

# Case presentation

Kazuhiro Murayama, Kazuhiro Katada  
Fujita Health University

# 症 例

- 50歳 女性
- 主訴：言葉の出にくさ
- 既往歴・家族歴：特記事項なし
- 現病歴：1か月前から言葉の出にくさが出現し、徐々に増悪するため他院を受診。頭部MRIで異常を認めため当院脳神経外科紹介となった
- 現症：JCS I-0, 体温37.7°C, 血圧111/57mmHg, 脈拍75/分. 発語の障害以外明らかな神経学的所見なし。

- A 50y.o. woman presented with speech disorder before a month. She has gone worse gradually. Multiple abnormal findings were revealed in MRI examination. So, she was admitted to our hospital.
- Vital signs: JCS I-0, 37.7°C, BP111/57mmHg, HR75/min
- Neurological examination findings were normal except for speech disorder.

# 検査所見

## 【血液検査】

末血・生化：特記すべき異常所見を認めず。

腫瘍マーカー：IL-2R 346U/m, CYFRA 0.3ng/mL, CEA 0.4ng/mL,  
CA19-9 14.6U/mL, SCC 0.6ng/mL, PIVKA II 1.2mAU/mL

## 【髄液検査】

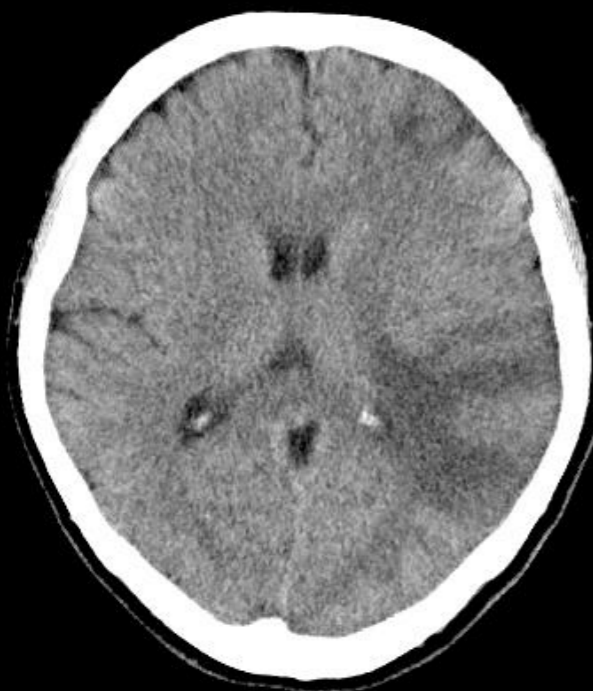
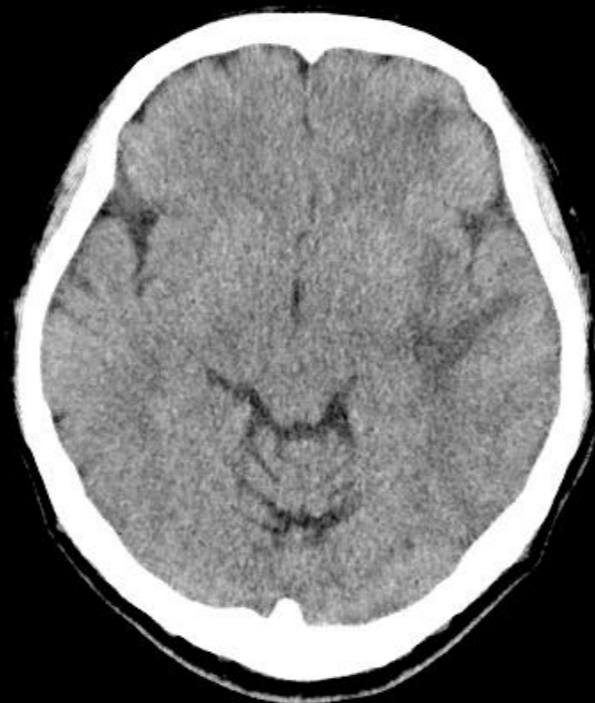
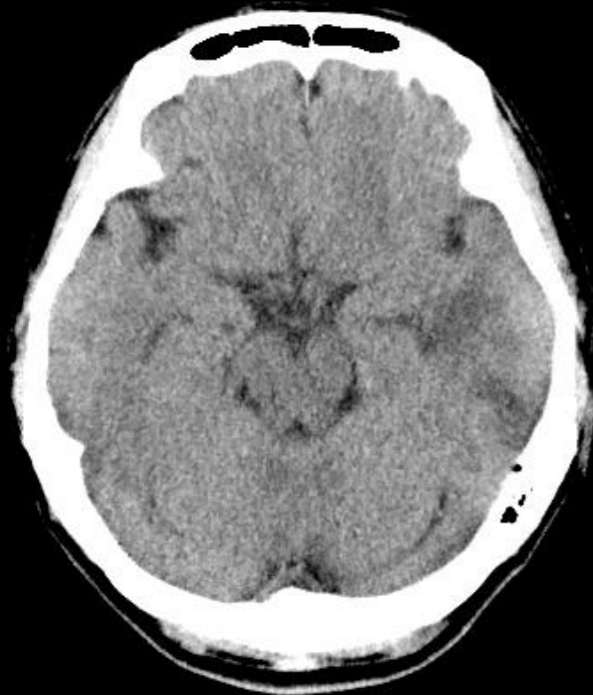
総蛋白量28mg/dL, 細胞数0/ $\mu$ L (MONO 0, POLY 0),  
糖量55mg/dL, 塩素量123mEq/L, 髄液トリプトファン反応(-)

