

# Neuroradiology Workshop 2014

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## 【16-year-old male】

### 【主訴】

意識障害、経口摂取不良

### 【現病歴】

生来健康。2年前より転びやすくなった。半年前より徐々に転ぶ頻度が増え、右手優位の手指の震えが目立つようになり、書字の乱れも認められた。昨年より集中力低下や学校の成績低下が起こり、同時期より視力も低下するようになったため、当院受診し、精査中であった。

またここ数カ月は食欲不振が続き、手すり歩行でないと移動できない状態であった。階段を降りようとしたところ、頭痛と悪心を認め、意識消失したため救急要請となった。

### 【C.C】

Disturbance of consciousness and poor appetite.

### 【History of present illness】

A previously healthy 16 year-old man who has been disposed to fall since 2 years ago. The number of times to fall was increased and finger tremor (mainly right hand) and dysgraphia have been appeared since a half year ago. Poor consciousness, decline in academic ability and reduced vision have been appeared 1 year ago, he visited our hospital and he has been examined carefully. Poor appetite have continued the last few months and he could not walk without handrail walking. When an attempt is made to get down the stairs, showed headache, nausea and disturbance of consciousness, so ambulance was requested.

## 来院時神経学的所見

- 意識清明
- 脳神経学的異常所見なし
- MMT  
Beceps 4/3 Triceps 4/3  
Wrist Ext.4/3 Wrist Flex 4/3  
Iliopsoas 3+/3 Quadriceps 4/3+  
Hamstrings 4/3  
Gastrocnemius 5/5  
Tibialis Ant 5/5
- 母指球筋の萎縮なし
- Spasticity なし rigidityなし
- Sensory 異常なし
- Babinski反射陰性
- 指鼻指試験陽性

## Neurologic examination

- Gen; alert awake
- Cranial nerve is all normal
- MMT  
Beceps 4/3 Triceps 4/3  
Wrist Ext.4/3 Wrist Flex 4/3  
Iliopsoas 3+/3 Quadriceps 4/3+  
Hamstrings 4/3  
Gastrocnemius 5/5  
Tibialis Ant 5/5
- No thenar eminence waste
- No pasticity, rigidity
- Sensory is all normal
- Babinski reflex negative
- Finger-nose-finger test positive

# Laboratory data

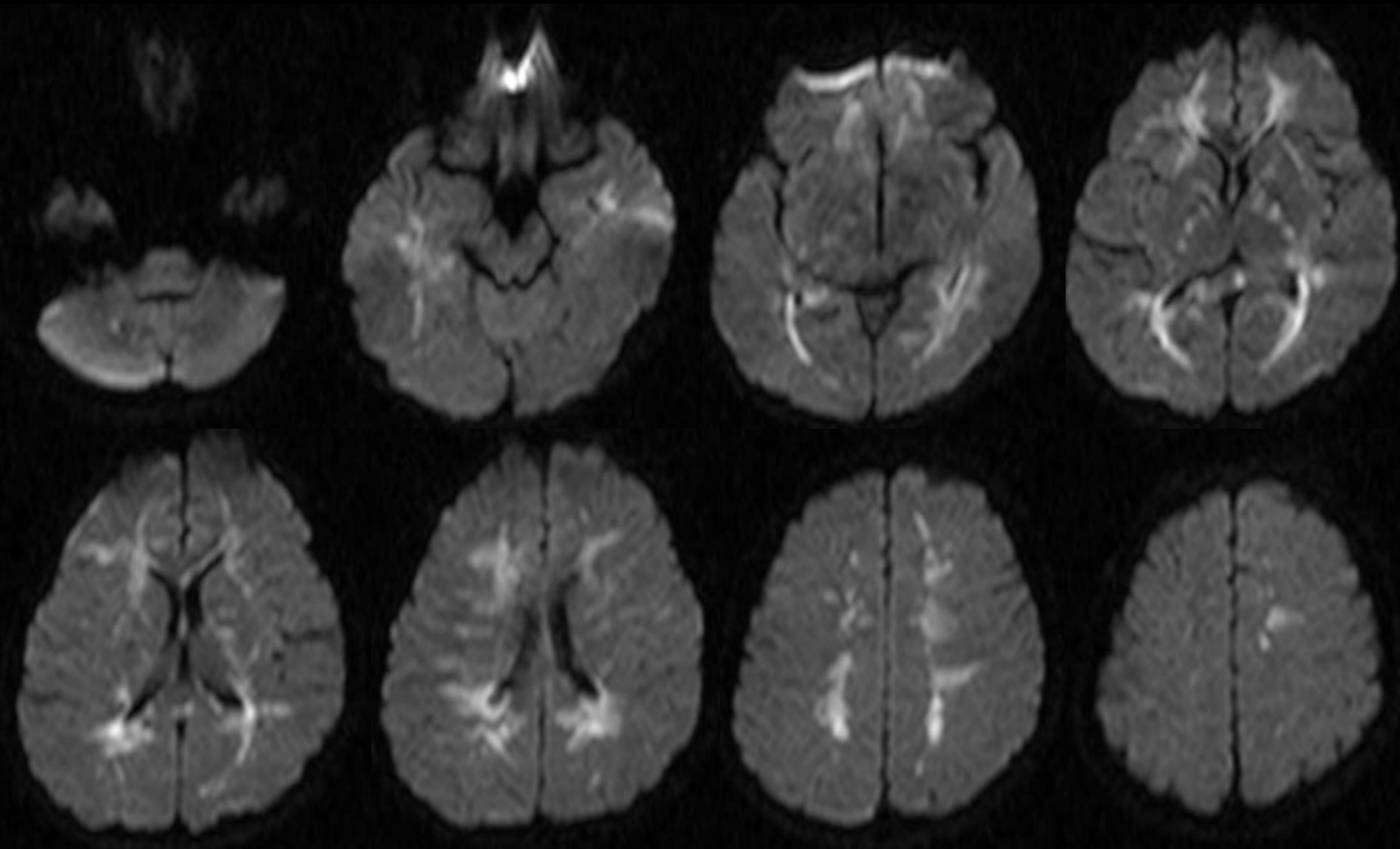
- Biochemical examination of blood

WBC	11300	/ $\mu$ L
RBC	560万	/ $\mu$ L
Hb	15.9g	/dL
Ht	44.5	%
Plt	13.7	/ $\mu$ L
GOT	14	U/L
GPT	9	U/L
LDH	128	U/L
$\gamma$ -GTP	45	U/L
Cr	0.79	mg/dL
BUN	13	mg/dL
Glu	199	mg/dL
Na	138	mEq/L
K	4.4	mEq/L
Cl	102	mEq/L
CRP	0.55	mEq/L

