

第34回神経放射線ワークショップ

国立病院機構 大阪医療センター
藤原 拓也

症例：55歳女性

【主訴】 左片麻痺 left hemiparesis

【現病歴】

突然左上下肢に力が入らなくなり、当院に救急搬送された。

A 55 year-old woman was admitted to our hospital with a history of sudden-onset left lower and upper limb weakness.

【既往歴】

幼少時より原因不明の眼球運動障害あり。

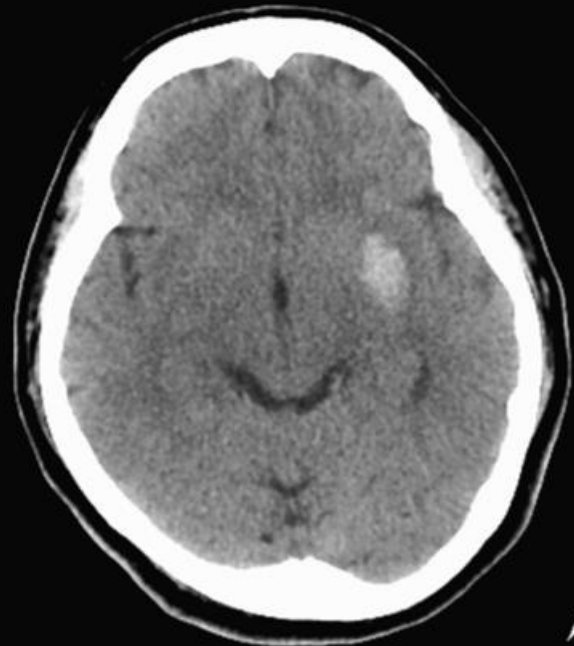
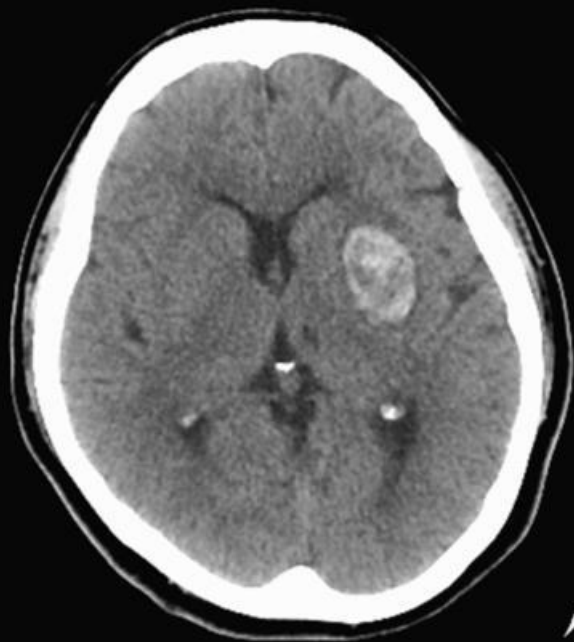
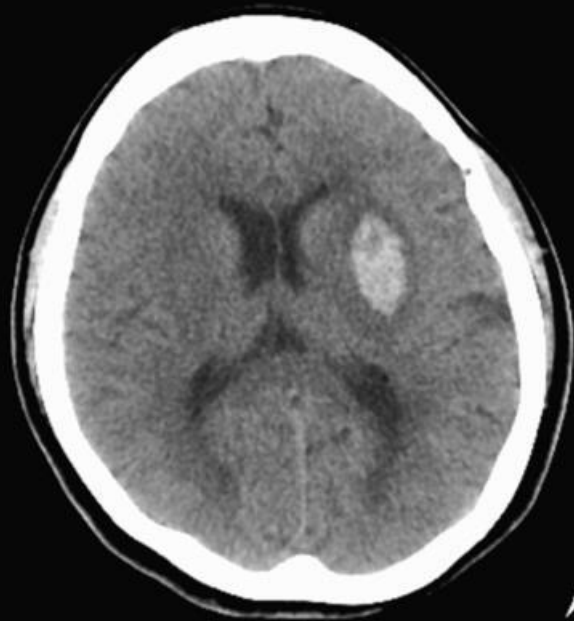
She had disturbance of eye movements with unknown causes from childhood.

