Osmootne demüelinisatsiooni sündroom

Ksenia Rumjantseva

Haigusjuht

* 10.11.2018 ITK EMOsse oli kiirabiga toodud segasusseisundis ning desorienteeritud 48a. naispatsient, kes, abikaasa sõnul, oli tarvitanud alkoholi üle viimase 3 nädala, öösel tekkis krambihoog ning peale seda naise käitumine muutus (kodus pidevalt tühjast klaasist joonud vedelikku, pani suhu suvalisi mittesöödavaid asju
* Etanool plasmas neg
* Aju KT – mõõduka kroonilise entsefalopaatia foonil värsket kahjustust esile ei tulnud.
* Hospitaliseeritud siseosakonda alkoholdeliiriumiga, krampidega.

Siseosakonnas…

* Patsient püsivalt rahutu, agressiivne, delirioosne, mistõttu saab maksimaalsetes annustes sedatiivseid ravimeid (antud juhul Sol.Dextor ning Diazepam i/v ja tbl)
* 13.11.2018 patsiendil tõsine hüpernatreemia (**180** ↑↑ (136 .. 145 mmol/L) ning polüuuria; kerge hüpokaleemia - Dexdori kõrvaltoimed, see koheselt lõpetatud
* Sama päeva õhtul patsient raskes üldseisundis, kontakti ei ole ->>> üleviidud AIROsse edasiseks raviks

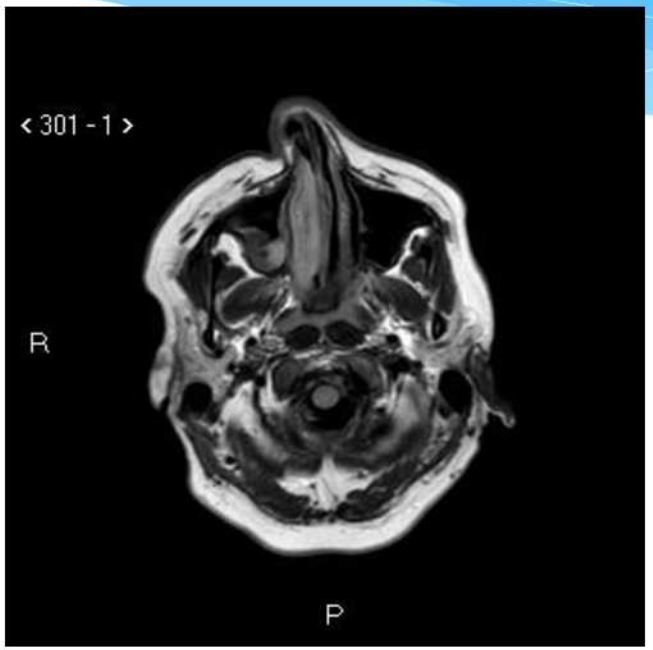
Anestesioloogia ja intensiivravi osakonnas….

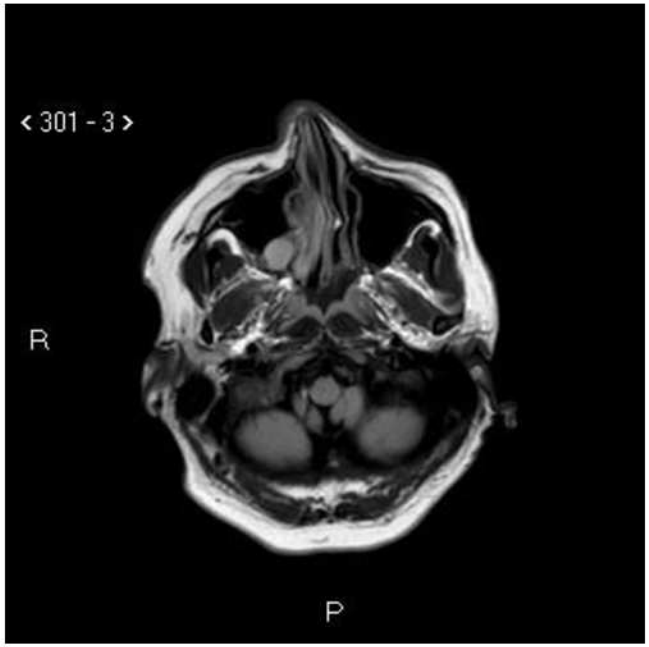
* 14.11.2018 hommikul Na* **167** ↑↑ (136 .. 145 mmol/L)
* 15.11.2018 hommikul Na* - 151 ↑, hüpokaleemia korrigeeritud
* 16.11.2018 - Na* 146 ↑
* 17.11.2018 – Na* 147 ↑
* 18.11.2018 – Na* 146 ↑

Viimane kontakt patsiendiga 13.11.2018 siseosakonnas →

kahtlus mittekonvulsiivsele epilepsiale. Difdgn: hüpernatreemiast tingitud muu orgaaniline ajukahjustus → MRT peaajust (6 päeva peale sümptomite tekkimist)