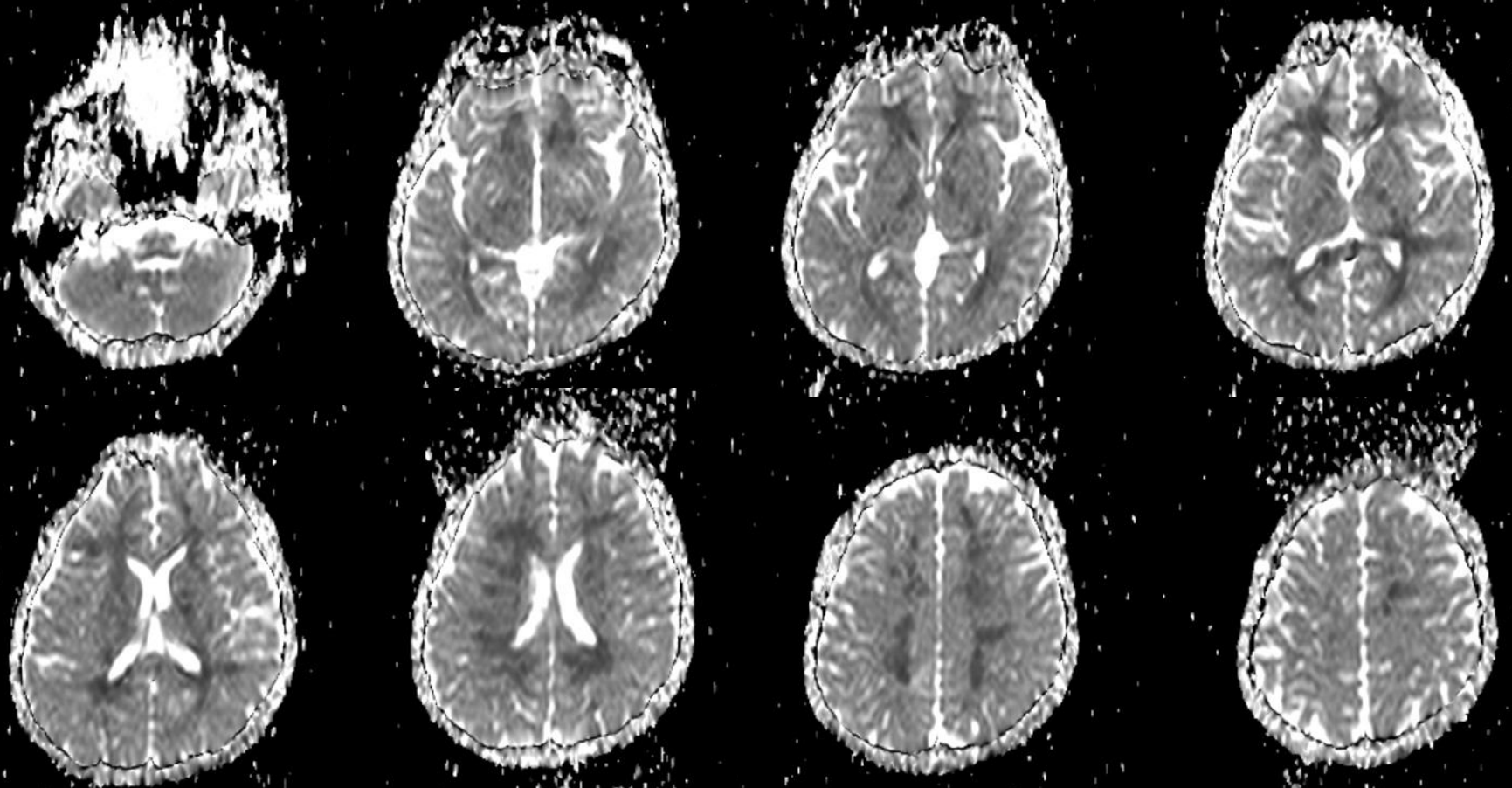
- Examination of cerebrospinal fluid (CSF)

appearance	: clear
Cl	: 123 mEq/L
Glu	: 58 mg/dL
proteins	: 446 mg/dL
specific gravity	: 1.008
count of cell	: 3