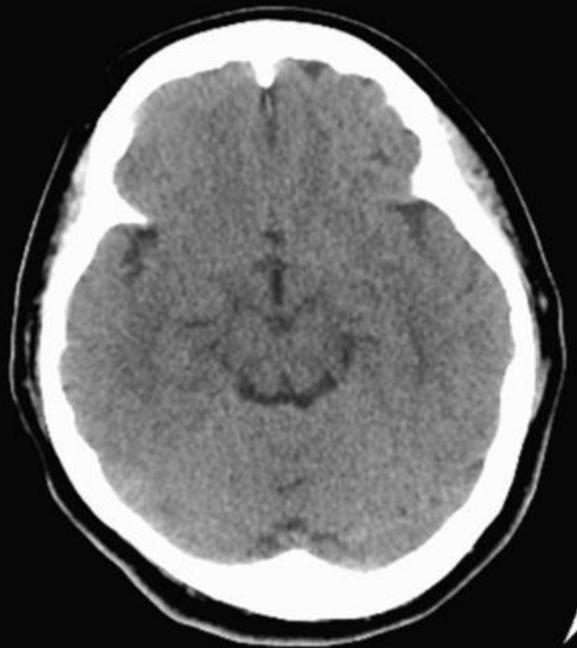
【現症】

左片麻痺、水平方向の注視麻痺あり。

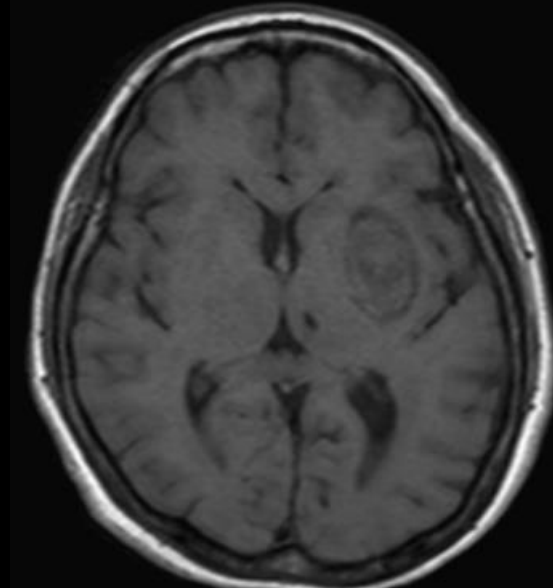
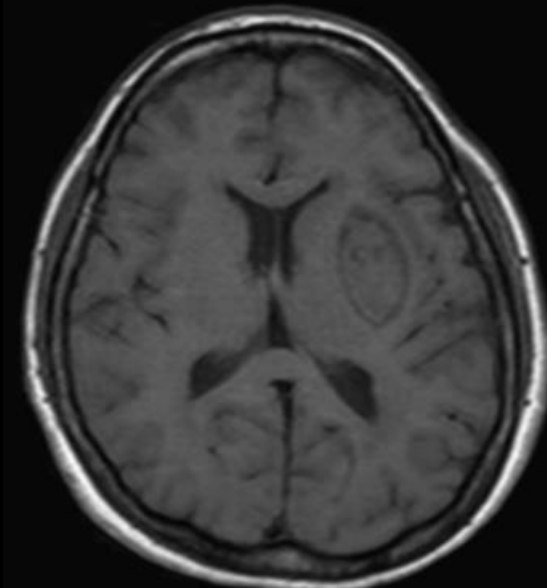
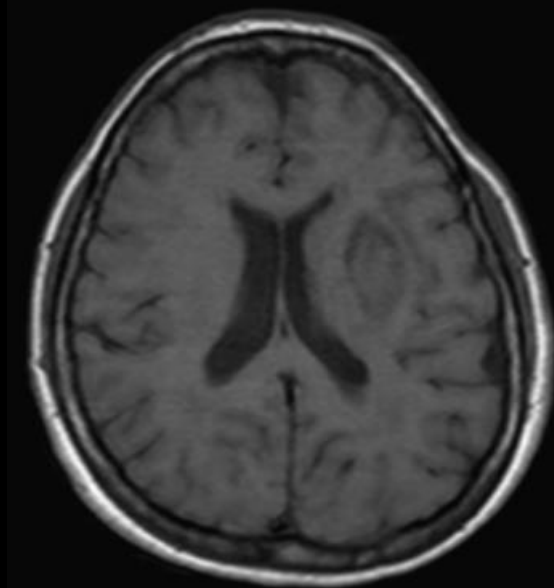
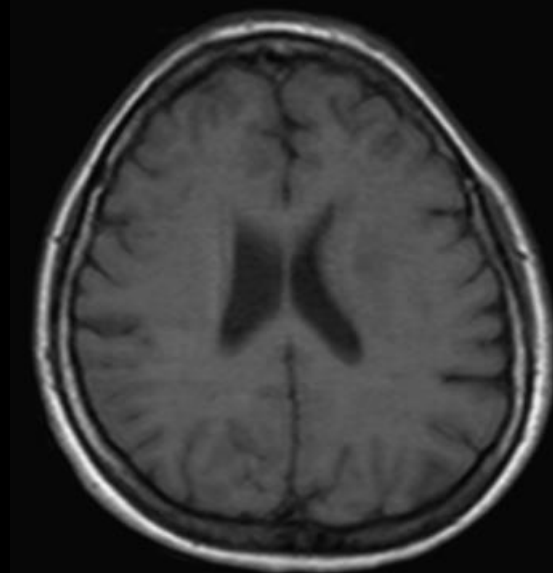
She had left hemiparesis and horizontal gaze paralysis.