FLAIR

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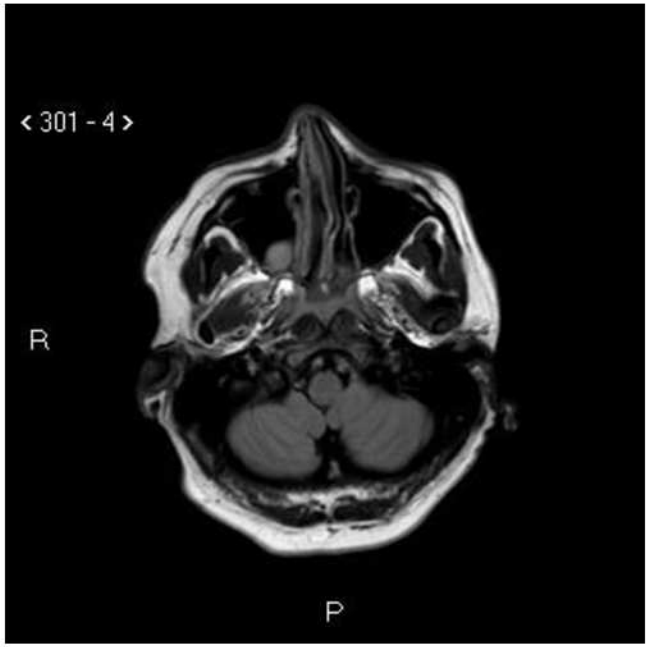
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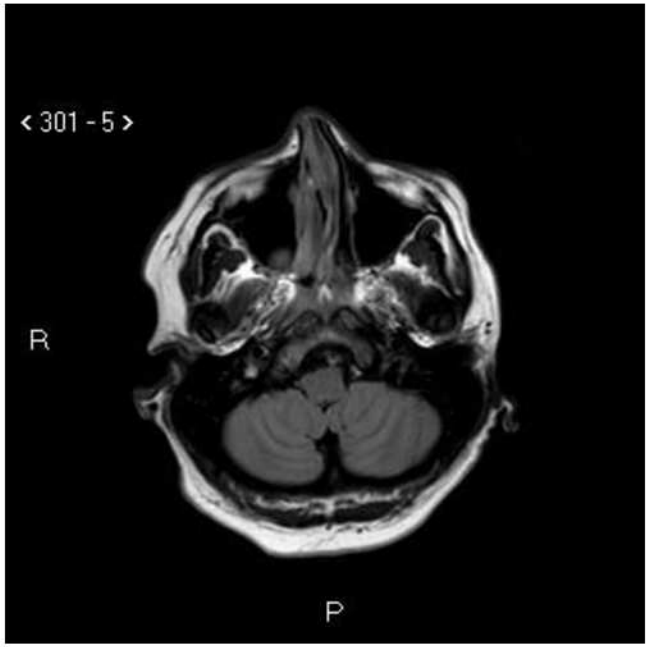
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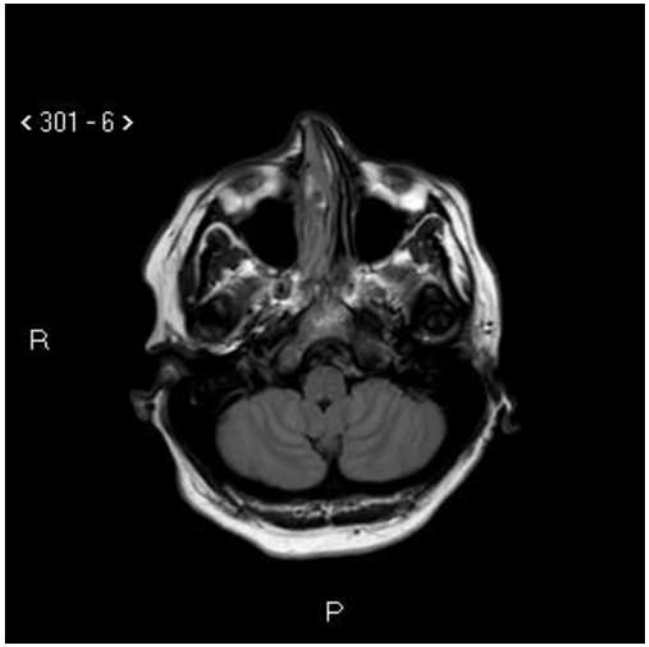
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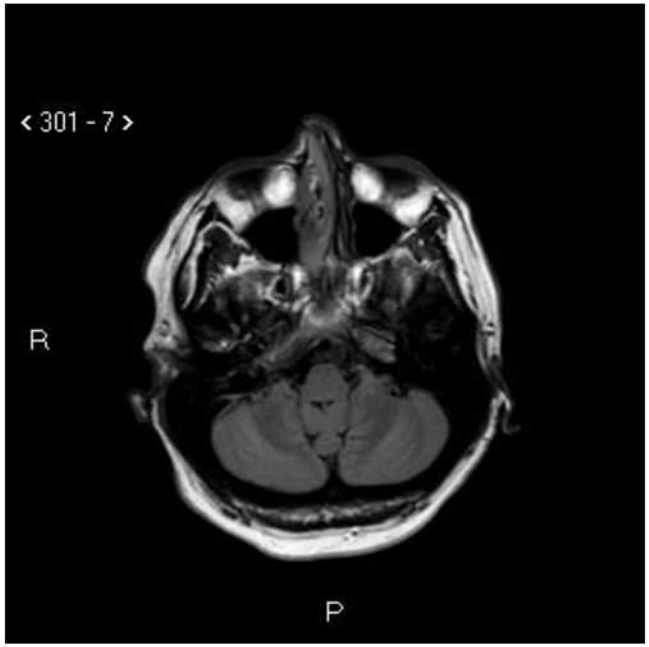
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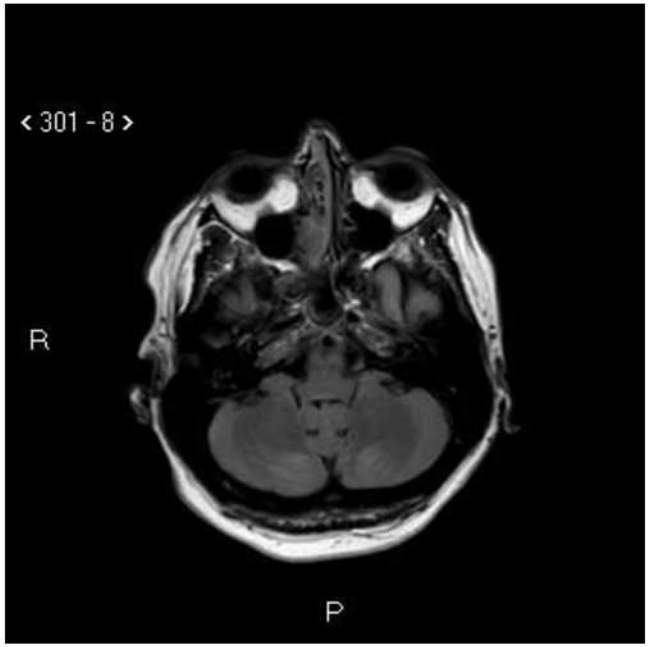
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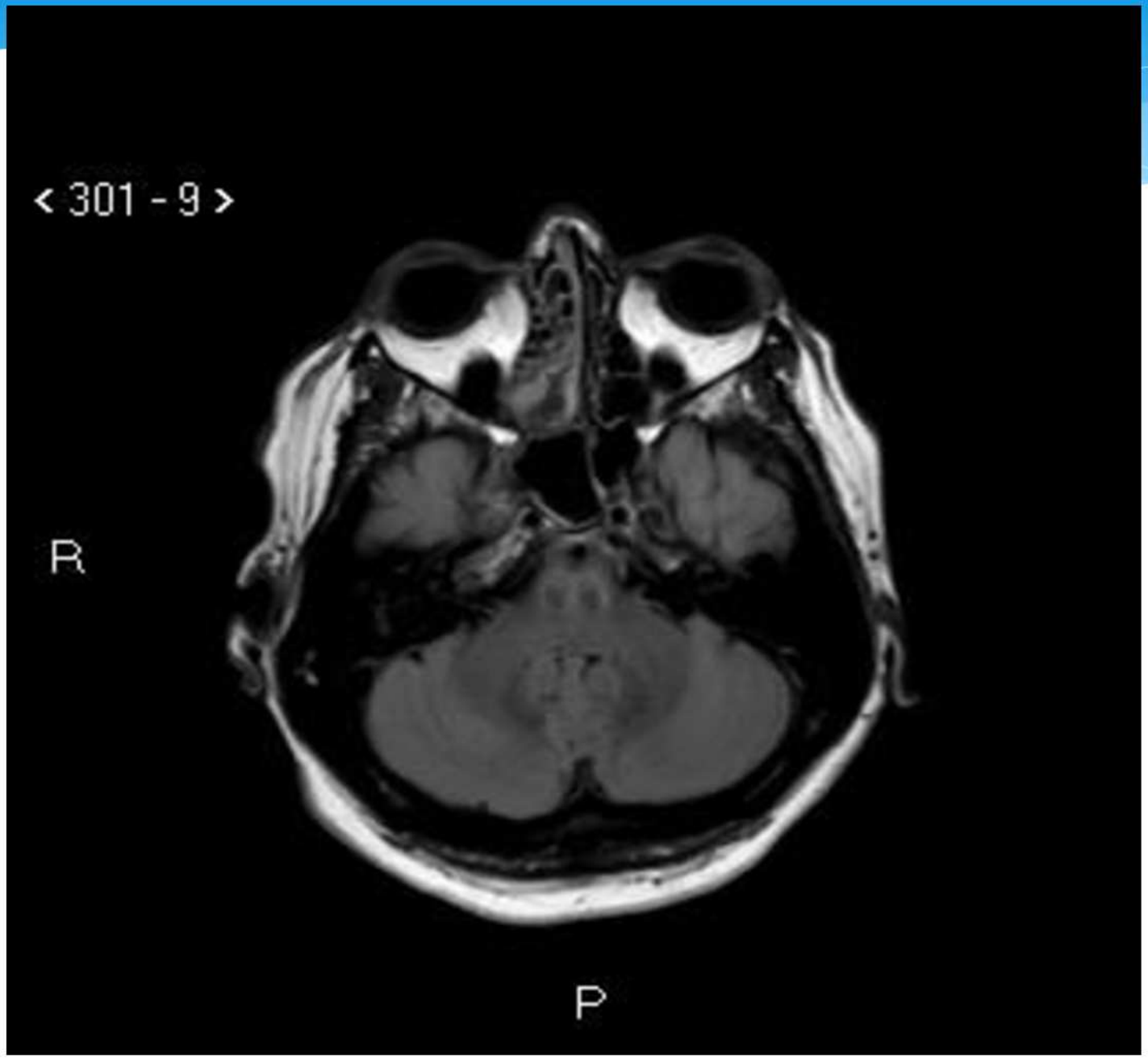
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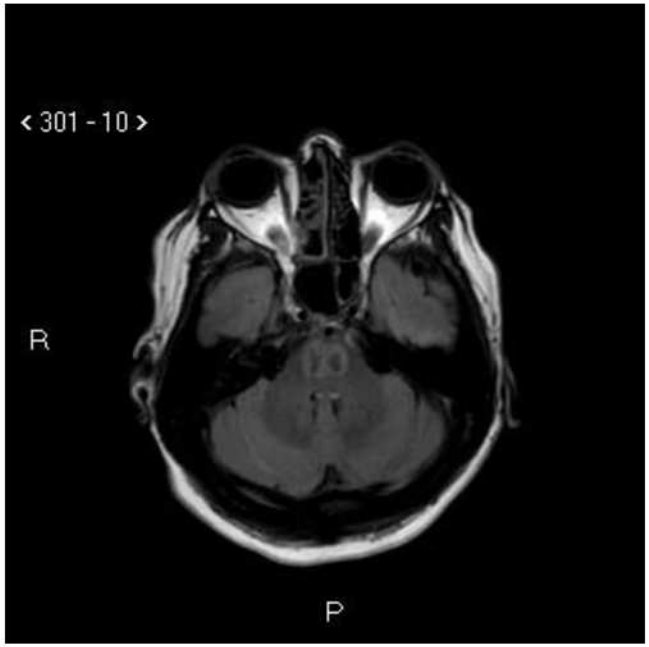