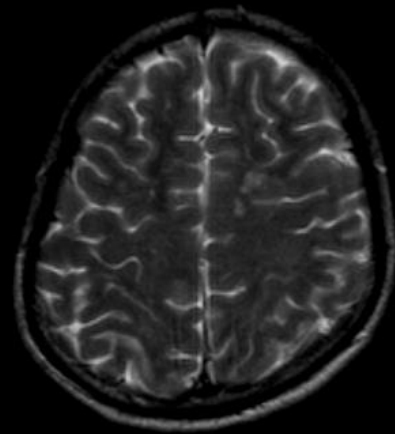
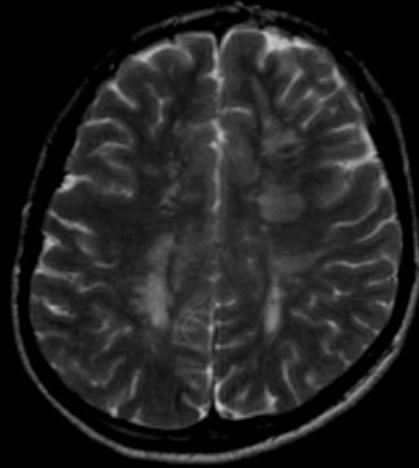
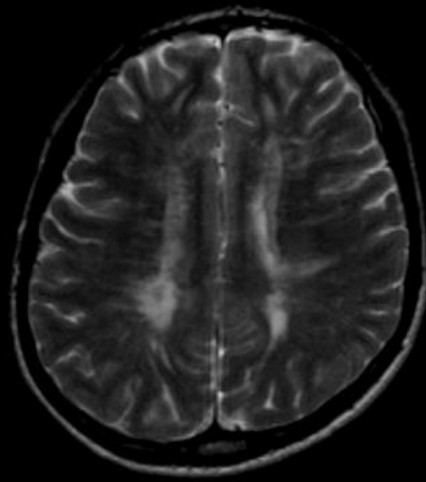
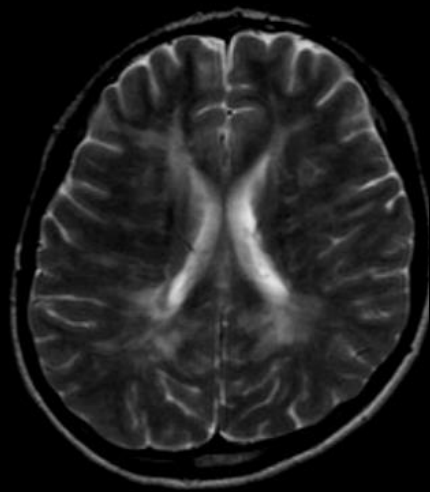
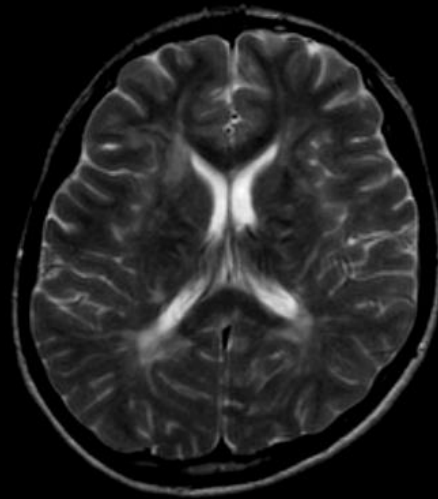
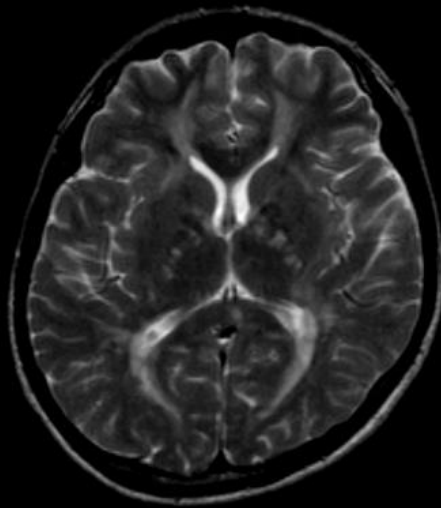
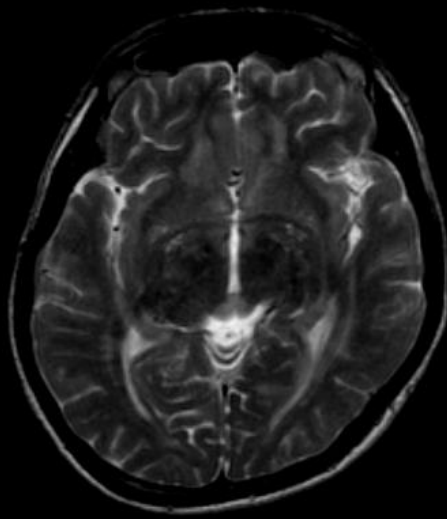
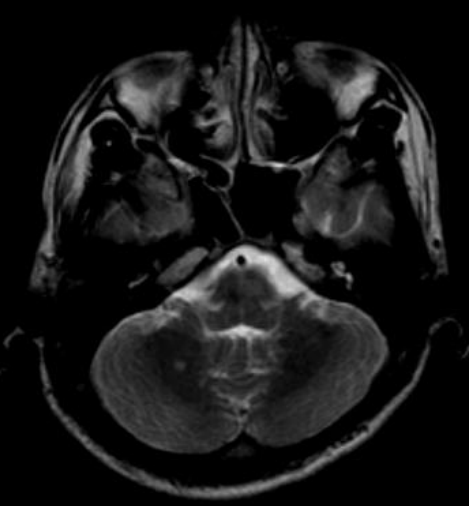
DWI