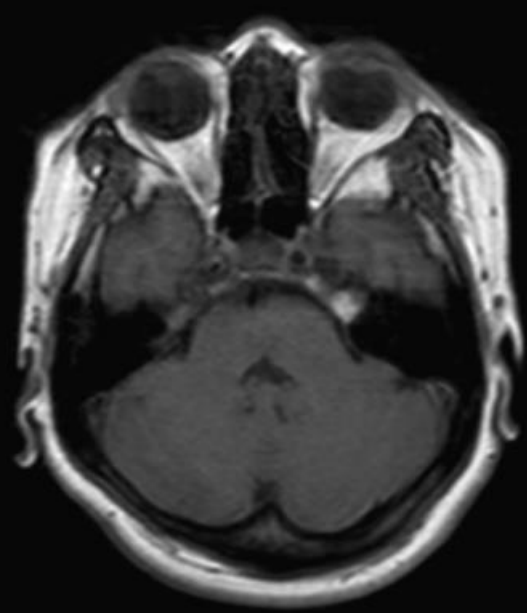
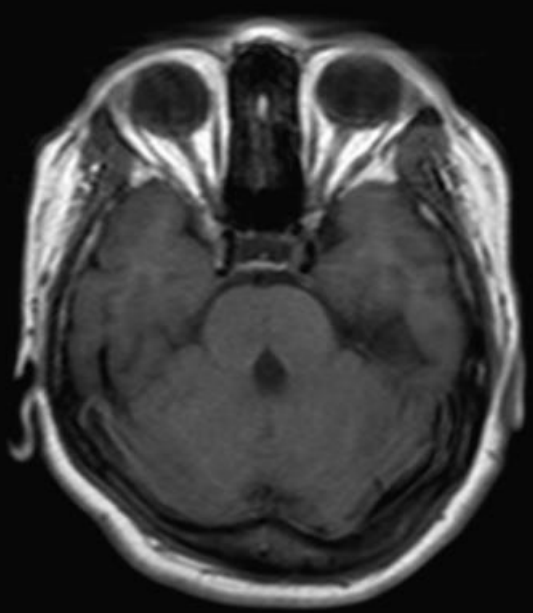
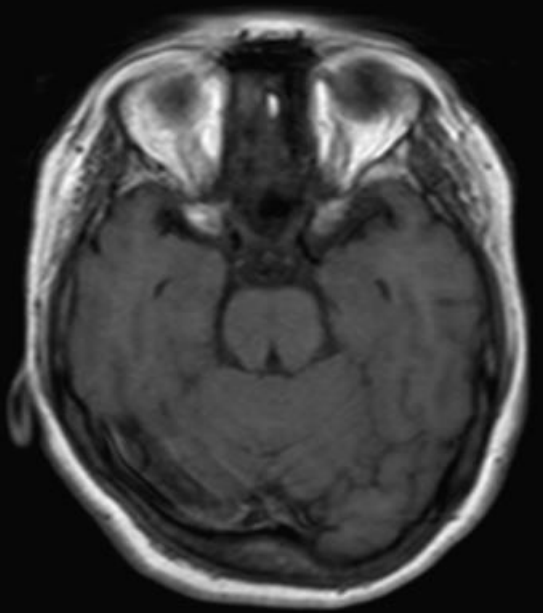
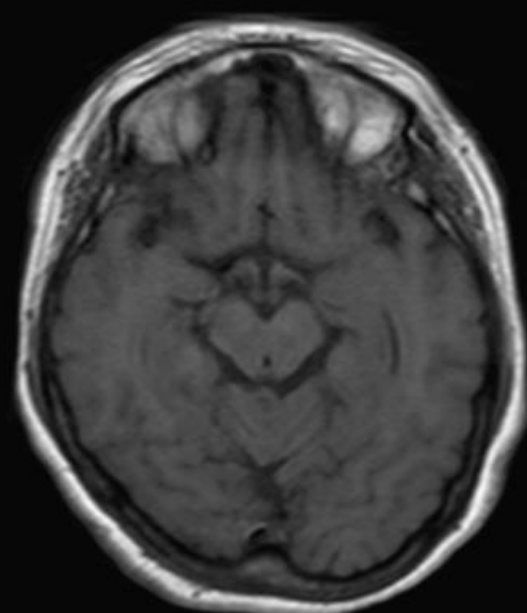
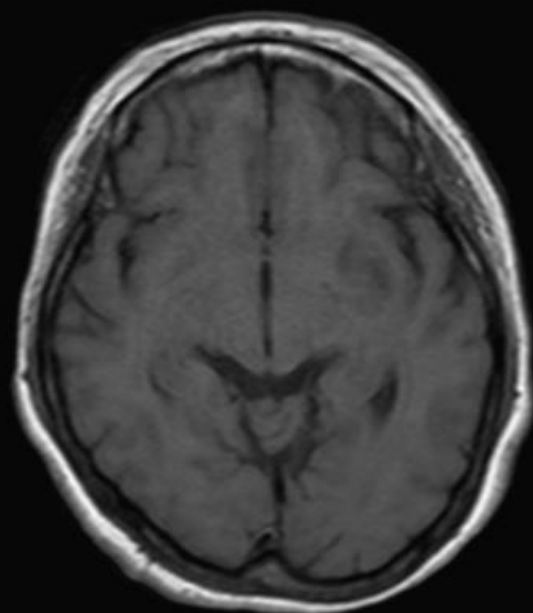
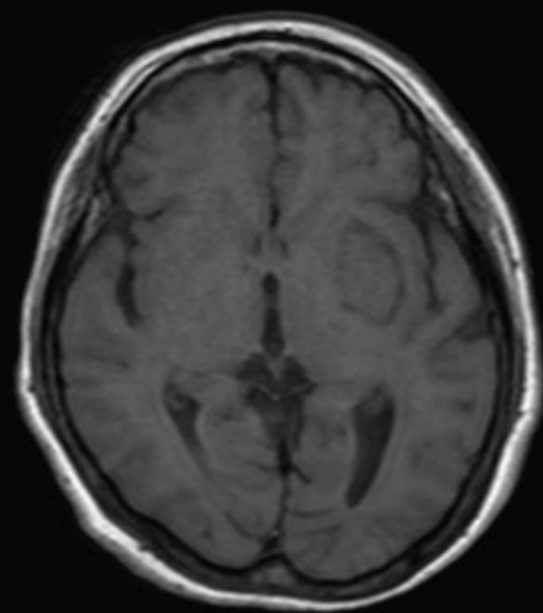