## 【その他の検査所見】

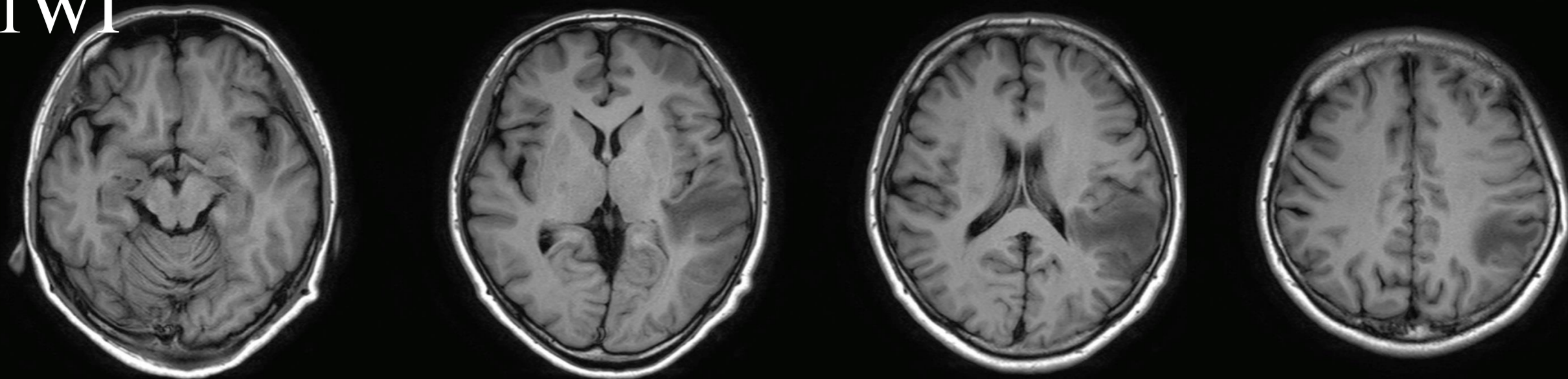
全身スクリーニングのために施行されたCTで縦隔腫瘍が認められ、後に生検で胸腺腫と診断された。

- Baseline haematological and biochemical investigations were normal; a lumbar puncture revealed a normal cerebral spinal fluid.
- A mediastinal tumor was revealed in screening CT, which was diagnosed as a thymoma.