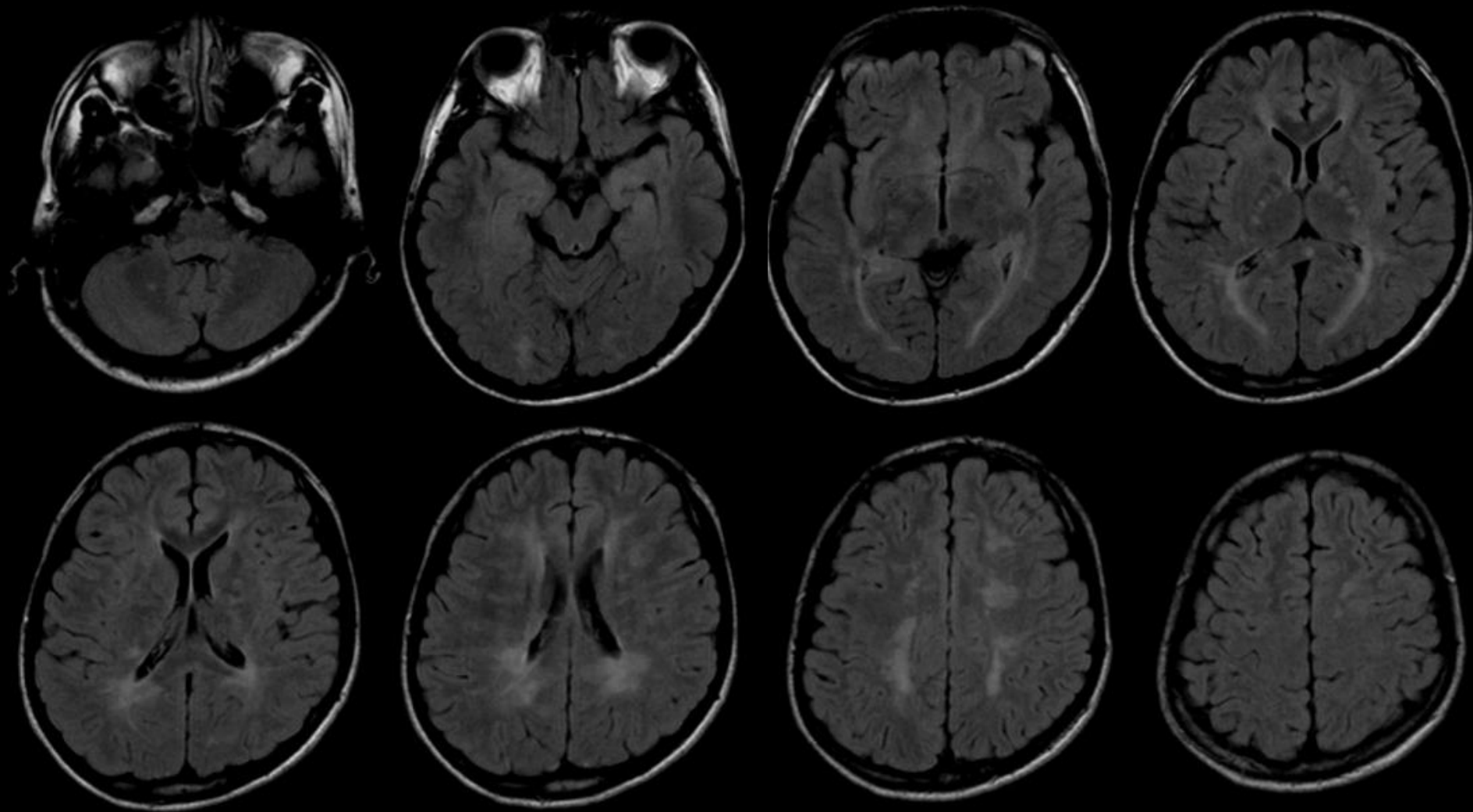
# ADC map



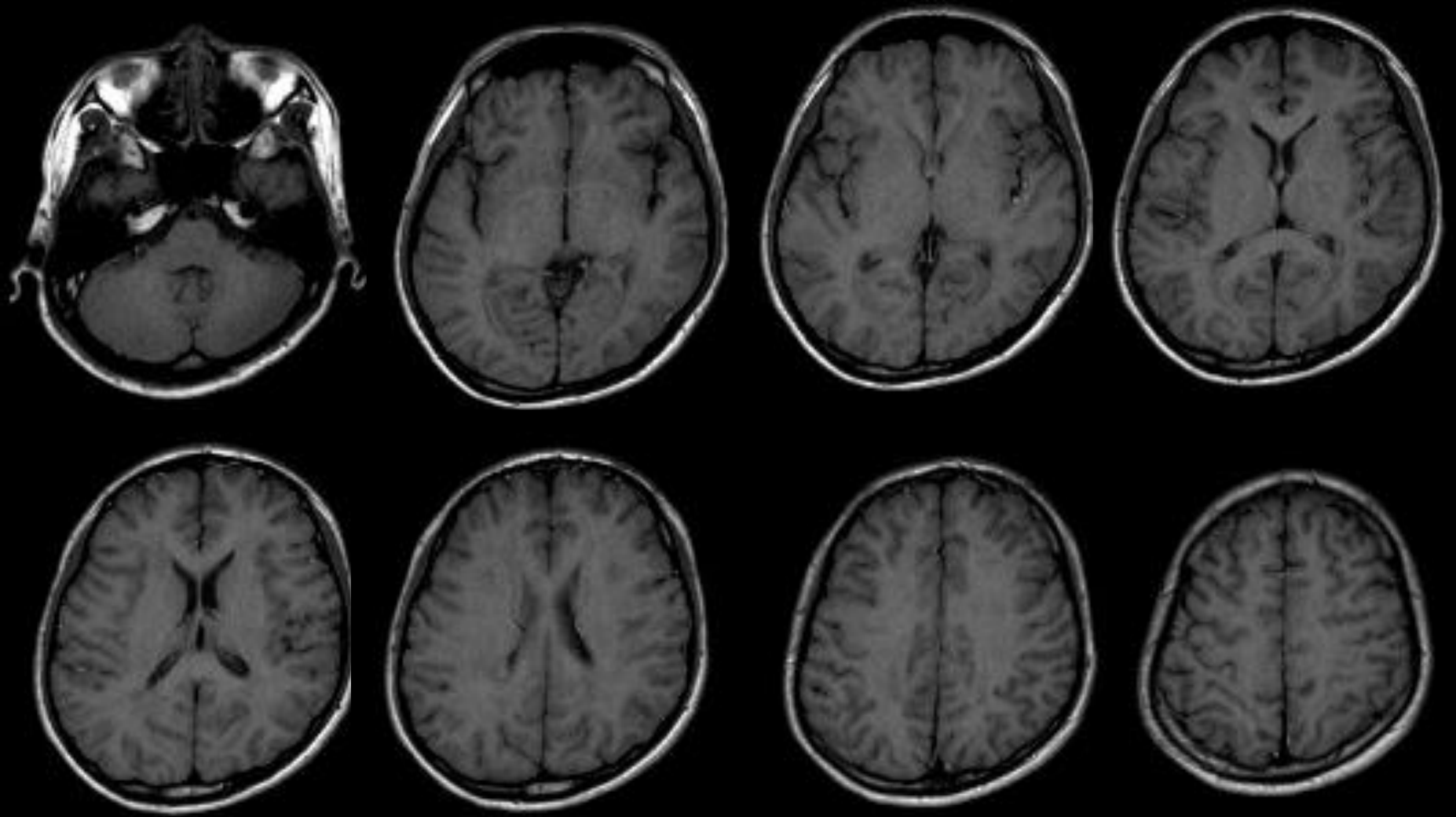
T2WI



FLAIR

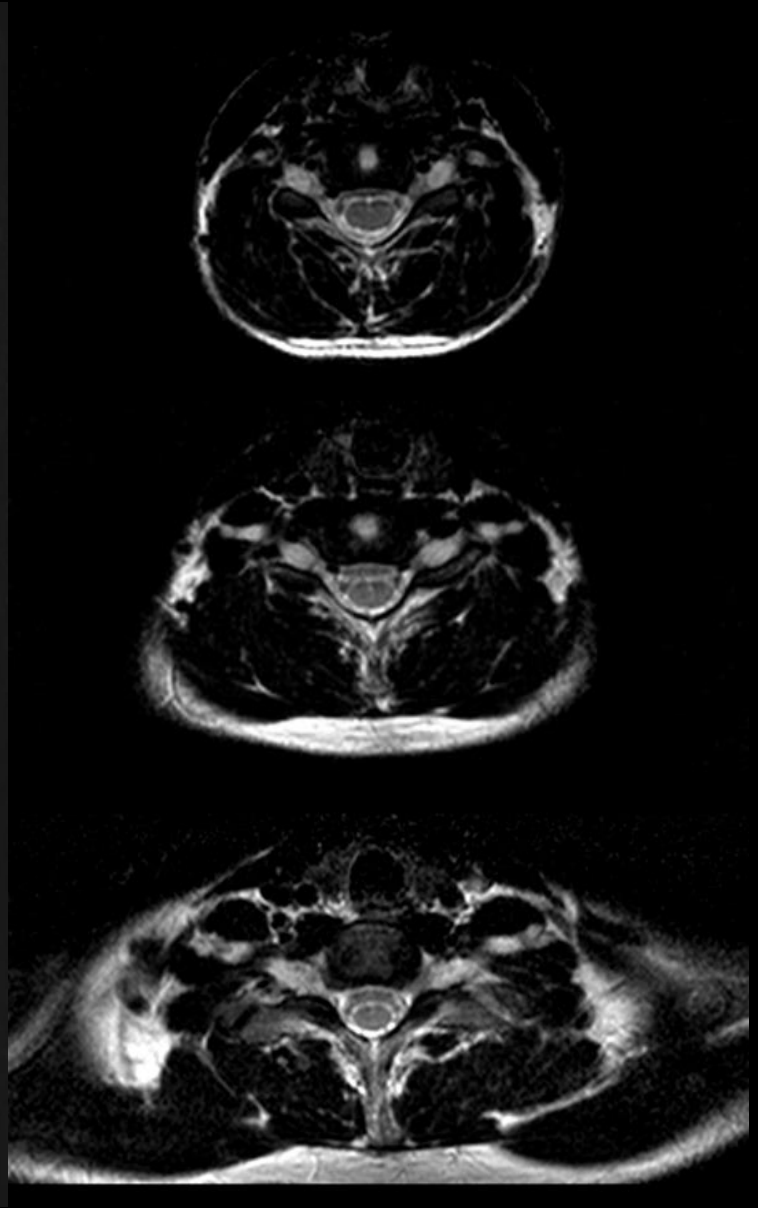


T1WI