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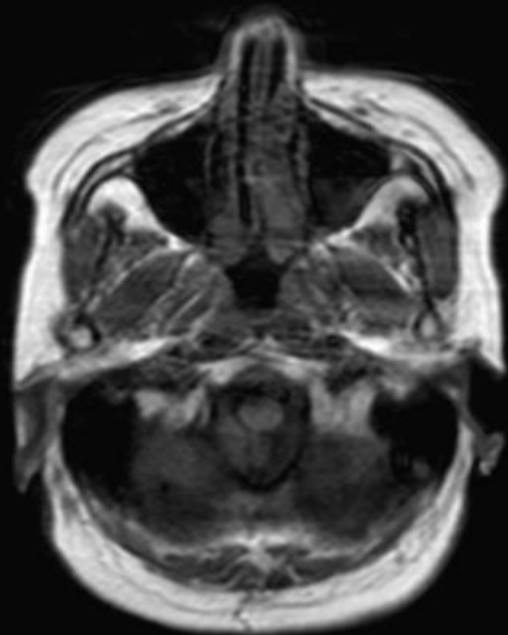
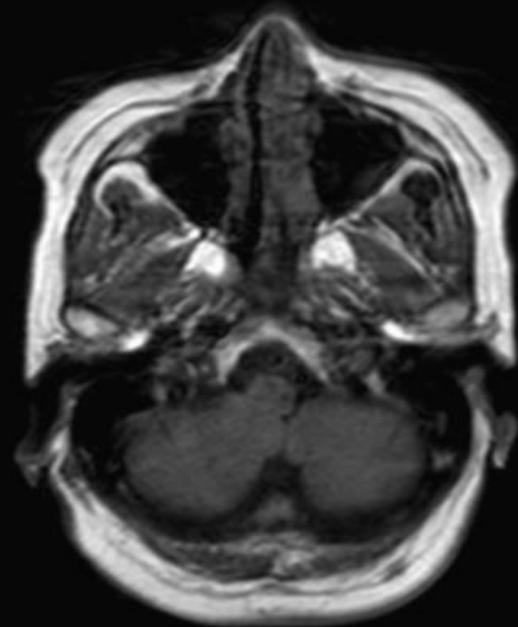
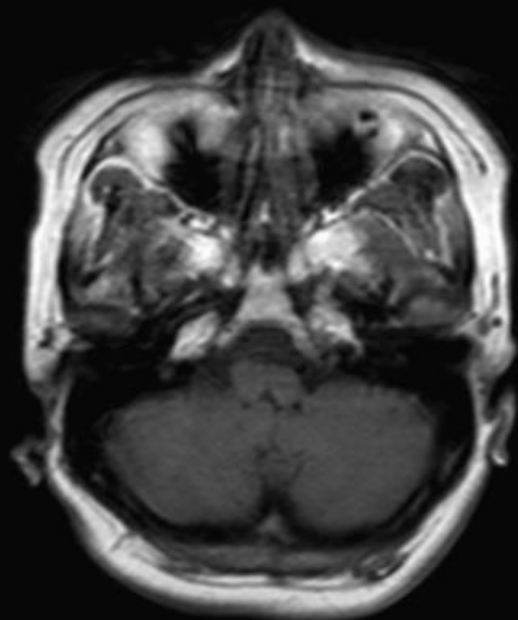
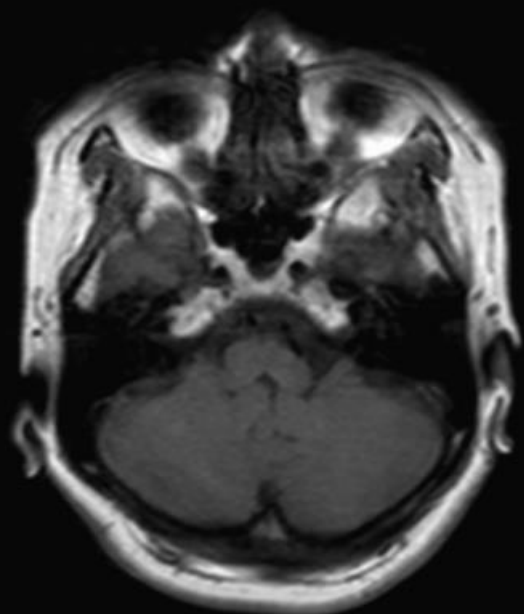




