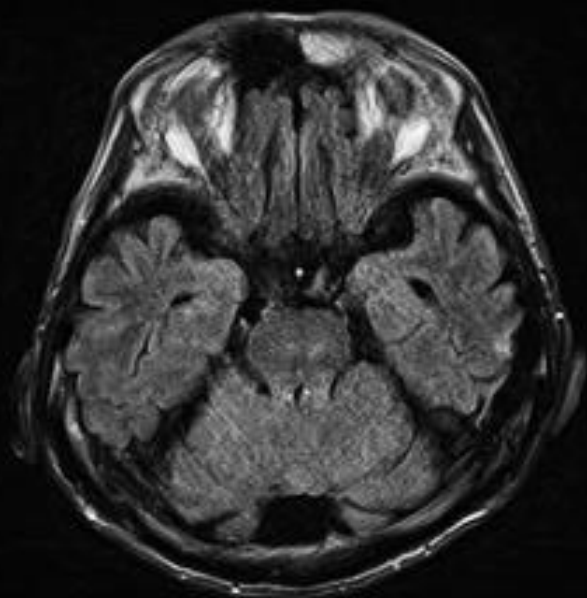
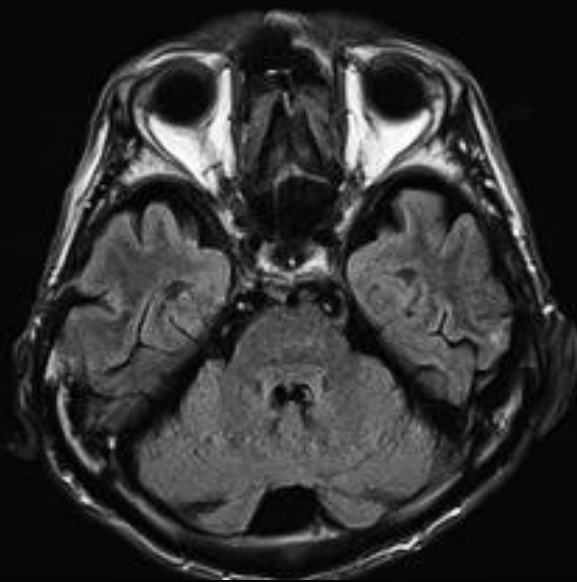
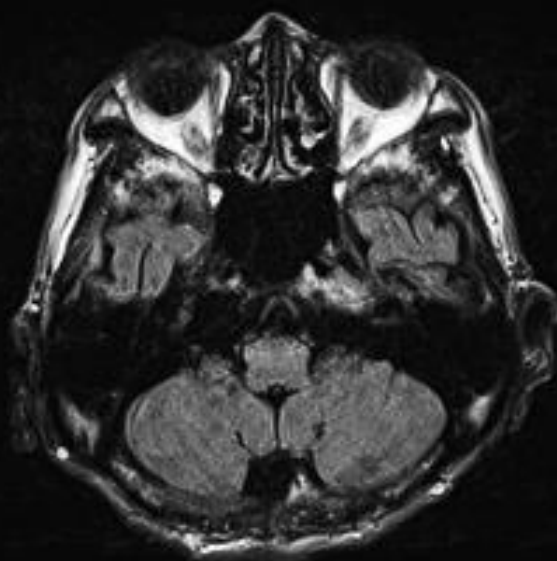
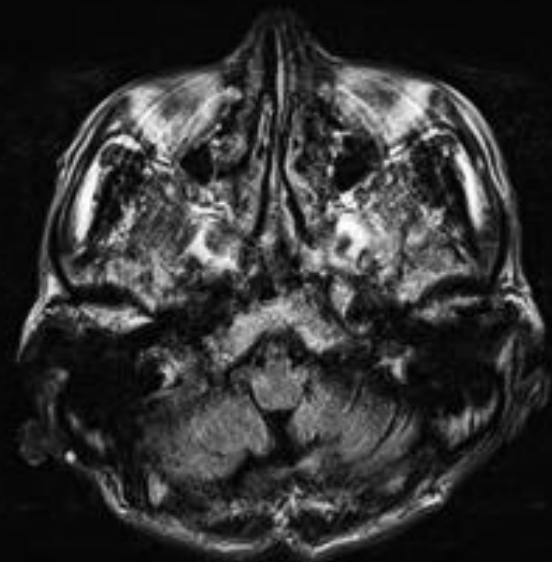
神経内科紹介約10ヵ月後T2WI