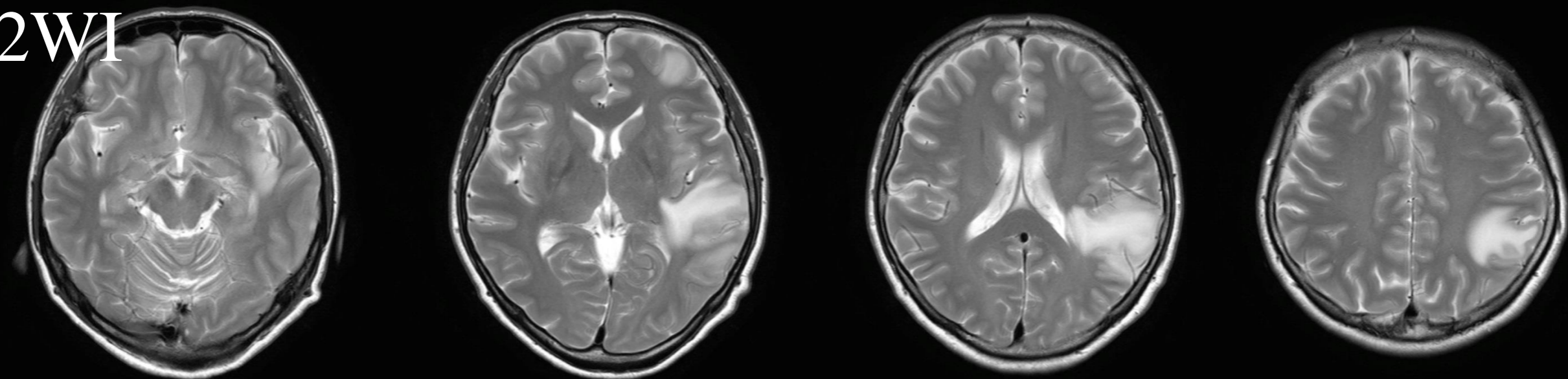
NC-CT



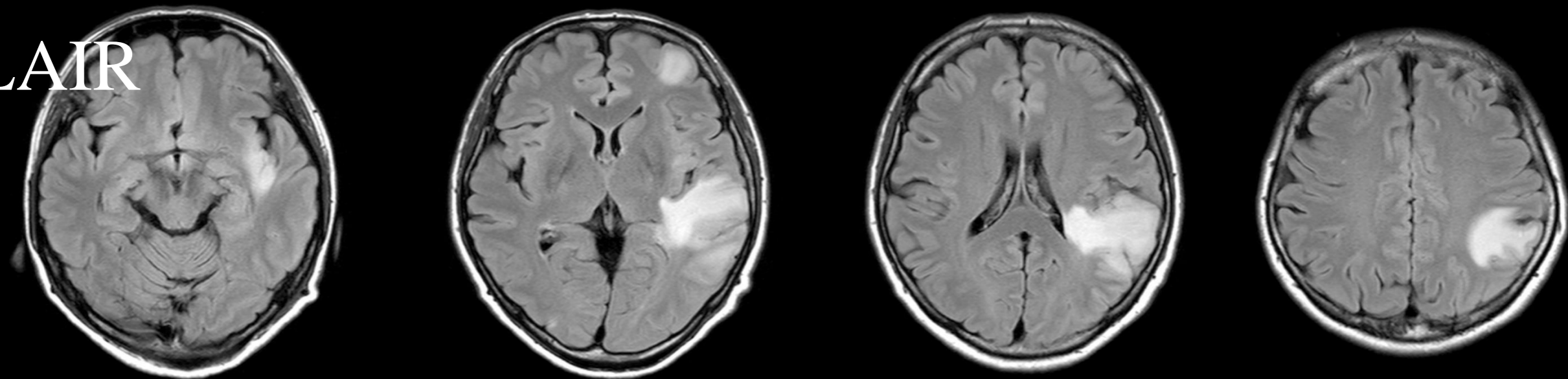
T1WI



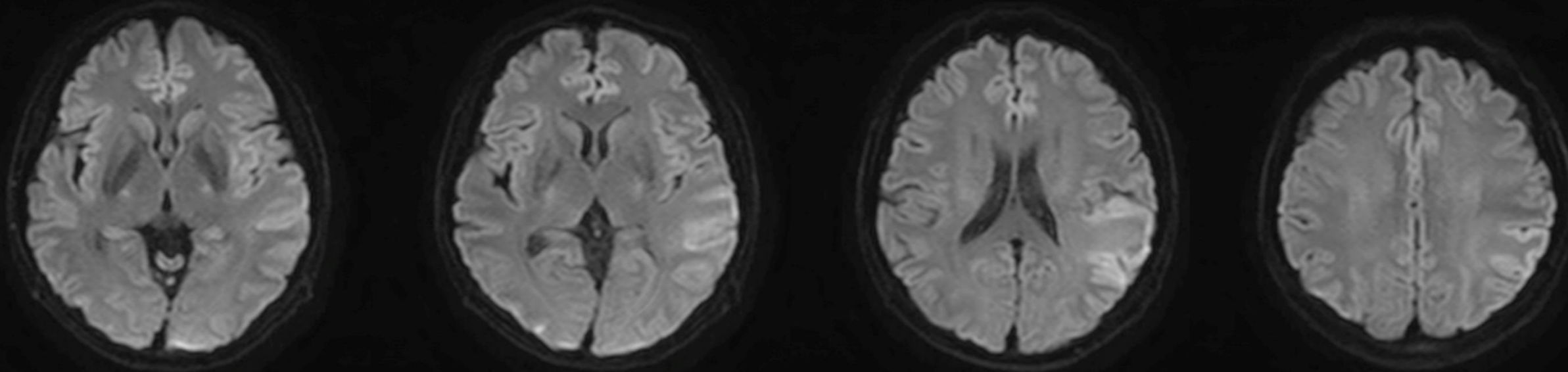
T2WI



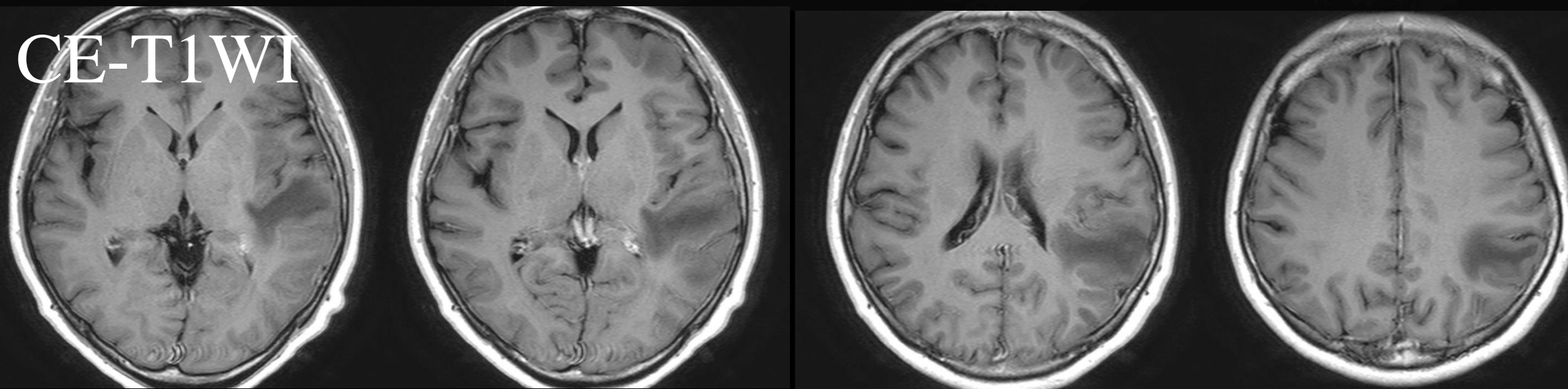
FLAIR



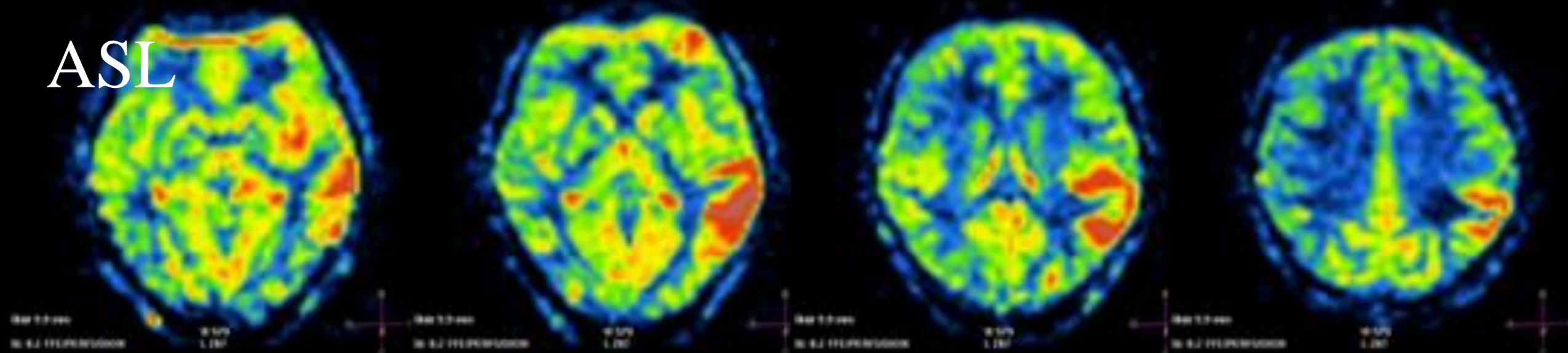