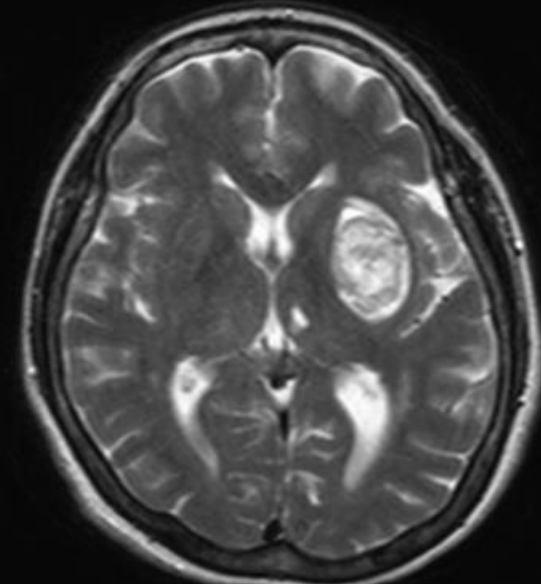
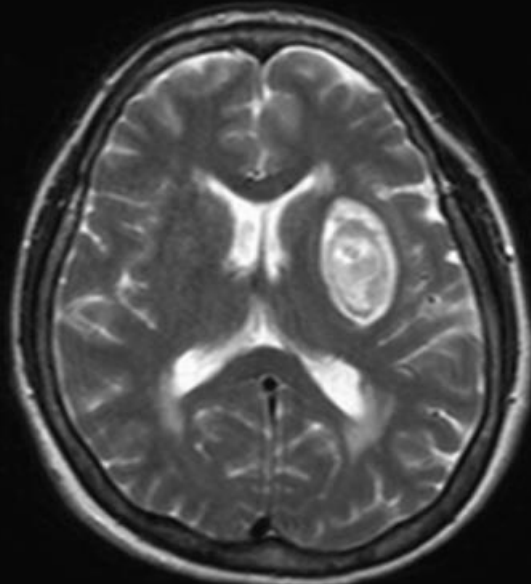
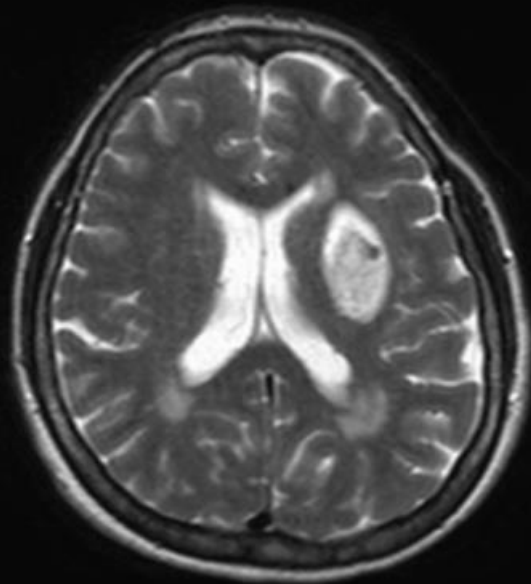
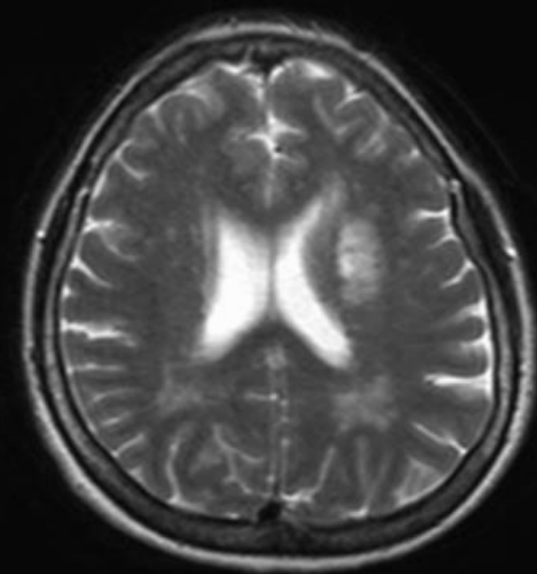
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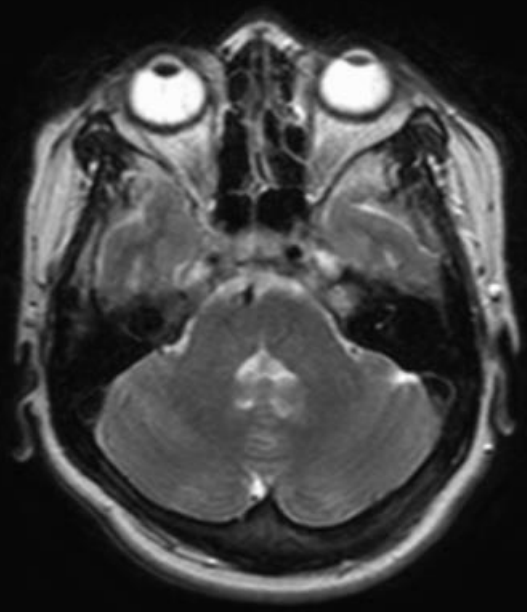