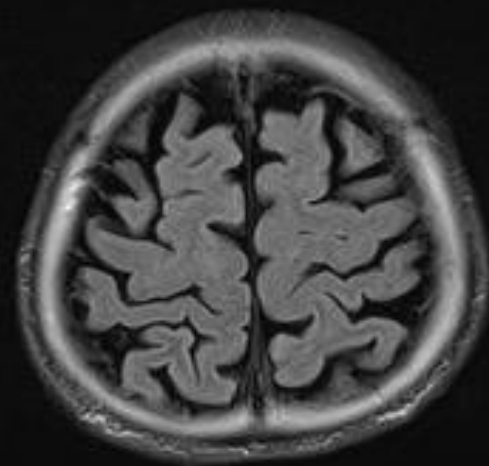
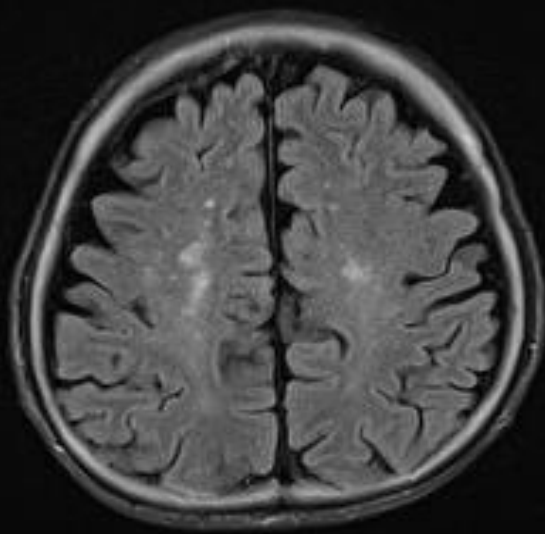
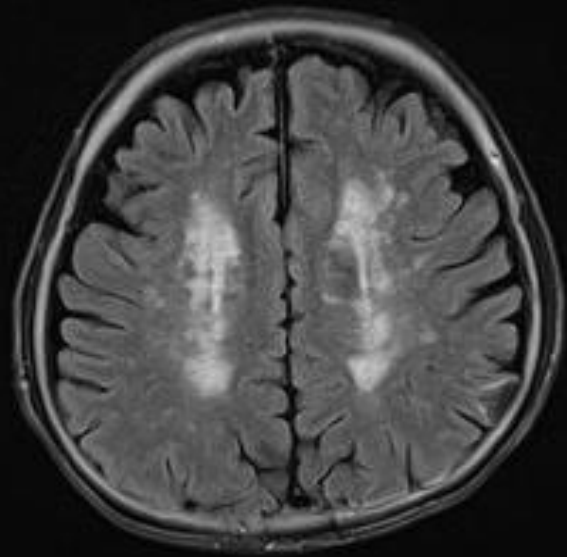