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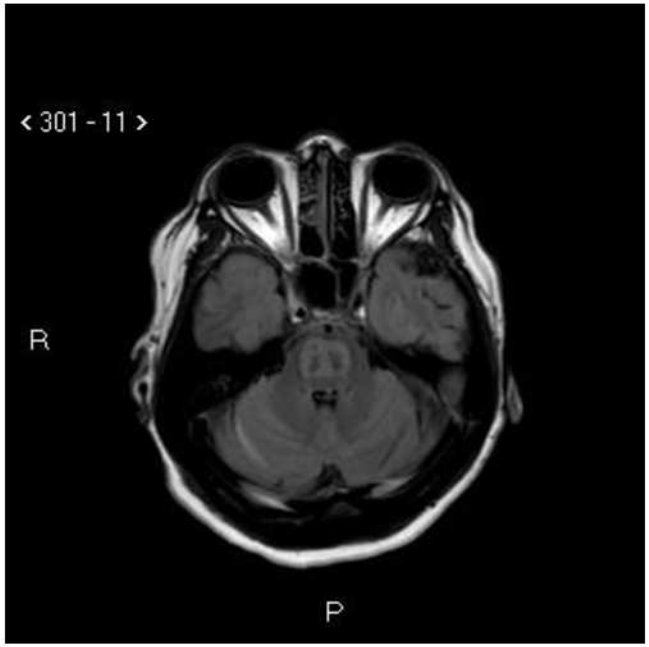
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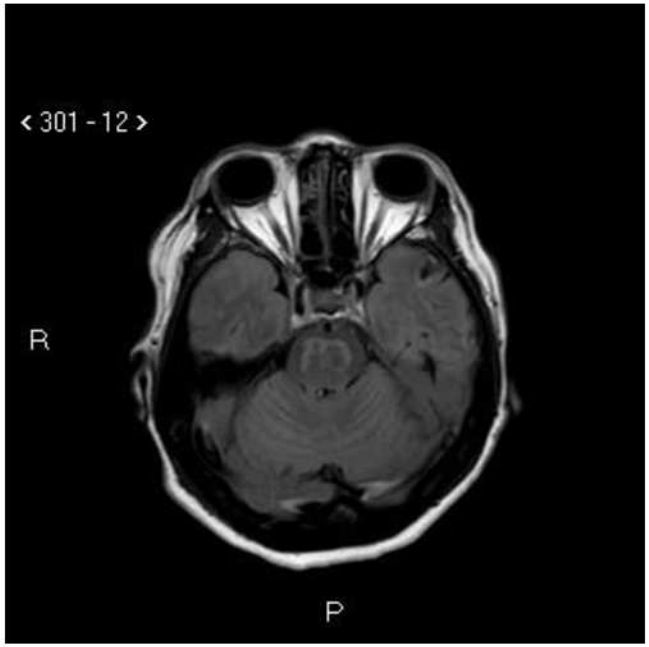
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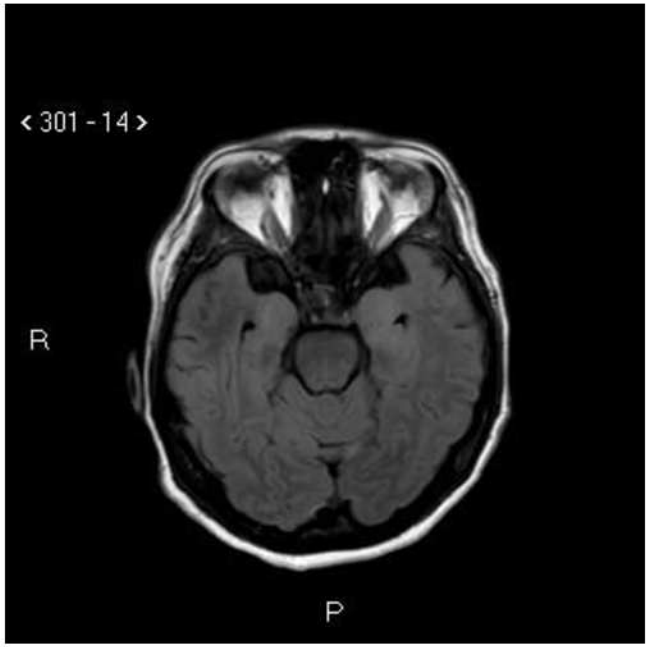
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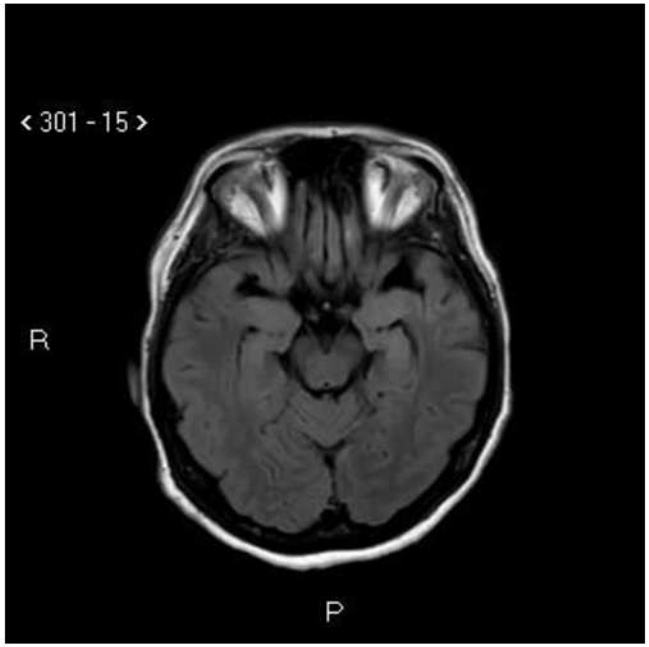
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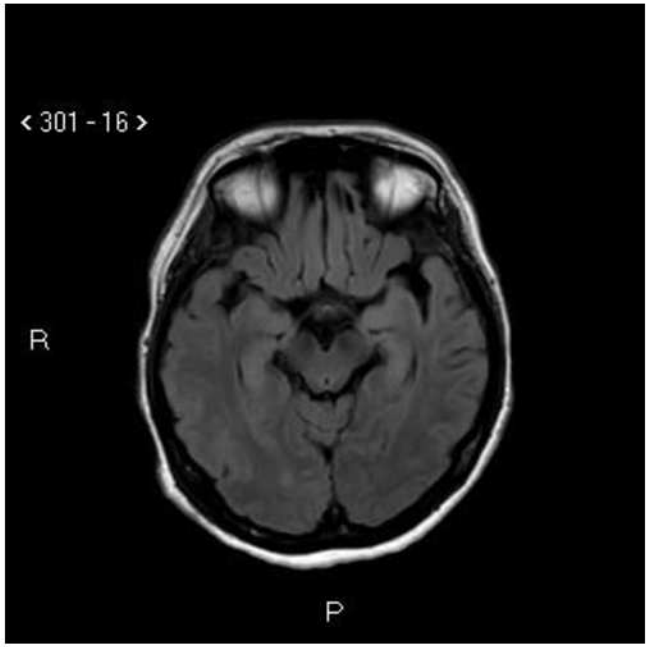
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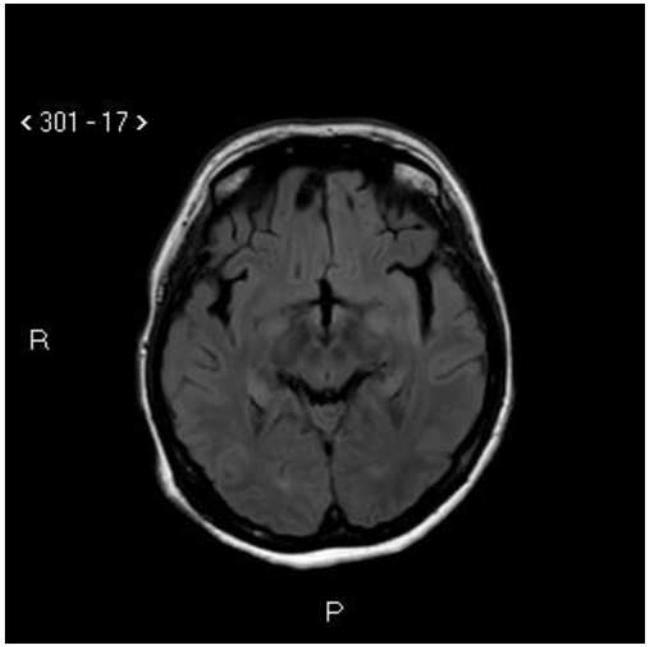
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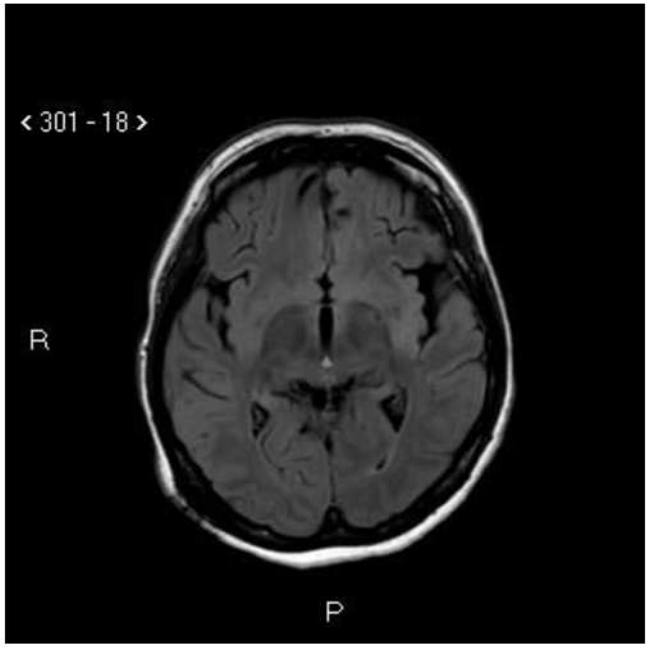
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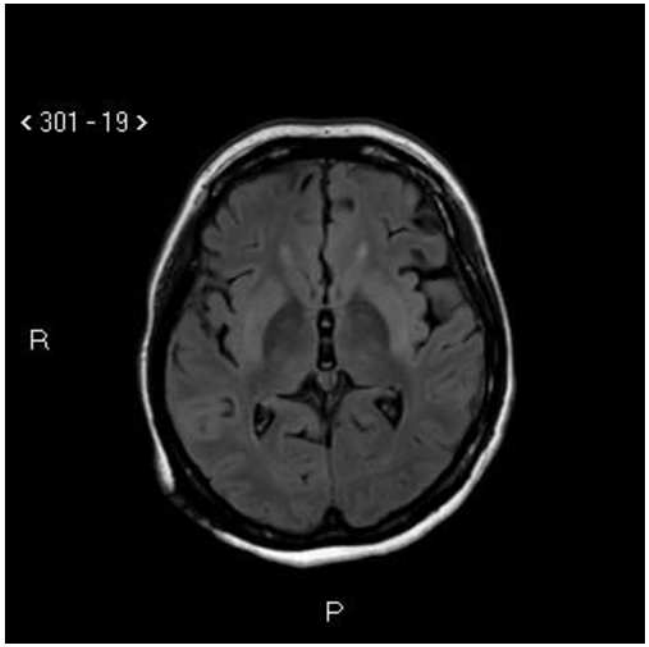