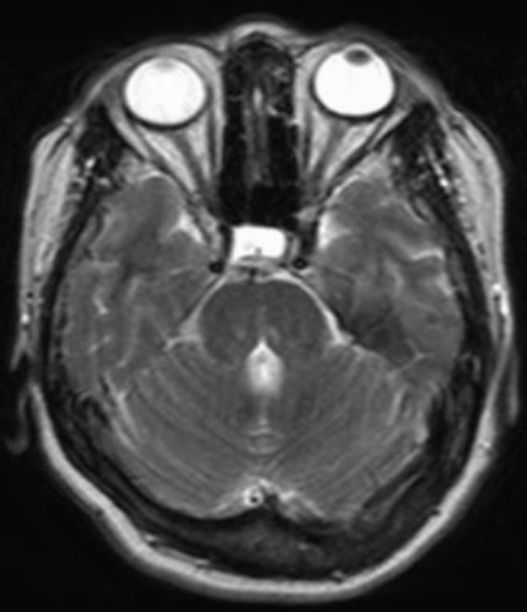
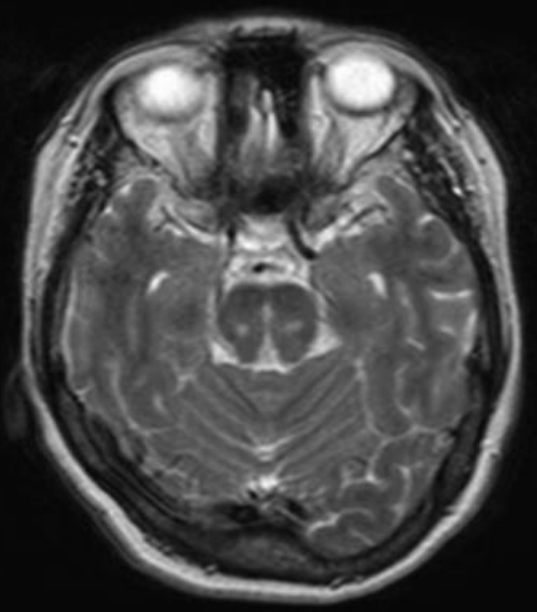
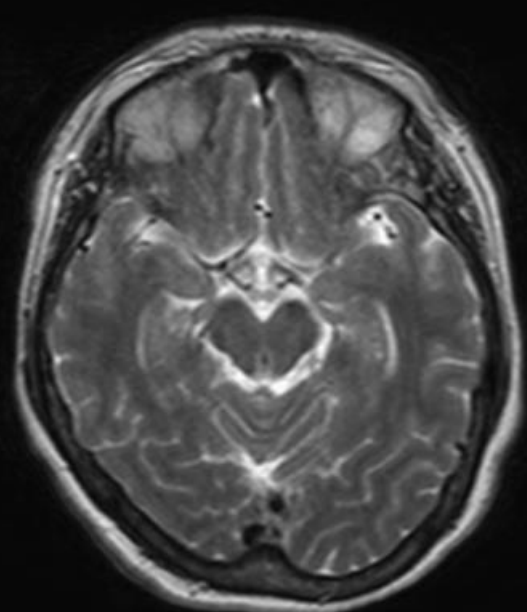
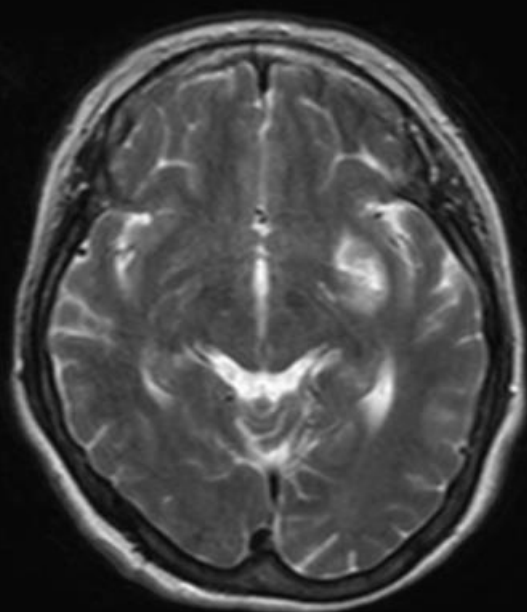
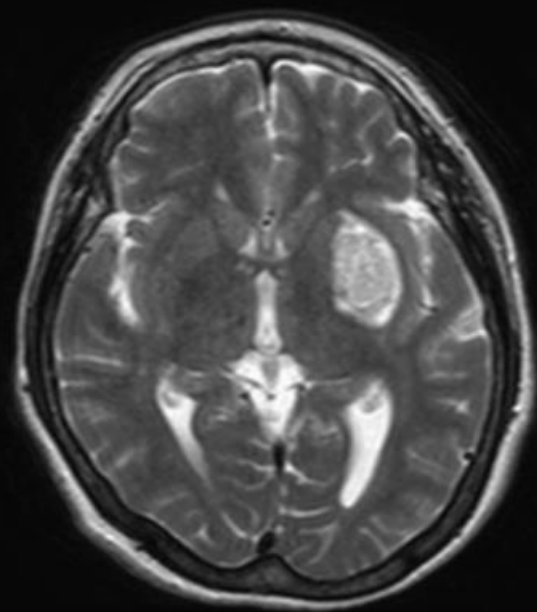