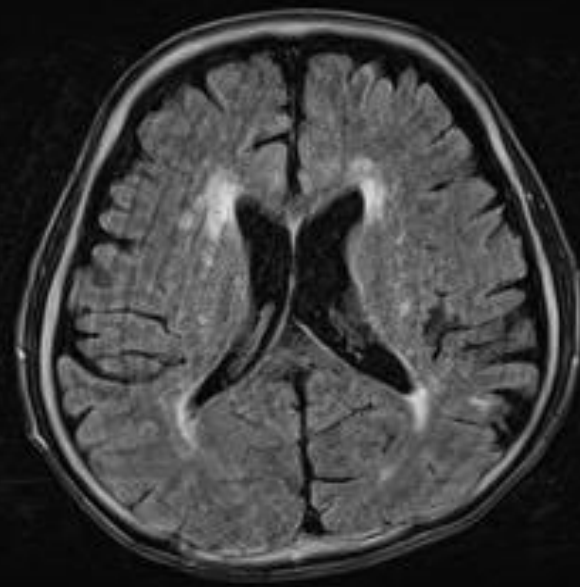
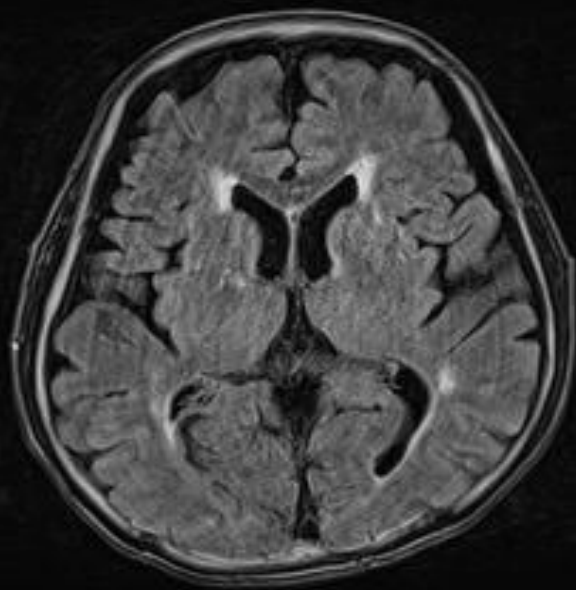
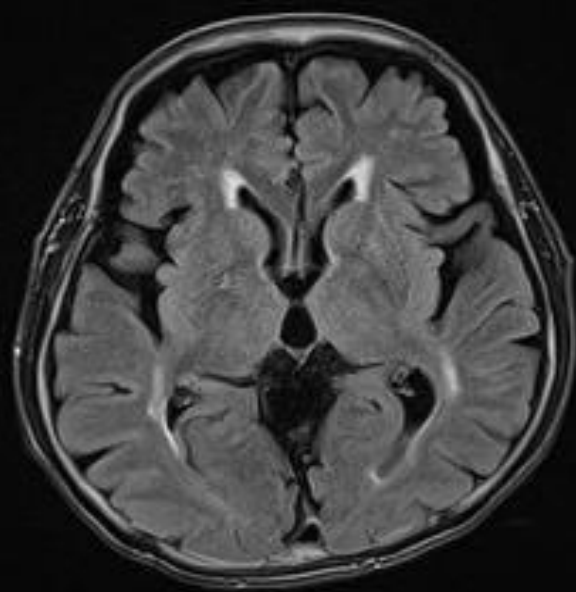
神経内科紹介約10ヵ月後T2WI