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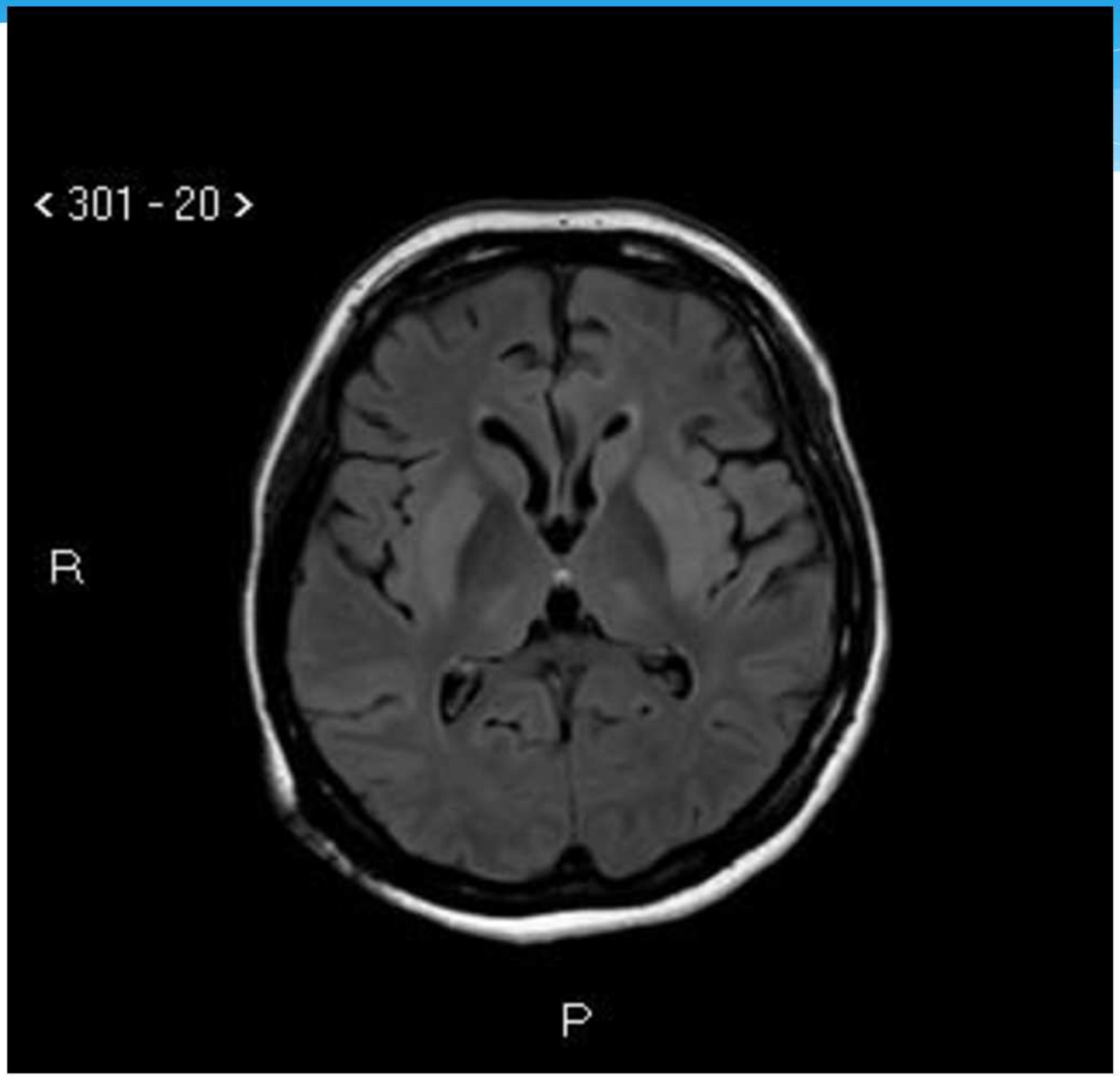
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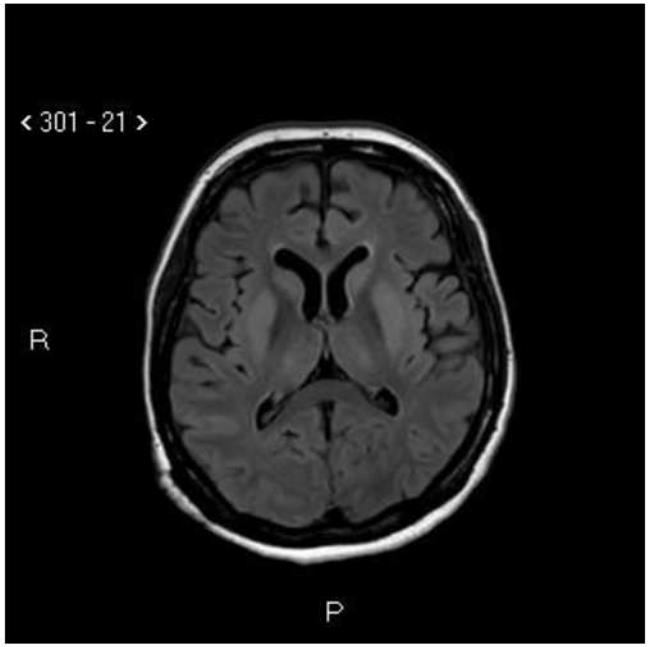
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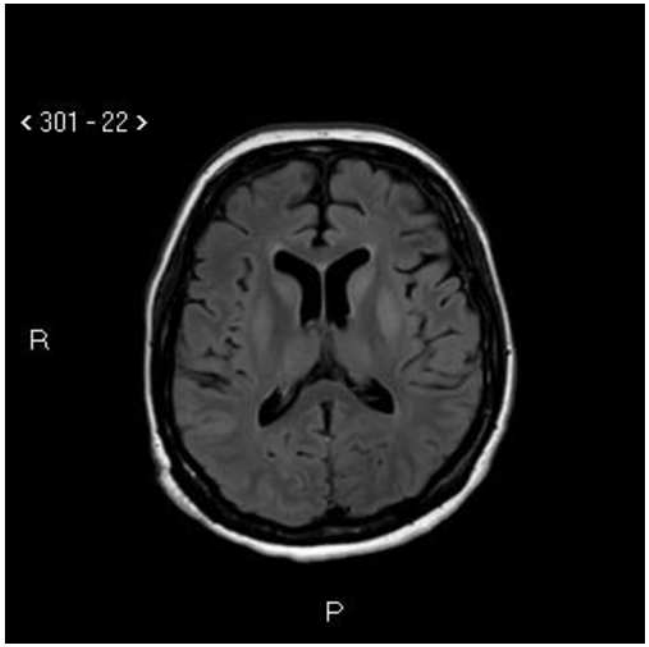
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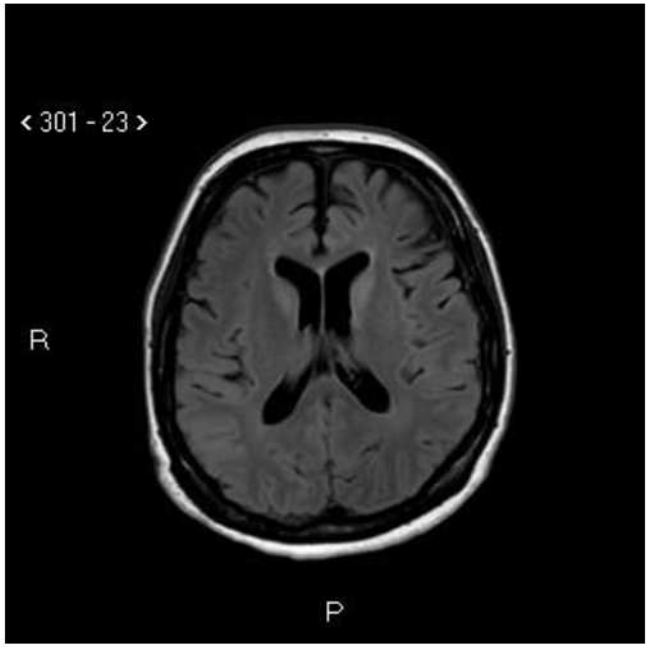
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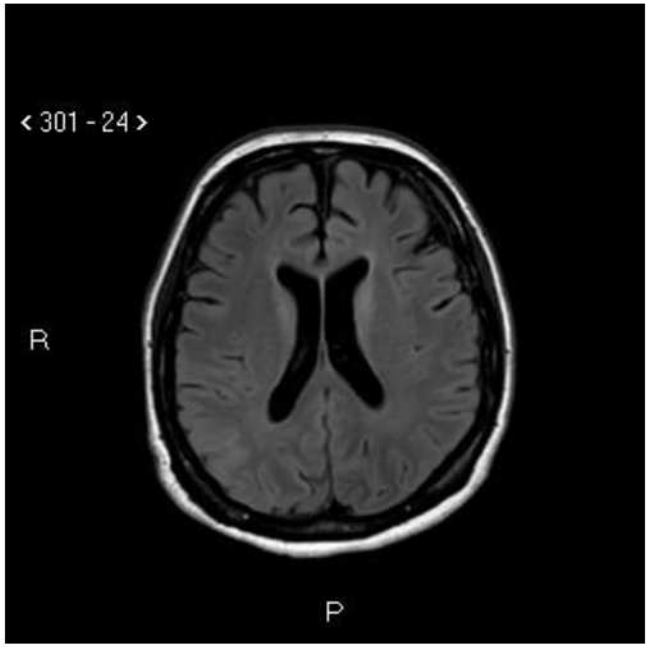
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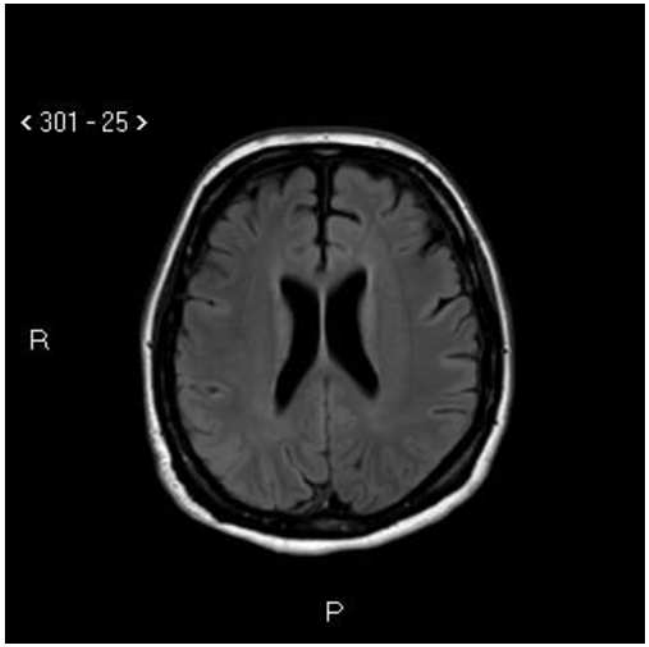
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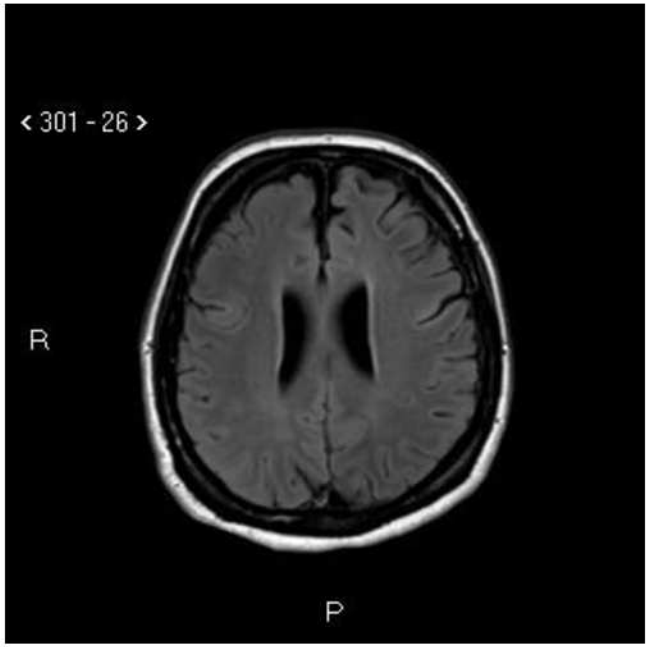