DWI



CE-T1WI



ASL



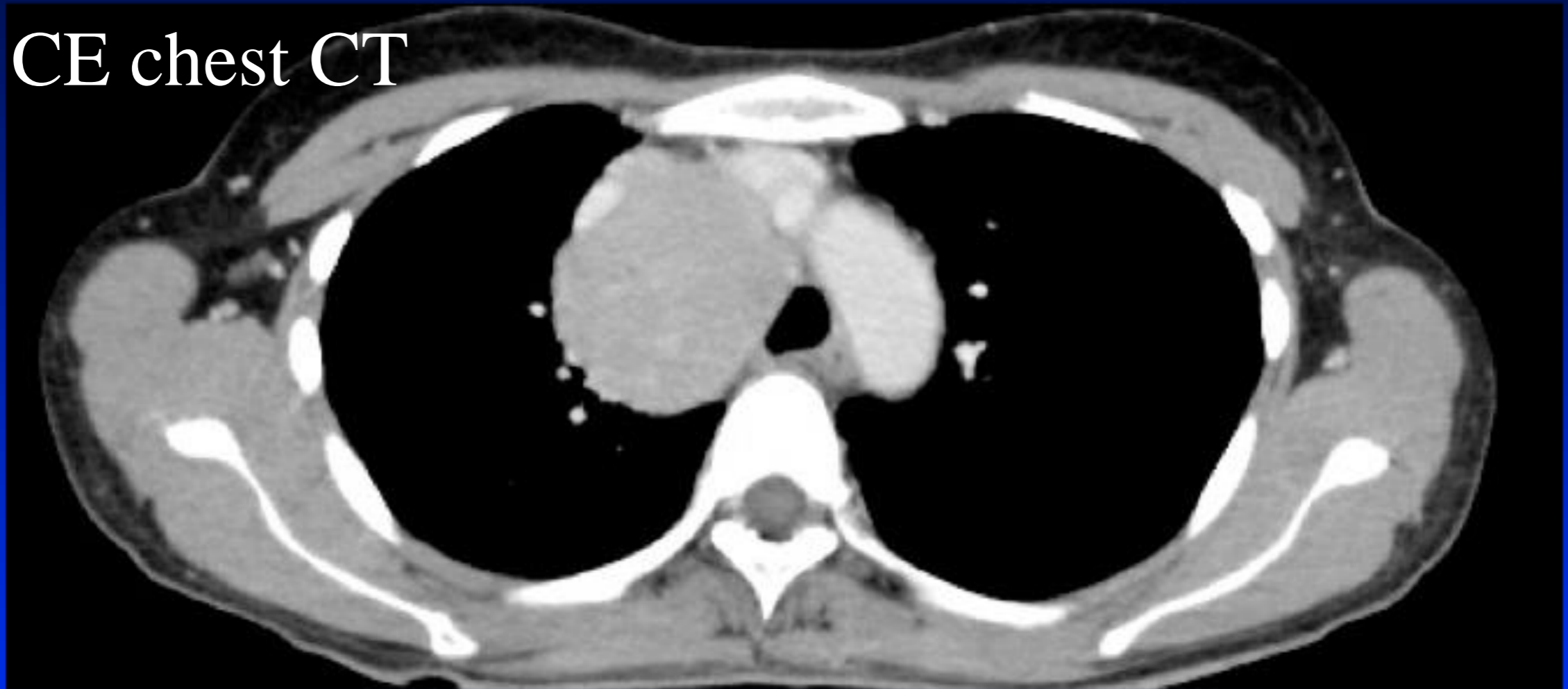
読影を宜しく願います。

Please diagnosis!



blank

CE chest CT



- A mediastinal tumor was revealed in screening CT, which was diagnosed as a thymoma.