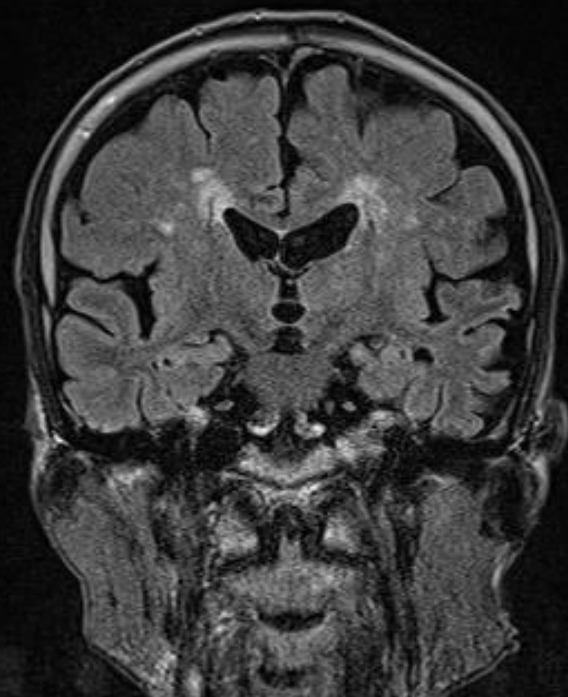
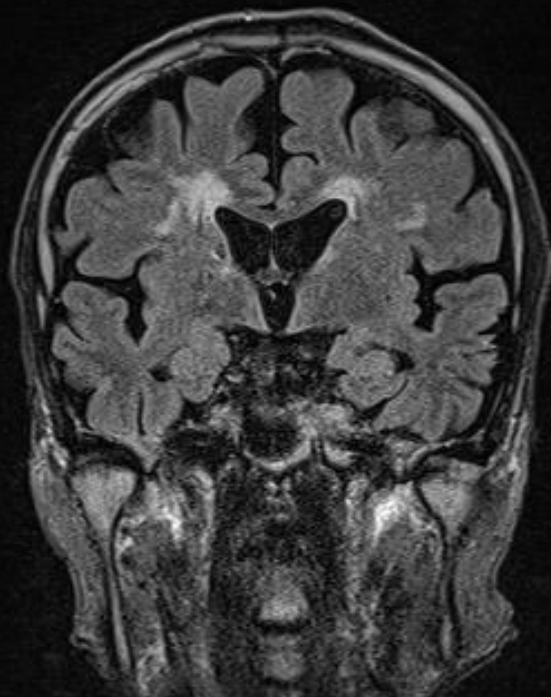
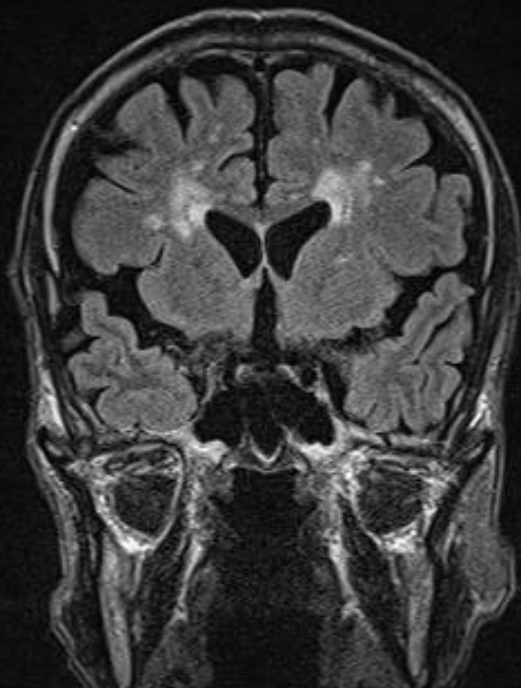
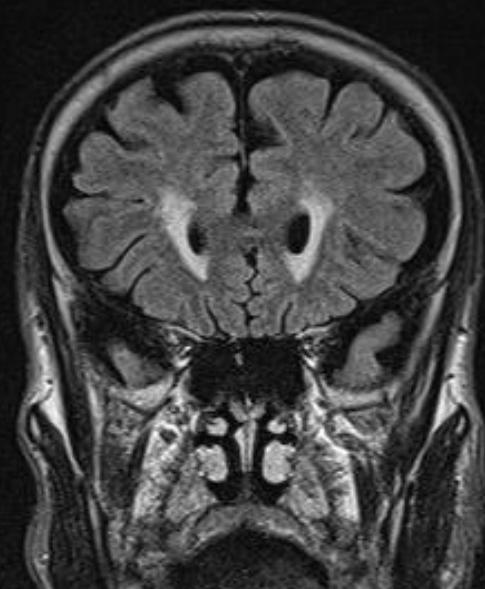
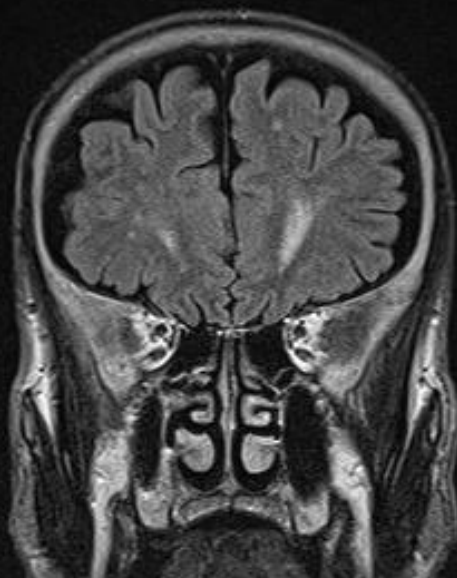