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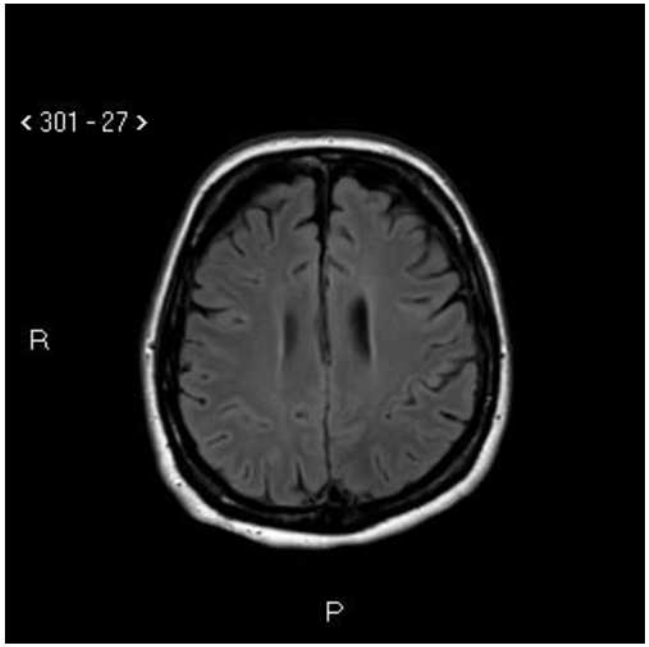
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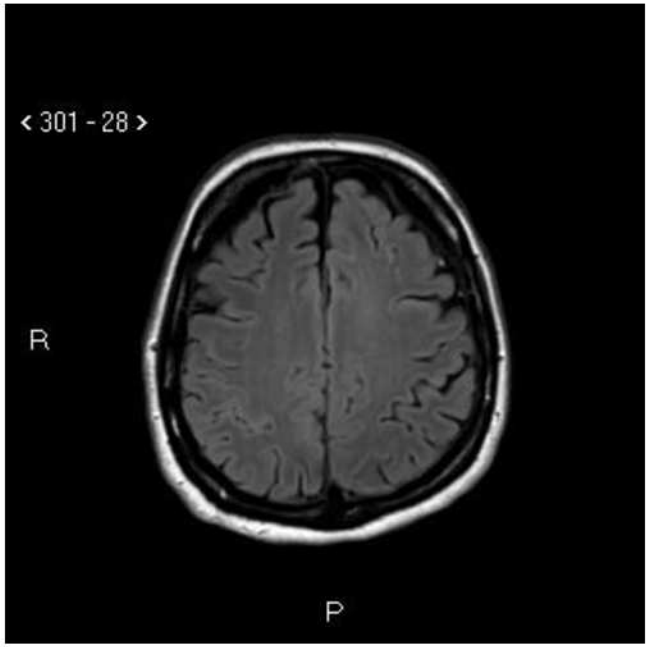
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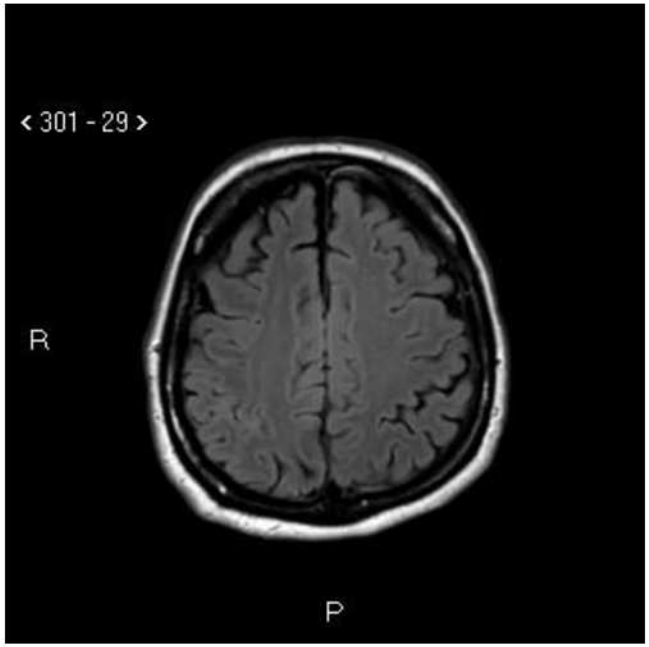
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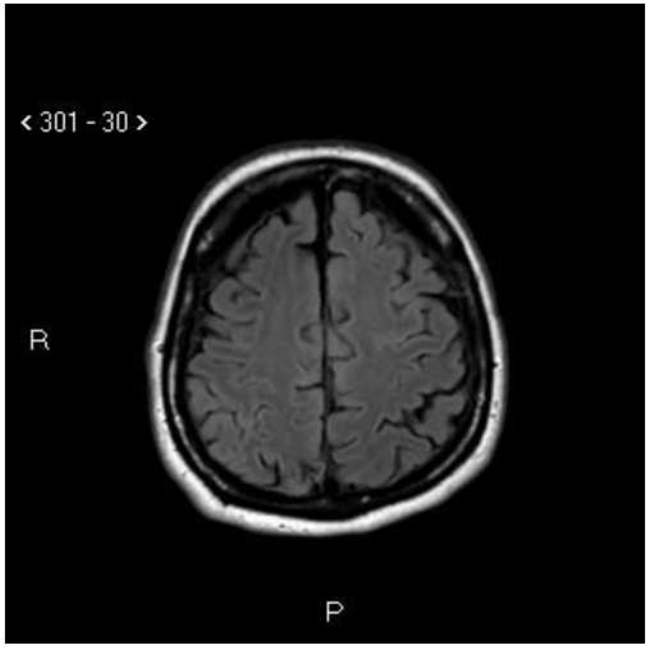
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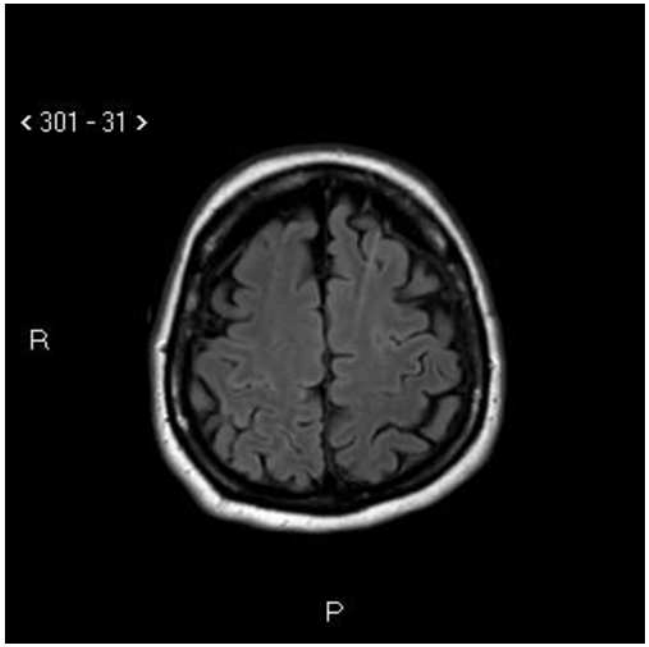
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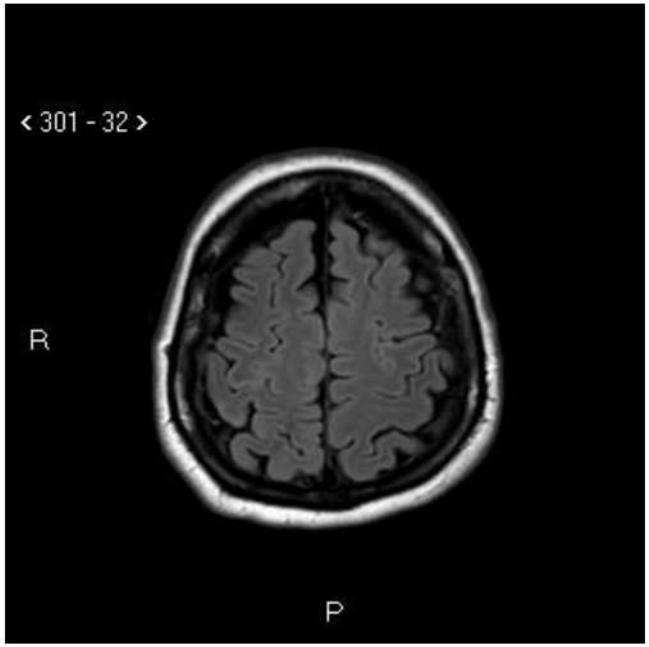
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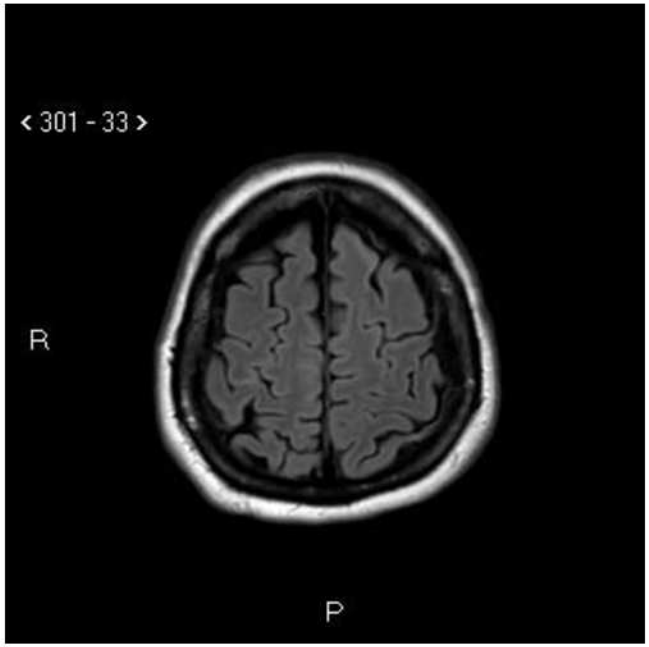
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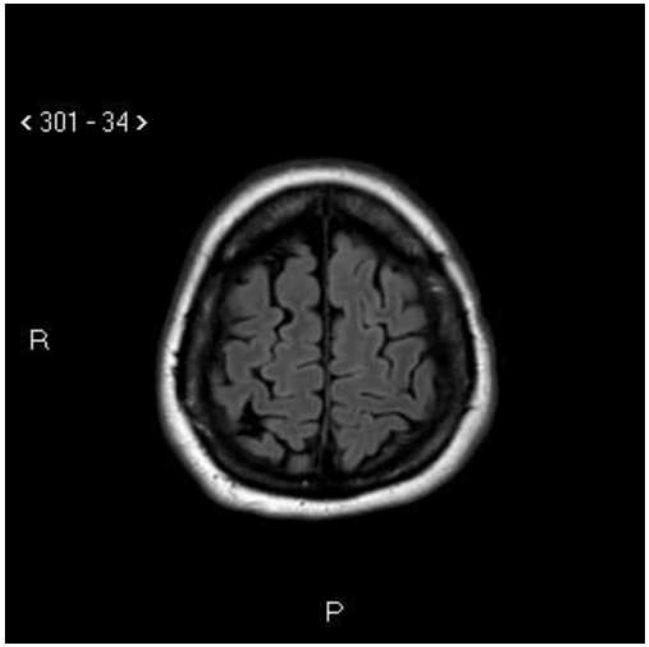