# Spine MRI cervix level

# T2WI axial

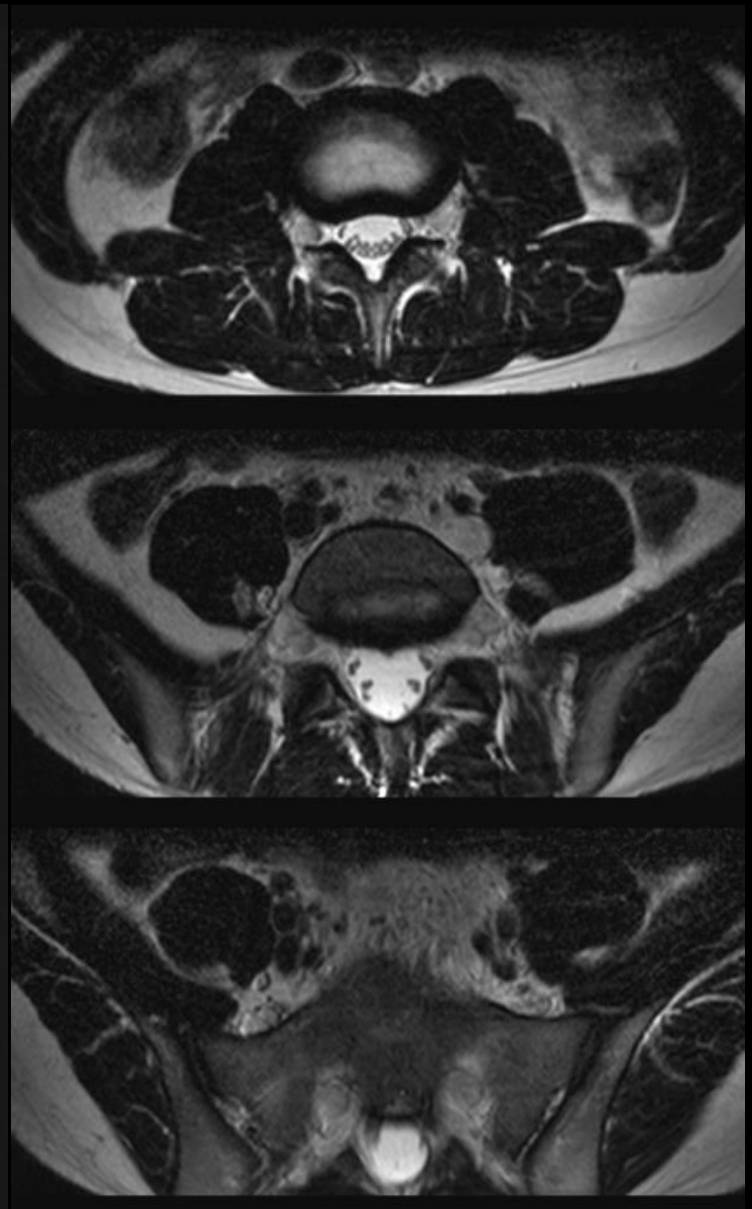


T1WI

T2WI

Spine MRI lumbar level

T2WI axial



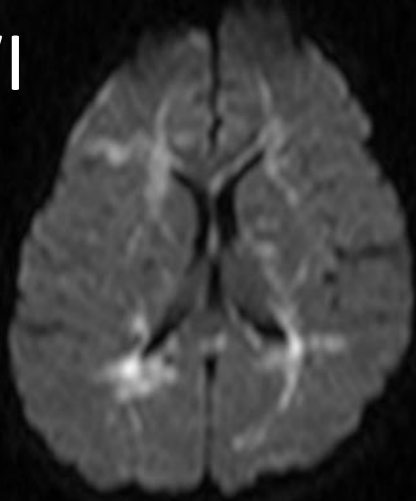
以上の所見から考えられる鑑別診断は？