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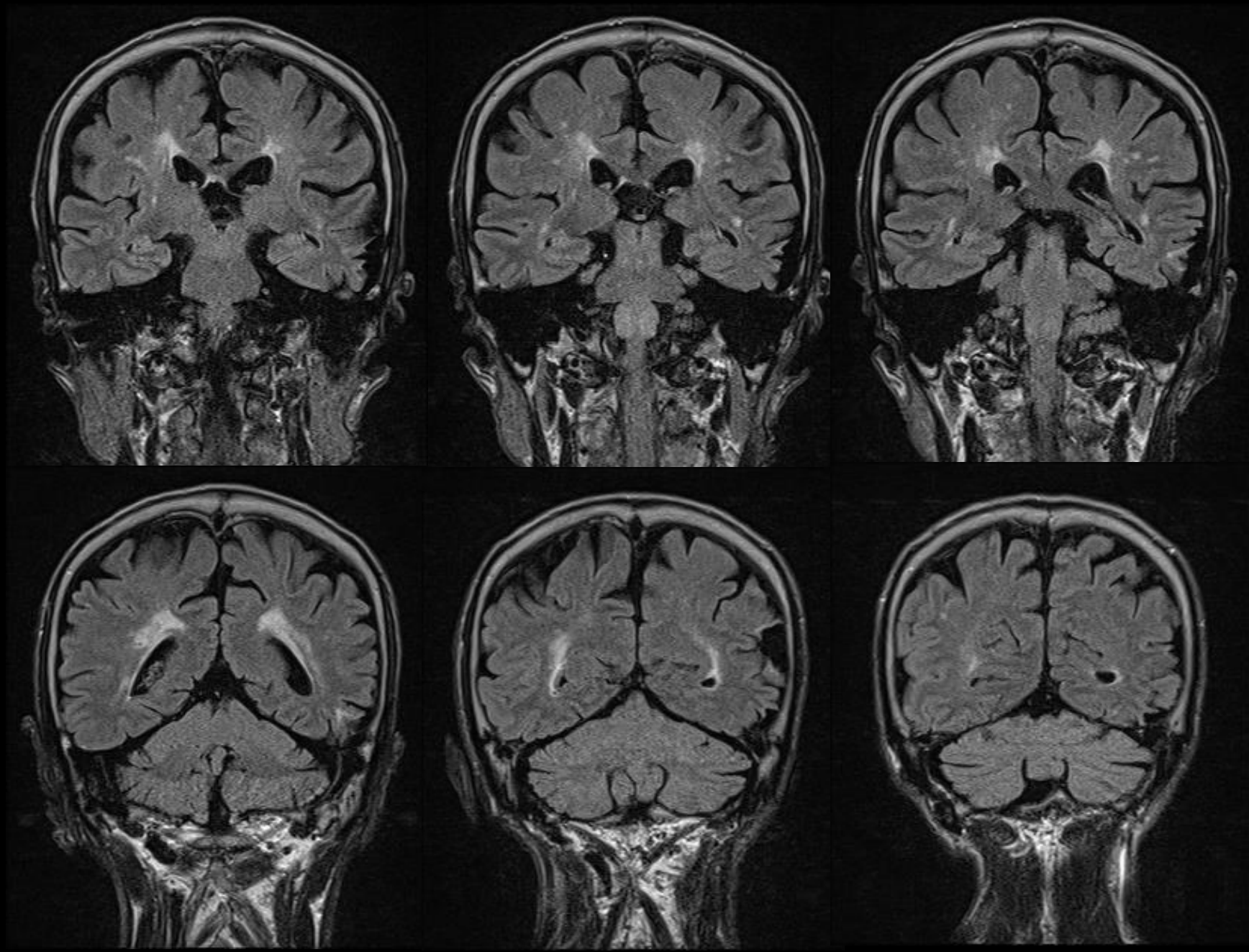
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神経内科紹介約10ヵ月後T2FLAIR 冠状断