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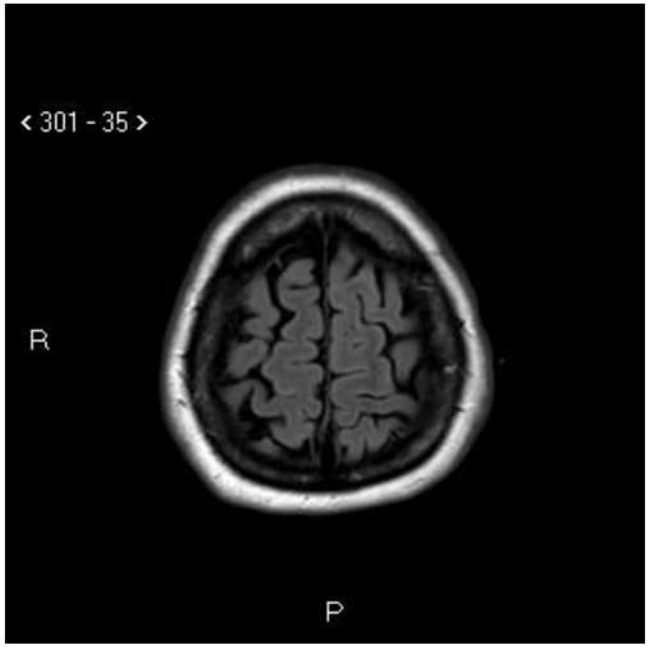
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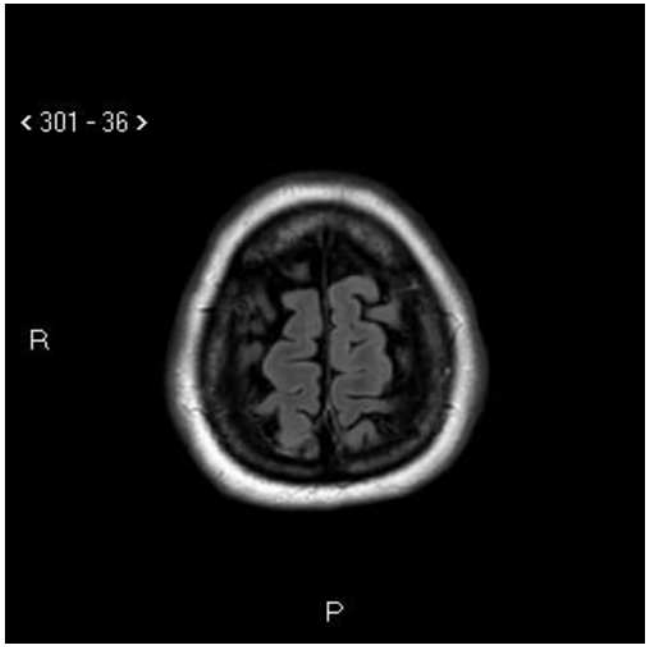
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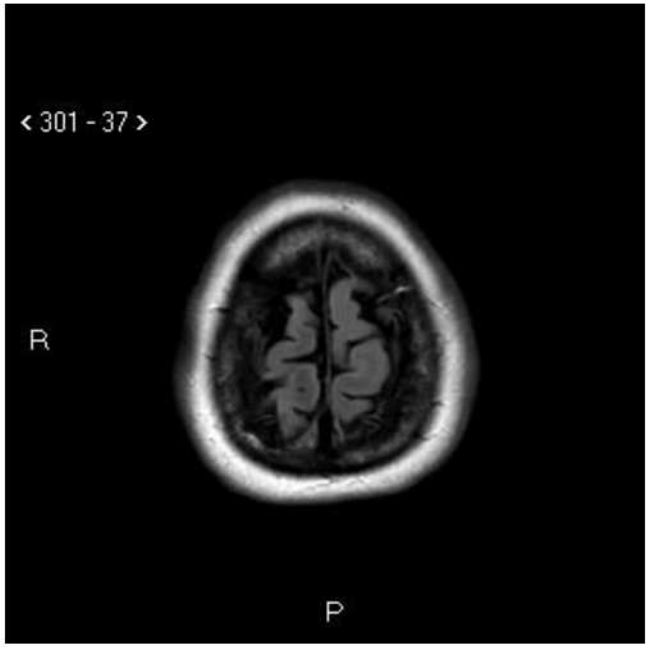
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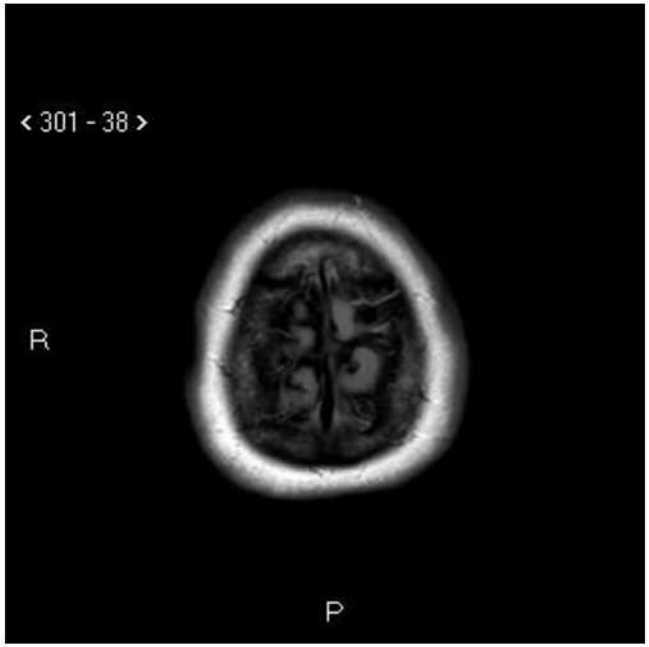
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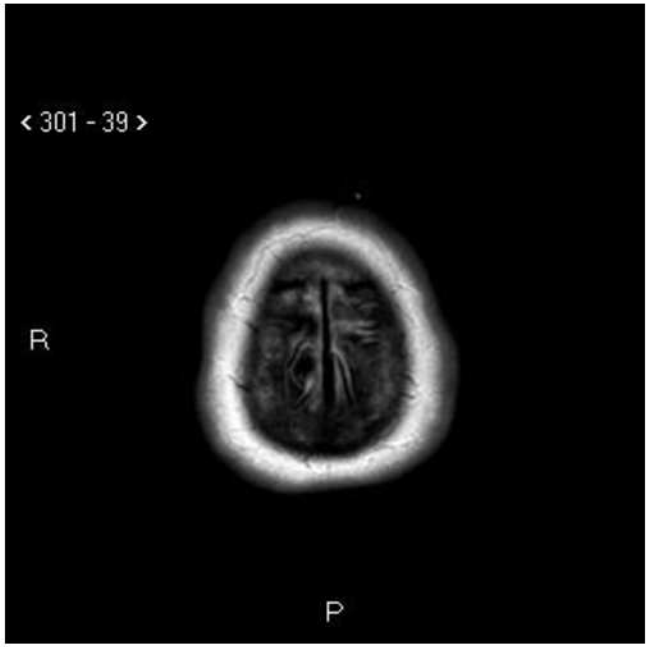
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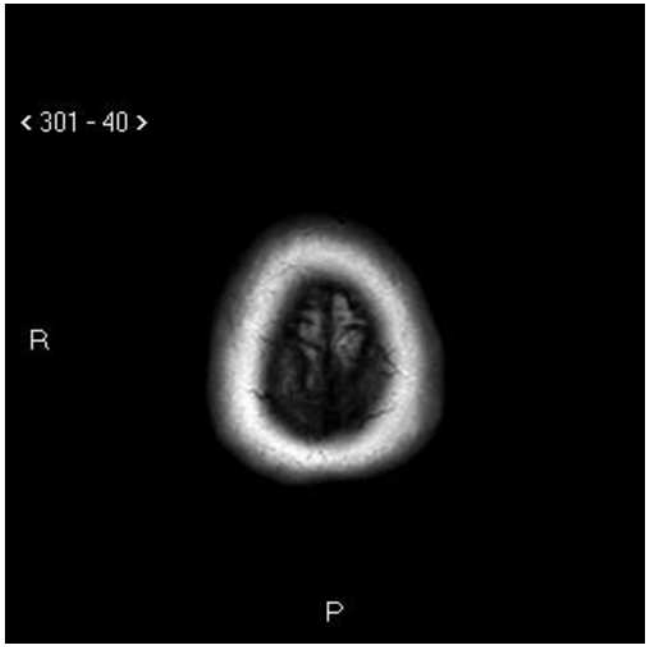
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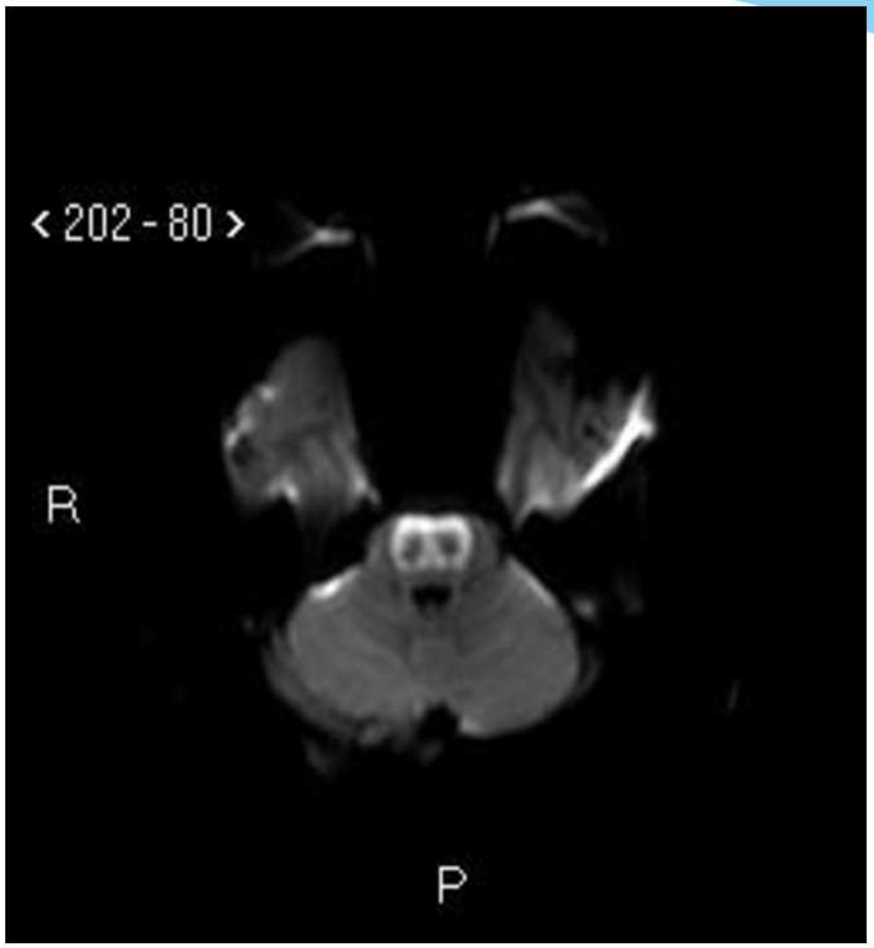