# $^1\text{H}$ -MRS

## Long TE



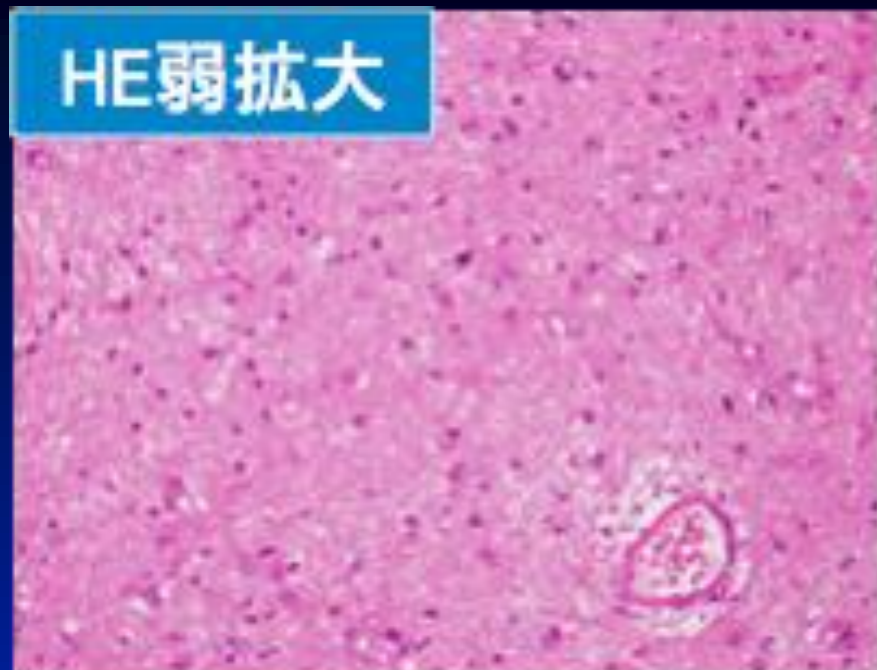
# Differential diagnosis of the multiple cerebrum lesion

- brain metastases no enhancement
- malignant lymphoma no enhancement, DWI↓
- brain abscess no enhancement, DWI↓
- vasculitis no enhancement, CBF↑
- ADEM/NMO
- MELAS Lactate↑
- PML regression and progression
- paraneoplastic syndrome CBF↑, extra limbic pattern, thymoma

# Differential diagnosis of the multiple cerebrum lesion

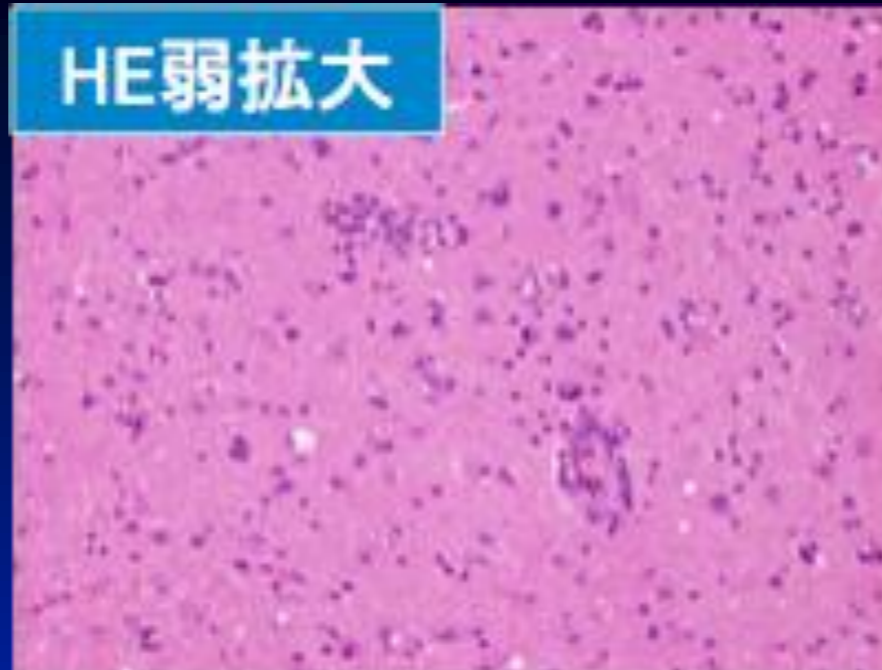
- brain metastases no enhancement
- malignant lymphoma no enhancement, DWI↓
- brain abscess no enhancement, DWI↓
- vasculitis no enhancement, CBF↑
- ADEM/NMO
- MELAS Lactate↑
- PML regression and progression
- paraneoplastic syndrome CBF↑, extra limbic pattern, thymoma