Please diagnose.

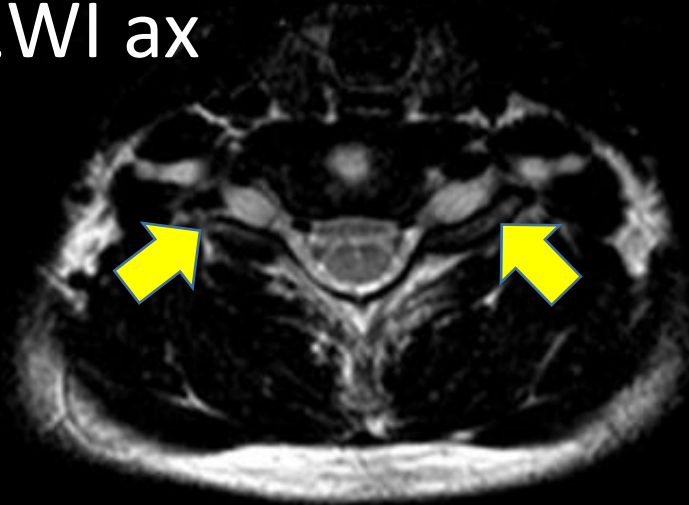


# Key Image

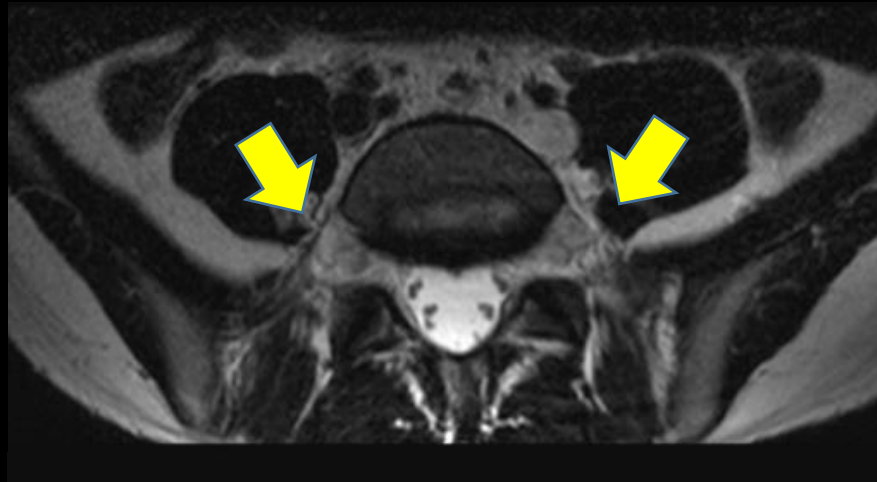
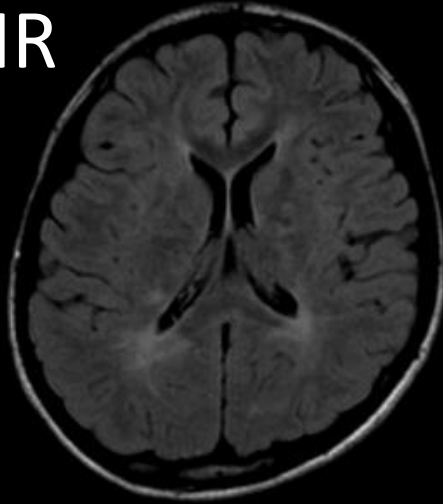
DWI