神経内科紹介約10ヵ月後T2FLAIR 冠状断



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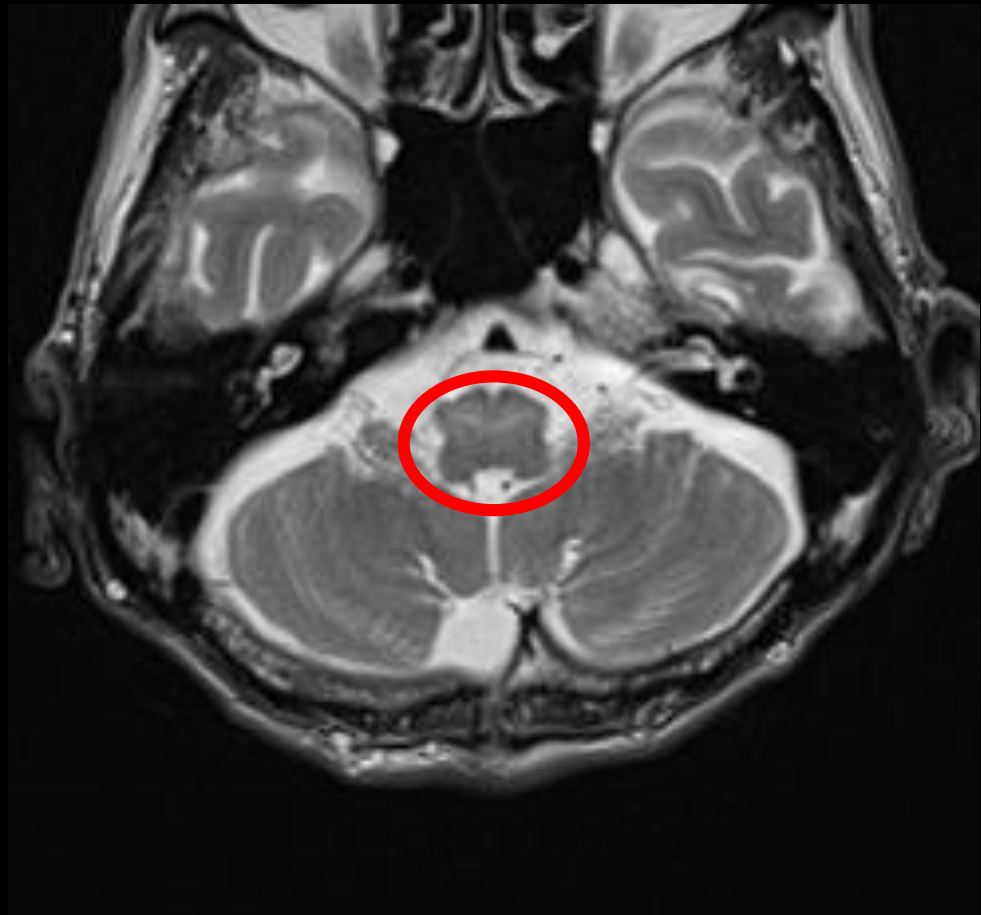
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臨床所見のまとめ

- ・口蓋振戦
- ・緩徐進行性の小脳失調の症状
- ・脳幹症状(MLF症候群)も合併
- ・家族歴はない

画像所見のまとめ

- ・下オリーブ核のT2WI異常高信号と肥大。
- ・小脳の軽度萎縮あり。



10ヶ月での経過の比較

神経内科紹介時

神経内科紹介約10ヶ月後



下オリーブ核のT2WI高信号が進行

下オリーブ核にT2WI高信号を認める疾患

1. 小脳歯状核には異常認めない

- ・進行性核上性麻痺(PSP-C)
- ・中間型延髄梗塞
- ・SCA1,2
- ・進行した多系統萎縮症
- ・Wilson病
- ・ミトコンドリア脳症

2. 小脳歯状核にも異常認める

- ・SCA20
- ・オリーブ核仮性肥大
- ・小脳歯状核腫瘍の摘出後
- ・メトロニダゾール脳症
- ・脳腱黄色腫症

progressive ataxia and palatal tremor (PAPT)

下オリーブ核の異常信号をとめない，口蓋振戦に進行性の小脳性運動失調を呈する変性疾患

口蓋振戦は，赤核，下オリーブ核，対側の小脳歯状核を結ぶGuillain-Mollaret 三角のいずれかの部位の障害により出現

Arch Neurol 1970;22:135-143.

< 診断 >

progressive ataxia and palatal tremor (PAPT)