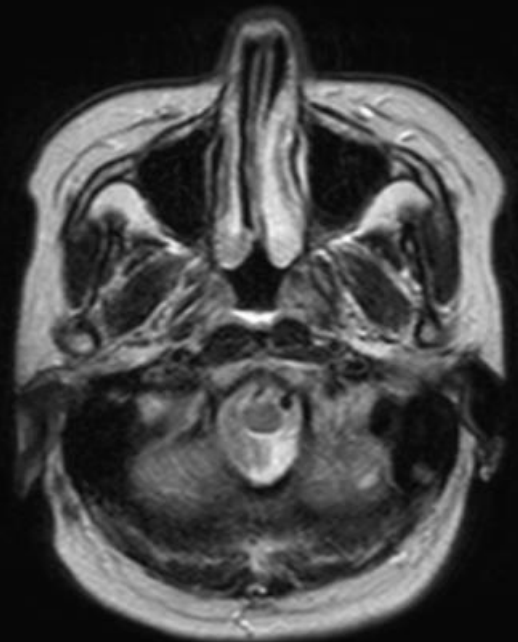
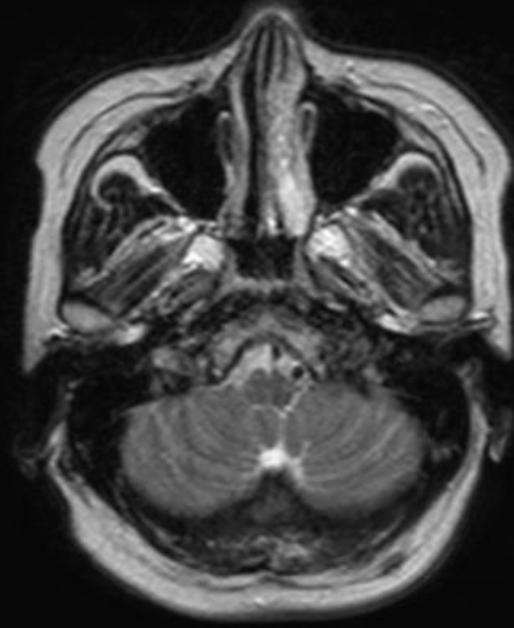
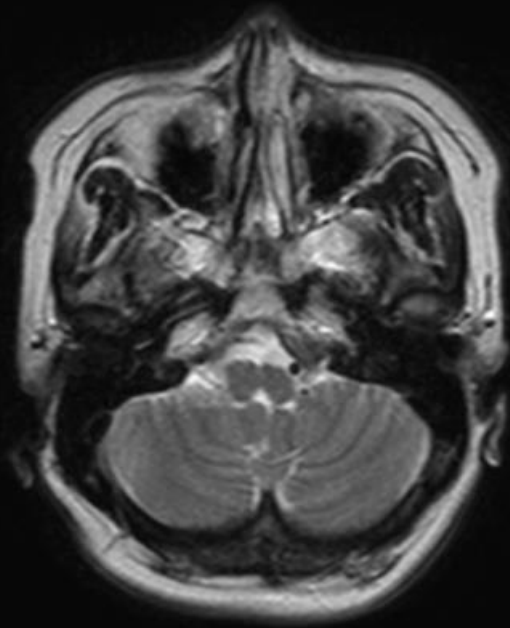
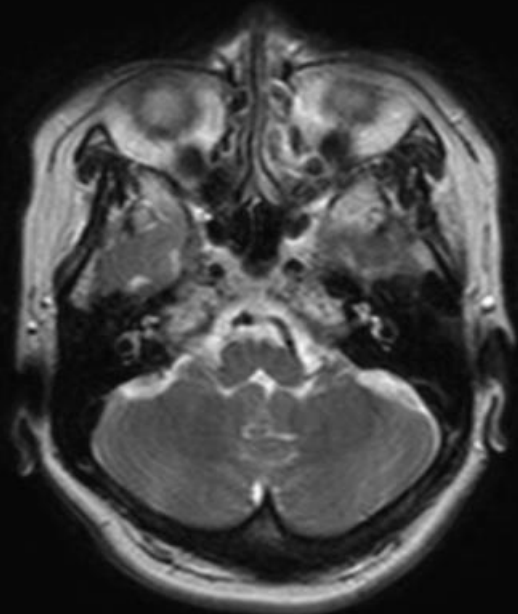