HE弱拡大



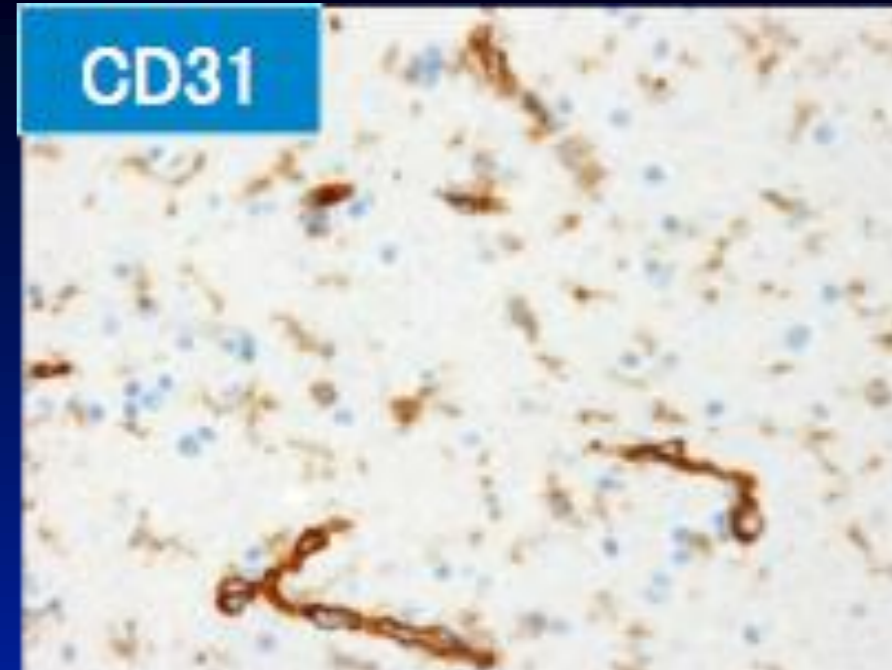
灰白質の構造破壊

HE弱拡大



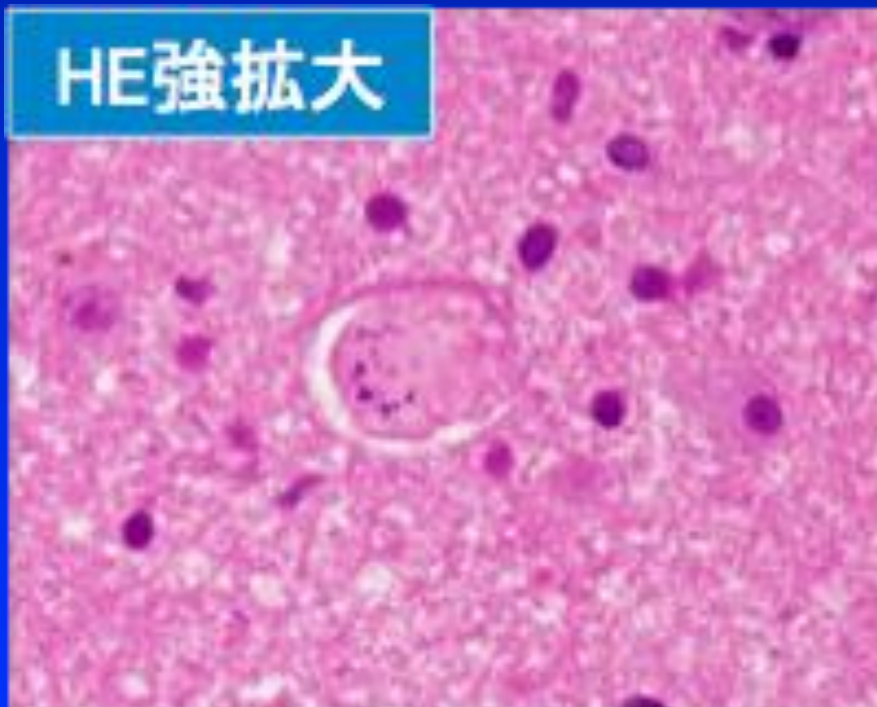
血管周囲リンパ球浸潤

CD31



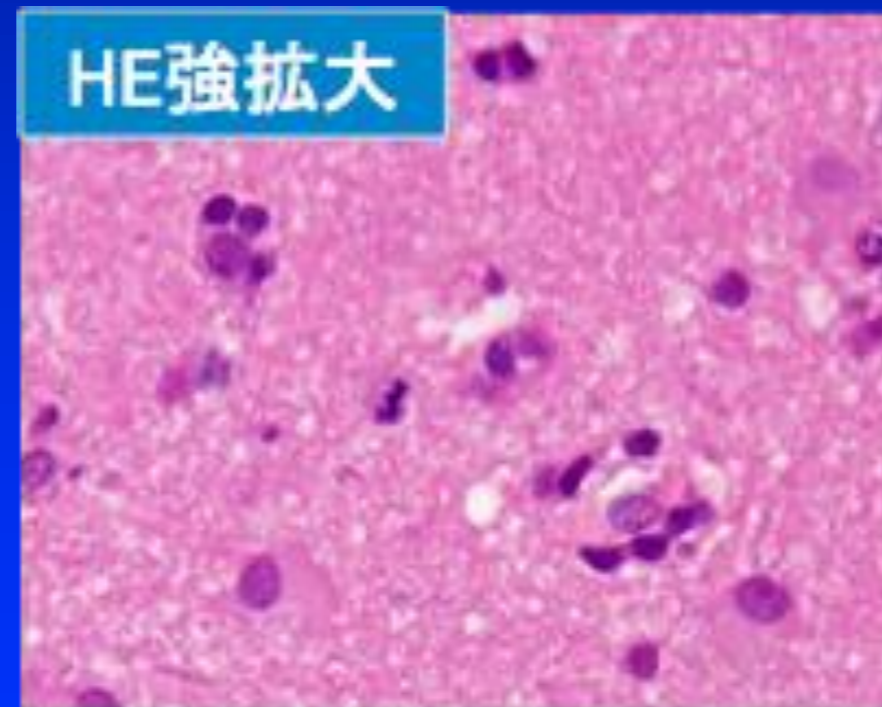
活性化ミクログリア浸潤

HE強拡大



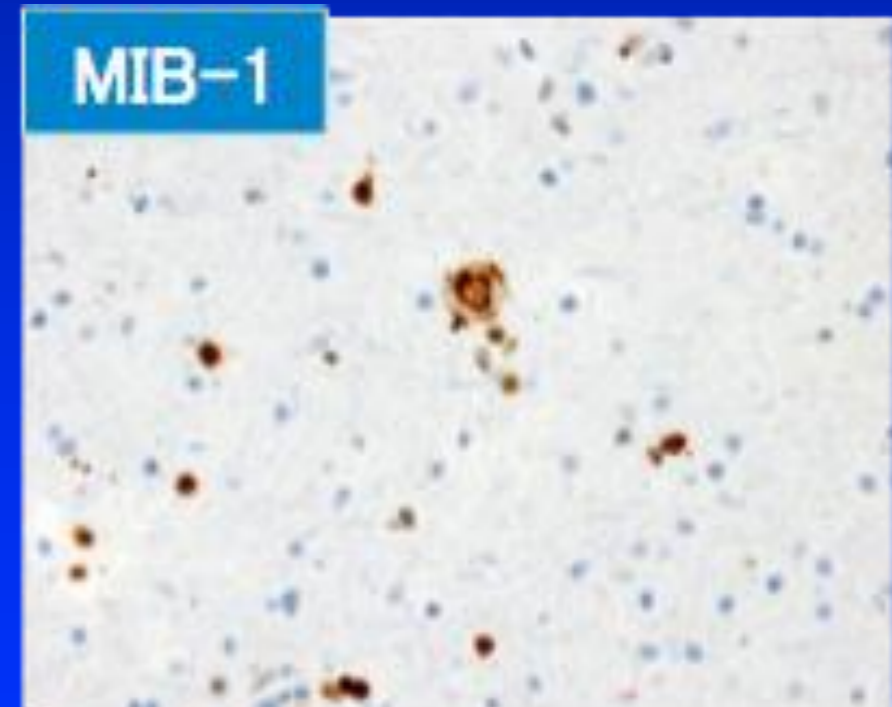
Creutzfeldt astrocyte

HE強拡大



細胞分裂像

MIB-1

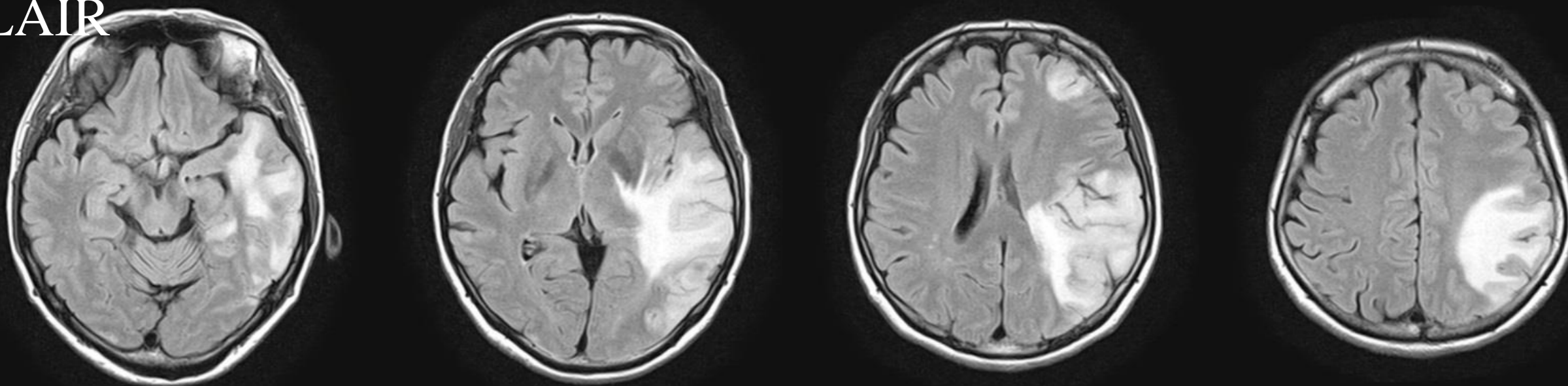


分裂期の細胞増加

平戸先生 宜しくお願いします。

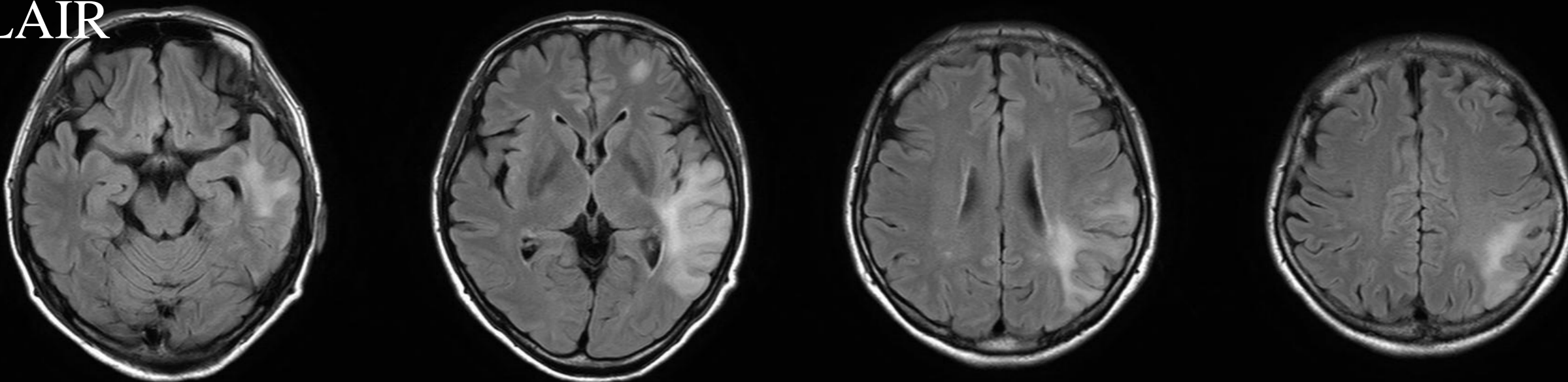
# Before thymectomy

FLAIR