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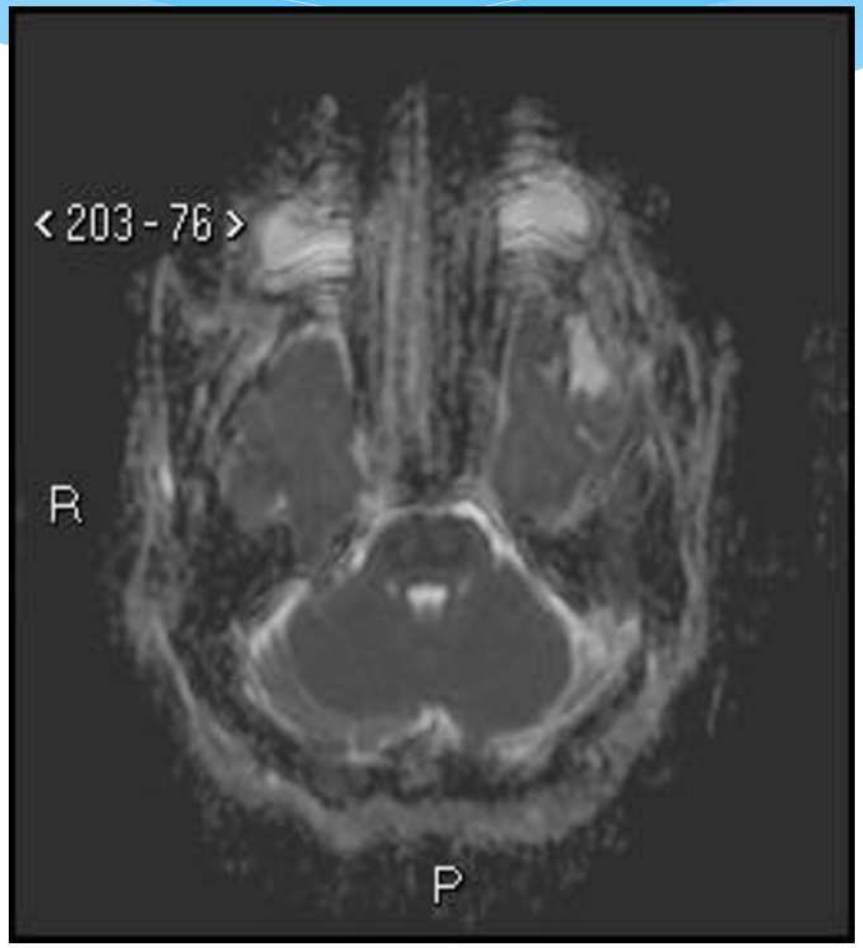