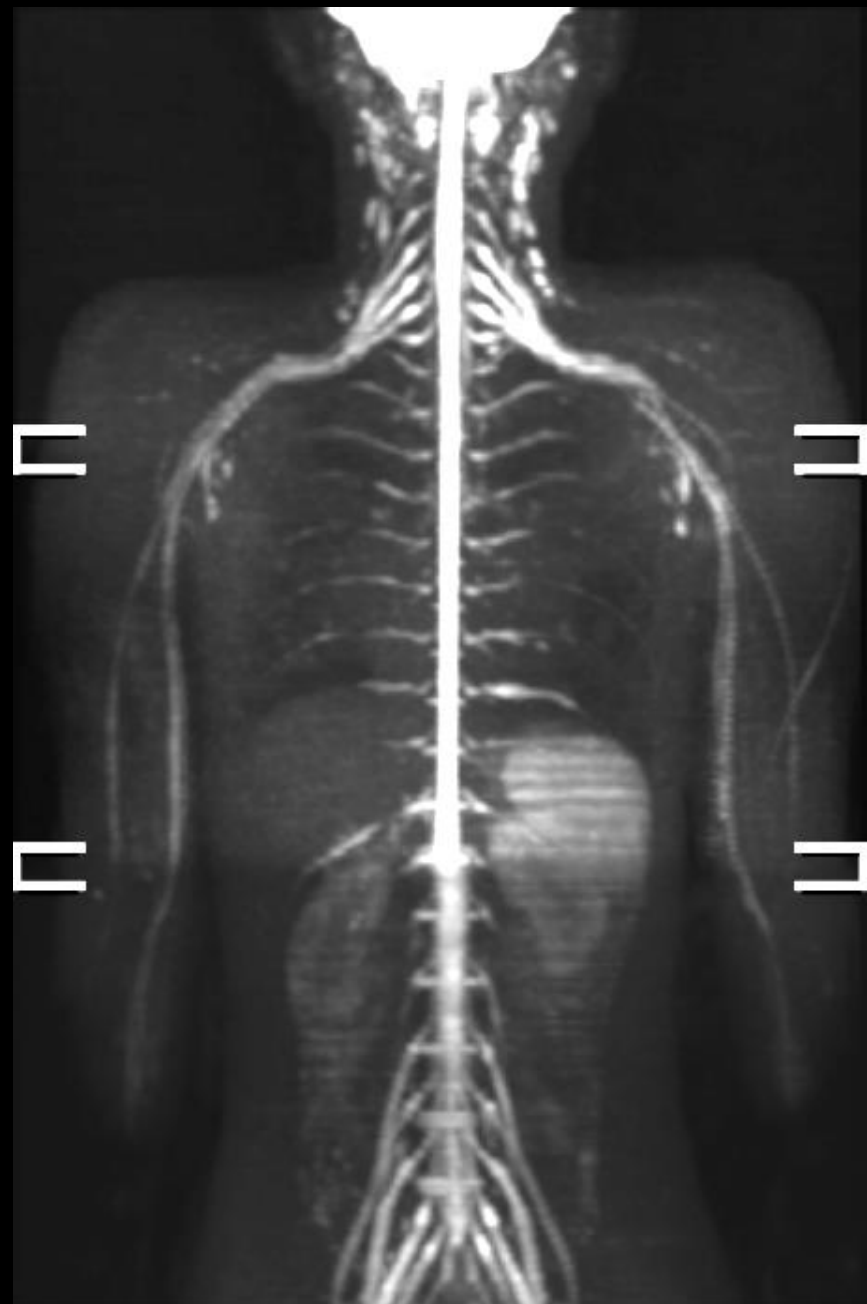
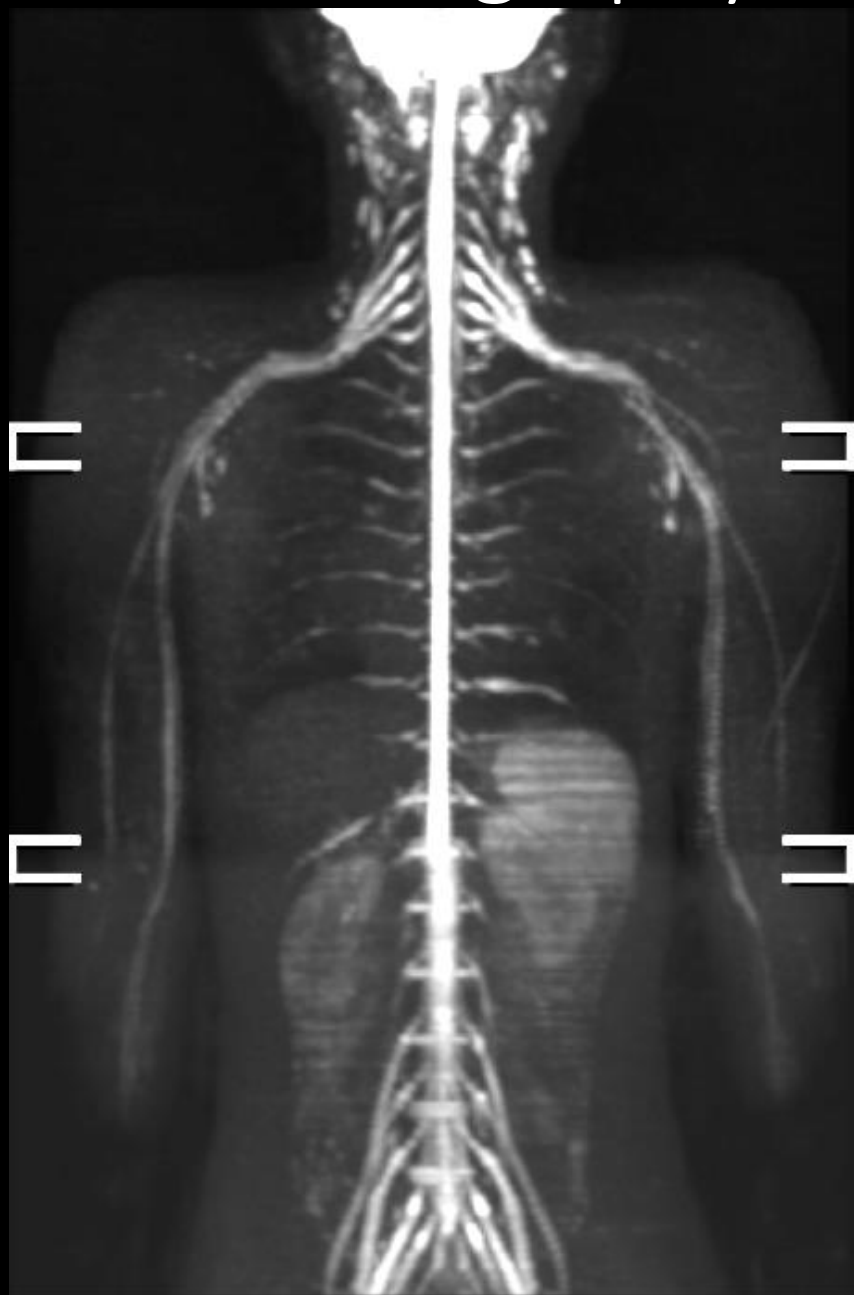
T2WI ax



FLAIR



# MR neurography



DWI高信号

びまん性大脳白質病変＋小脳半球病変

＋

びまん性両側性の神経根腫大



・ミトコンドリア病

・PRES

・MS

・傍腫瘍症候群

・サルコイドーシス

・ベーチェット病

・Guillan-Barre Synd.