# After thymectomy 14POD

FLAIR



# Diagnosis

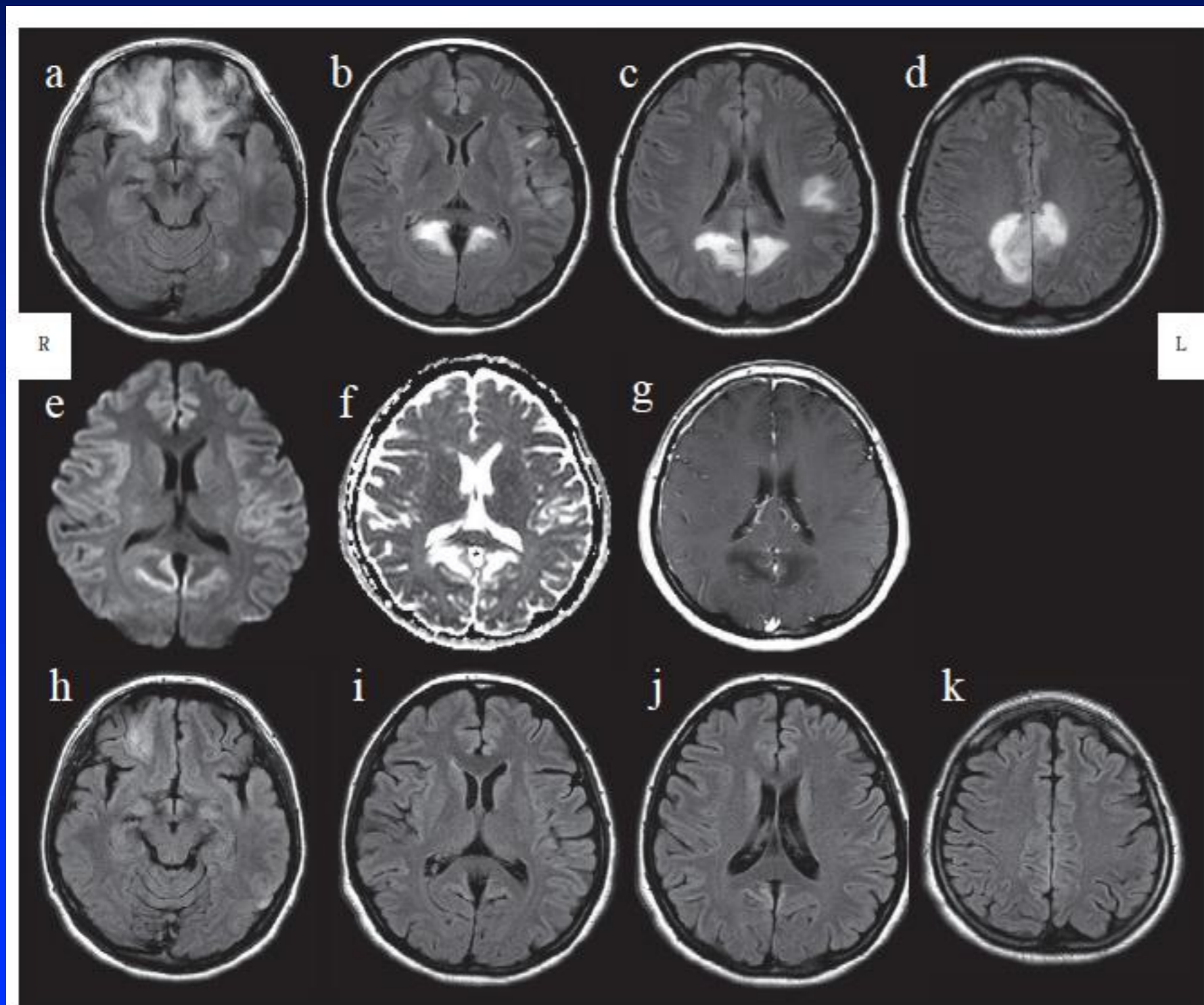
paraneoplastic extra limbic encephalitis/encephalopathy  
associated with thymoma

胸腺腫関連 傍腫瘍性非辺縁系脳炎・脳症

# Paraneoplastic extra limbic encephalitis/encephalopathy associated with thymoma

- MRI findings are similar to common PNS **except for location of lesion**
  - Cortex and subcortical white matter involved
  - High intensity signal in DWI
  - High CBF in ASL
- Rare disorder, pathogenesis and clinical management of the disease remains uncertain.
- Recognition of paraneoplastic extralimbic encephalitis is important because this disorder are **treatable.**

# Immune-mediated encephalopathy associated with anti-GAD antibody



On admission

After plasma exchange

Kobayakawa et al.  
Clin Neurol 2010;50:92-97

Fig. 2 Brain MRI on admission and after treatment.