PAPTの画像所見

下オリーブ核のT2WI異常高信号

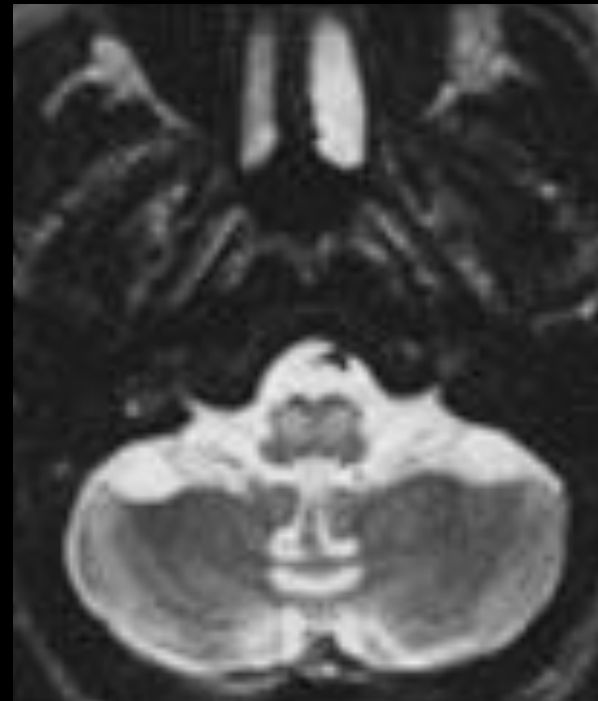
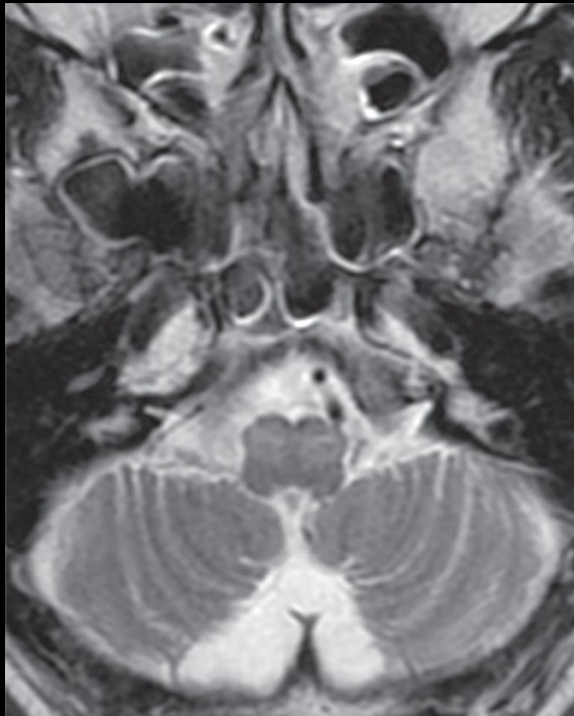


Table 1 Clinical and imaging details of our case and previously reported sporadic PAPT.

Author (Reference)	Age of onset/sex	Clinical features other than PT and ataxia	Hyperintensity of the IO	Hypertrophy of the IO	Other MRI features
Our case	71/M	none	(+)	(+)	CA
Kulkarni et al. ²⁾	32/M	dysarthria, nystagmus, abducent, oculomotor and facial nerve palsies	(+)	(+)	CA, pons and bilateral temporal lobes atrophy
Sperling et al. ⁵⁾	40/F	dysarthria, hyperactive tendon reflexes	NA	NA	CA
	60/F	dysarthria, external ophthalmoplegia, ear clicks	(+)	(+)	unknown
Samuel et al. ⁶⁾	40/M	diplopia, SEM, skew deviation, gaze-evoked nystagmus, left MLF syndrome	(+)	(+)	CA
	53/M	dysarthria, SEM, oscillopsia, vertical nystagmus, depression	(+)	(+)	CA
	63/M	dysarthria, SEM, bradykinesia of foot, postural tremor, difficulty with focusing, dementia	(+)	(+)	CA
	58/M	dysarthria, SEM, oscillopsia, downbeating nystagmus	(+)	(+)	CA
	63/M	dysarthria, SEM, exotropia, bilateral MLF syndrome	(+)	(+)	CA
	58/M	dysarthria, SEM, torsional nystagmus, dysphagia, incontinence, deafness	(+)	(-)	CA Brain stem atrophy
Cilia et al. ⁷⁾	50/M	dysarthria, SEM, bradykinesia of the left hand	(+)	(+)	CA
Bassani et al. ⁸⁾	53/M	Oscillopsia, nystagmus	(+)	(+)	CA mild hyperintensity of the SCP
Brinar et al. ⁹⁾	Middle-aged/F	none	(+)	(+)	CA
de Jong et al. ¹⁰⁾	66/M	dysarthria, SEM, swallowing difficulties	(+)	(+)	none
	61/F	dysarthria, SEM, swallowing difficulties	(+)	(+)	none

PT=palatal tremor, CA=cerebellar atrophy, IO=inferior olive, NA=not available, SEM=saccadic eye-movement, SCP=superior cerebellar peduncles

本症例の問題点

- PATAは小脳に病変が限局するのか、脳幹に限局するのか、両者が混在するのか、病態ははっきりしていない。本症例を含め、今後多数の臨床像の検討が必要である。
- PAPTと診断するには他の下オリーブ核に異常を認める疾患を除外する必要がある。

結語

- PAPTが最も考えられる症例を経験した。
- 下オリーブ核の異常信号はPAPTを疑うきっかけとなった。