・CIDP

・ADEM

・Charcot Marie Tooth

?

# Clinical course

- Examination of cerebrospinal fluid (CSF)

appearance : clear

Cl : 123 mEq/L

Glu : 58 mg/dL

**proteins** : **446 mg/dL**

specific gravity : 1.008

count of cell : 3

- 筋電図 → perioheral nerve disorder

- 各種抗体提出 : **抗neurofascin 抗体陽性** + その他の抗体は陰性

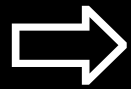


Combined central and peripheral demyelination  
with anti-neurofascin antibody (CCPD)

(抗neurofascin抗体陽性中枢・末梢連合脱髓症)

# central and peripheral demyelination

- myeline supporting cellsの崩壊による



中枢神経 (CNS) : oligodendrocytes

末梢神経 (PNS) : schwann cells

## Central demyelinating disease

- Multiplesclerosis
- ADEM
- acute hemorrhagic leucoencephalitis
- Osmotic demyelination syndromes
- PML

## Peripheral demyelinating disease

- Guillan-Barre syndrome
- Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
- Paraproteinemic demyelinating neuropathy
- Charcot Marie Tooth
- Copper deficiency

# combined central and peripheral demyelination with anti-neurofascin antibody

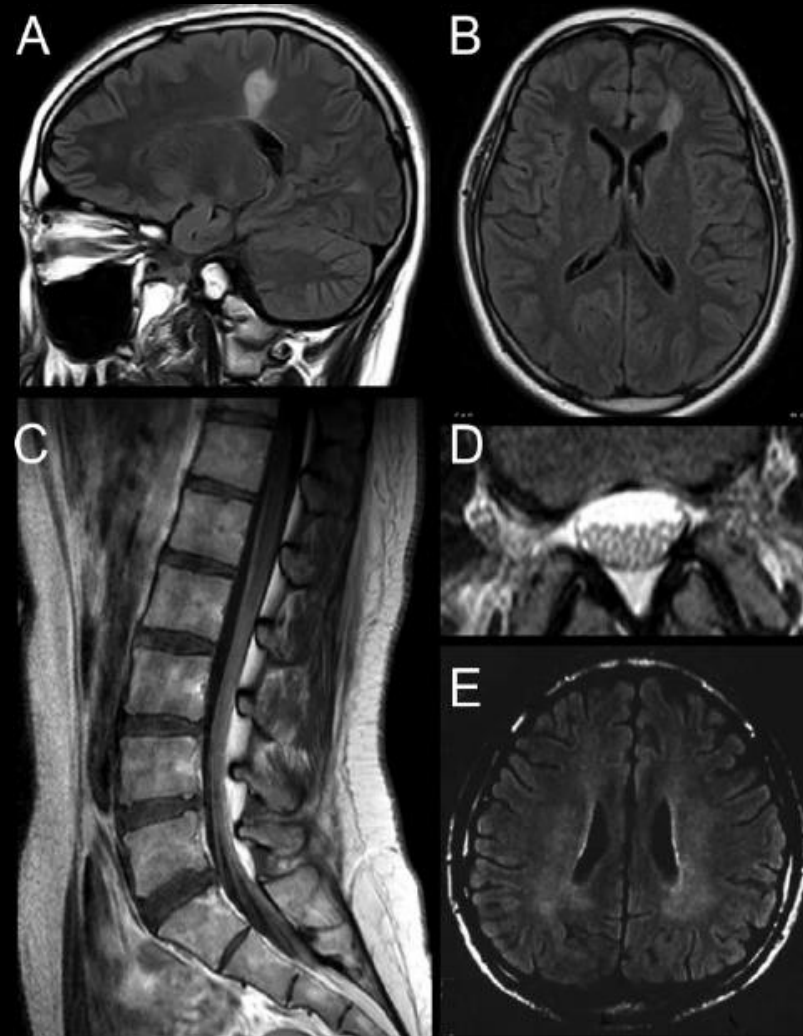
- 中枢神経 (CNS) や末梢神経 (PNS) の両方に生じる稀な炎症性脱髄症。
- Oligodendrocytes (CNS) と schwann cells (PNS) に共通の自己免疫抗体反応が機序とされている。
- 抗neurofascin抗体陽性になる頻度は高い。
- 稀な病態であり海外含めて報告は多くないが、MSに合併したものやGBSに合併するもの等、様々な病態の報告がある。

# Neurofascin

- 中枢神経・末梢神経の両方に共通して発現する細胞接着分子。
- immunoglobulin superfamily (IgSF)と呼ばれる。
- Subtype : 複数存在
  - NF155 ➡ brain white matter
  - NF186 ➡ diffuse both gray and white matter
- 抗NF155抗体や抗NF186抗体が両方関与するcrossed reactionやMS、Guillan-Barre Syndromeなどの脱髄性疾患でも陽性となり混在した報告例も。

# combined central and peripheral demyelination with anti-neurofascin antibody

- 画像所見：
    - CNS demyelination  
(ex. MS like lesions)
    - +  
PNS demyelination  
(ex. CIDP like lesions)
  - 抗neurofascin抗体陽性CCPD
    - 1) IV immunoglobulin
    - 2) plasma exchange
- が治療として効果的と報告



# 結語

- 抗neurofascin抗体陽性CCPDの1例を経験した。
- 中枢神経(CNS) + 末梢神経(PNS)の両方に生じる脱髄性疾患が疑われた場合、稀な疾患ではあるがcombined central and peripheral demyelination (CCPD) という病態を鑑別として考える必要がある。

# References

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- Tanja Adamovic et.al. Acute Combined and Peripheral Nervous System Demyelination in Children. Pediatneurool.2008 Vol39:307-316