Al. 13.11.2018 patsient on koomas, kontakti temaga ei ole

25.11.2018 ex.letalis

Osmootne demüelinisatsiooni sündroom

Müelinolüüs:

I. Tsentraalne pontiinne (CPM)

* Ponsis

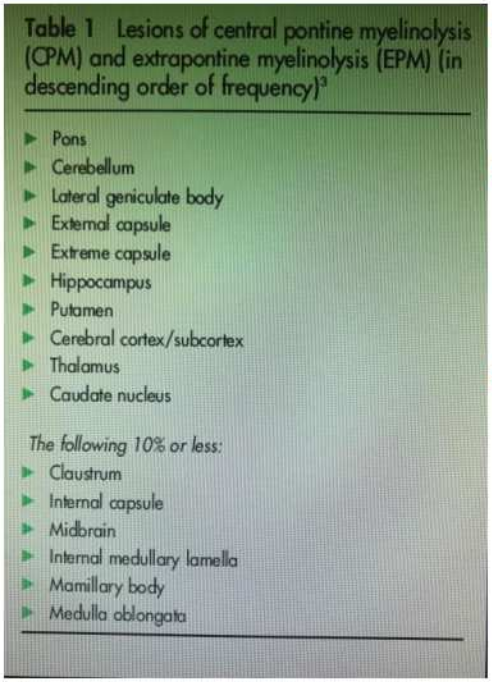


Table 1 Lesions of central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM) (in descending order of frequency)[3]

- Pons
- Cerebellum
- Lateral geniculate body
- External capsule
- Extreme capsule
- Hippocampus
- Putamen
- Cerebral cortex/subcortex
- Thalamus
- Caudate nucleus

The following 10% or less:

- Claustrum
- Internal capsule
- Midbrain
- Internal medullary lamella
- Mamillary body
- Medulla oblongata

II. Ekstrapontiinne

* Basaalganglionid ning aju valgeaine
* Harvem: ajukorteks, hippocampus, coprus geniculatum laterale
* Tavaliselt koos tsentraalse müelinolüüsiga, samas võib olla ka isoleerituna

CPM soodustavad faktorid

* • Krooniline alkoholism
* • Puudulik toitumine
* • Pikaajaline diureetikumide kasutamine
* • Psühhogeenne polüdipsia
* • Põletushaavad
* • Maksahaigused (maksapuudulikkus)
* • Maksatransplantatisoonijärgselt (cyclosporine)
* • Hüpofüüsiop.-ijärgselt

Kliiniline kulg

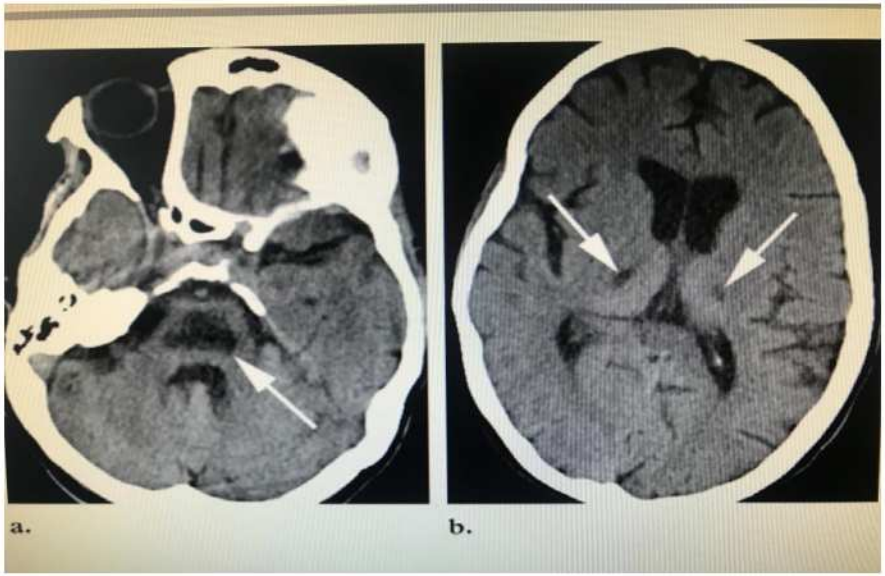
* Tavaliselt seotud hoopis hüponatreemiaga (selle kiire korrigeerimisega)
* Pt-l tõsised elektrolüütide tasakaalu häired → krambihood, entsefalopaatia
* Normonatreemia saavutamise pärast pt. seisund paraneb 48-72t jooksul, kuid mõni päev pärast hakkab kiiresti halvenema (lisanduvad düsartria, düsfaagia, kvadriparees (hiljem spastiline) jne –

→kooma/deliirium

* Prognoos varieeruv, ei ole ennustatav kliiniliselt ega radioloogiliselt (täielikust paranemisest surmani)

Imaging (KT,MRT)

* KT vähem tundlik kui MRT
- hüpodensiivsed alad ponsis (basaalganglionid, talamus), ei põhjusta mass-efekti

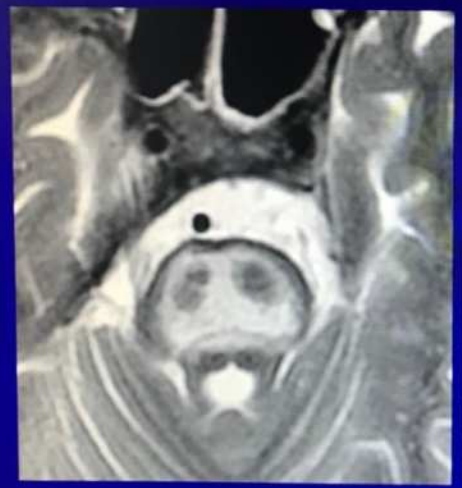


MRT

* Muutused tavaliselt ilmestuvad 10-14.päeval peale sümptomite tekkimist
* Klassikaline tunnus: iseloomulikud sümeetrilised T2 hüperintensiivsed alad ponsis („trident sign“ „piglet sign“
* Ekstrapontiinselt bilateraalsed sümeetrilised T2 hüperintensiivsed alad basaalganglionites, talamustes)
* Ei kontrasteeru

Piglet sign

Face of the Piglet sign:
Central Pontine Myelinolysis



Face of the Piglet sign

The pons with its characteristic appearance resembles the snout, ICAs and the fourth ventricle constitute the eyes and mouth of the piglet respectively [5, 6].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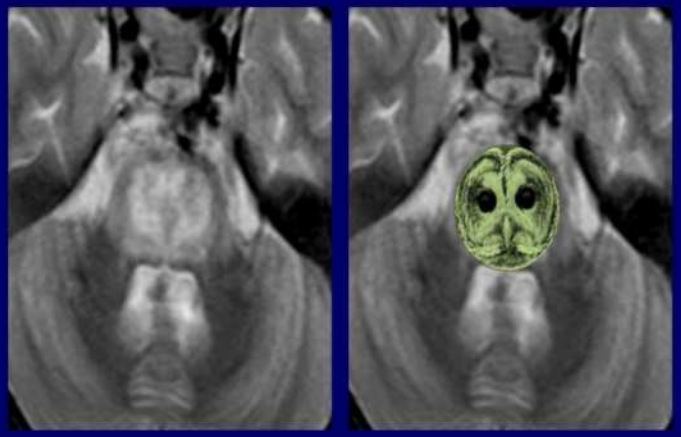
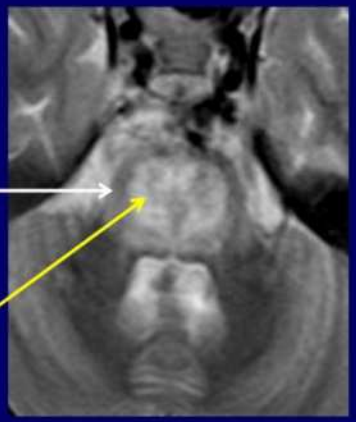
Owl's eye sign

Owl's eye appearance:
Central Pontine Myelinolysis



Owl's eye appearance

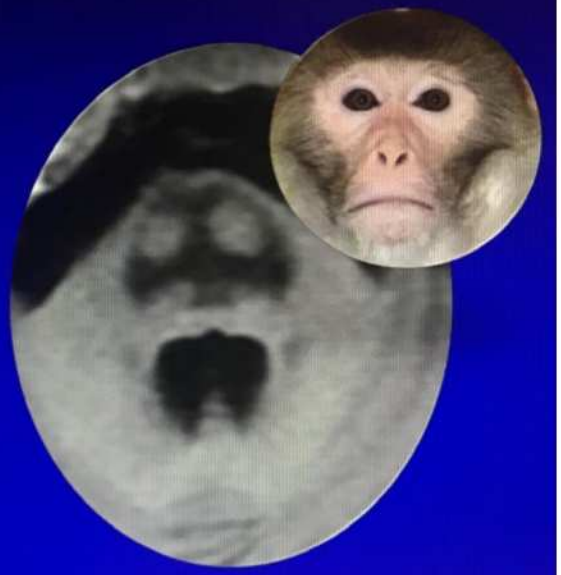
- Central Pontine Myelinolysis affects the basal pons with sparing of the descending corticospinal tracts as well as the peripheral pontine tissue.
- The corticospinal tracts may appear as preserved islands within a zone of hyperintense pontine demyelination giving an Owl's eye appearance [4].



Monkey sign

Monkey sign: Central Pontine Myelinolysis

- Corresponding T1-weighted MR images may show this characteristic pattern of signal alteration in the basal pons as resembling the face of a monkey - also referred to as the 'Monkey sign' of CPM [7].



DDx

1. A. basilaris'e harude infarktid
- tavaliselt asümmetrilised muutused ponsis
2. Ponsi haarav demüeliniseeruv haigus(SM)
- asümeetrilised muutused
3. Ponsi neoplasm (glioom)
- lapsed, noored täiskasvanud
- pons suurenenud, T2 hüperintensiivne

Kasutatud kirjandus

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