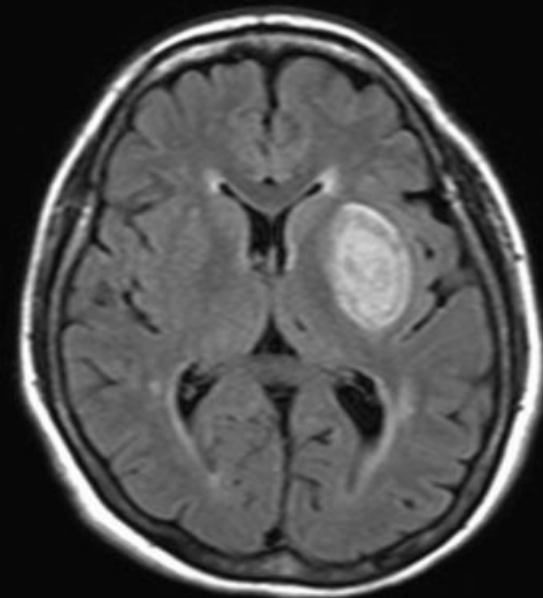
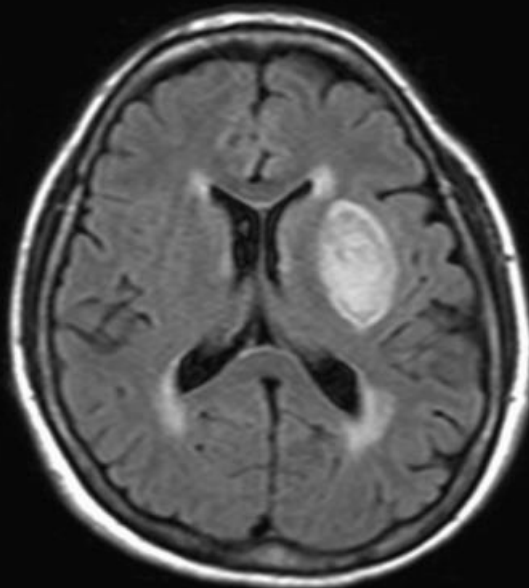
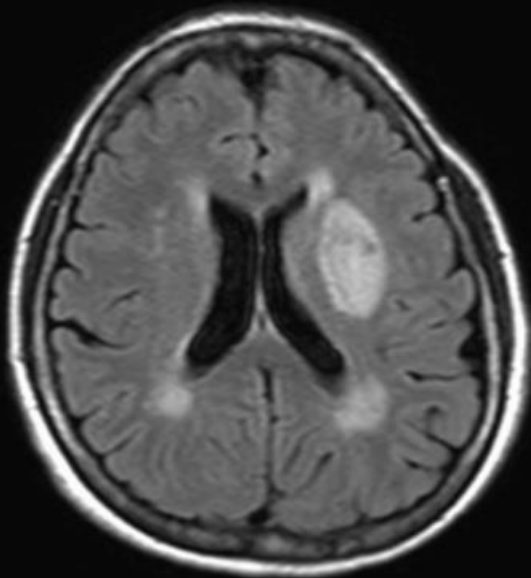
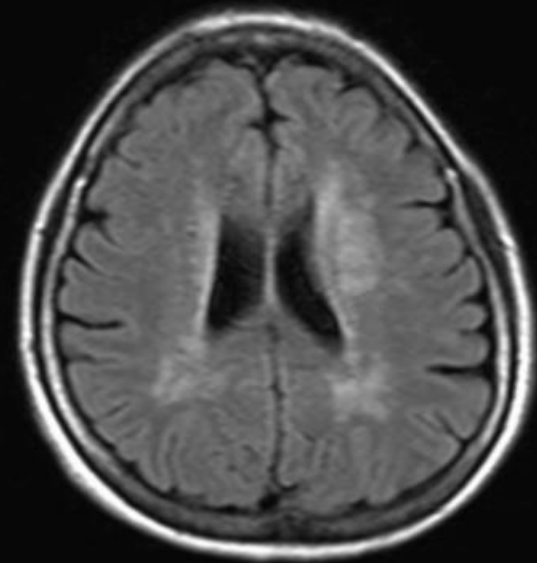
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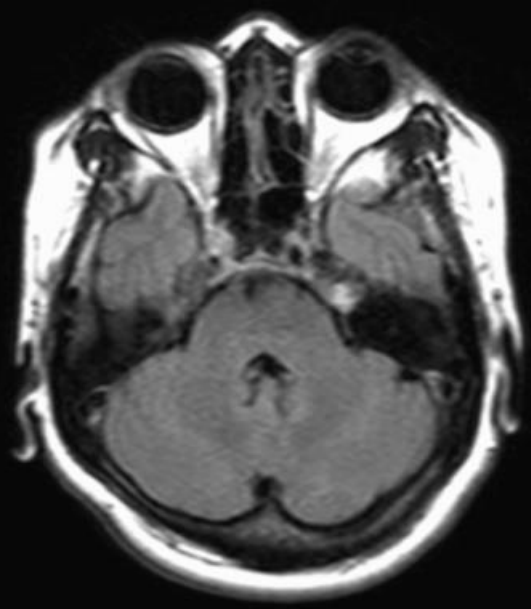
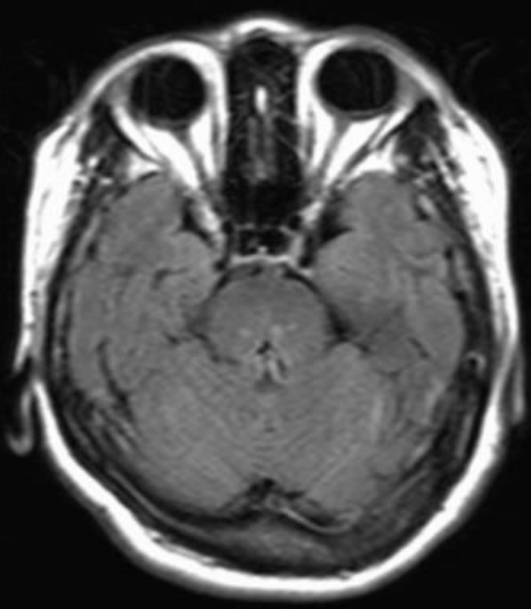
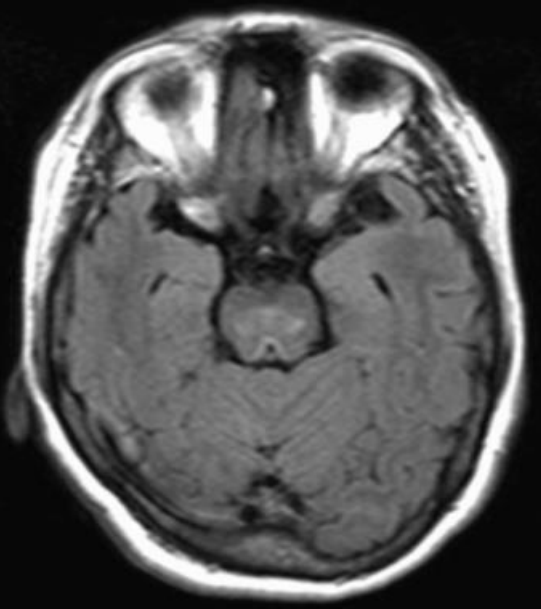
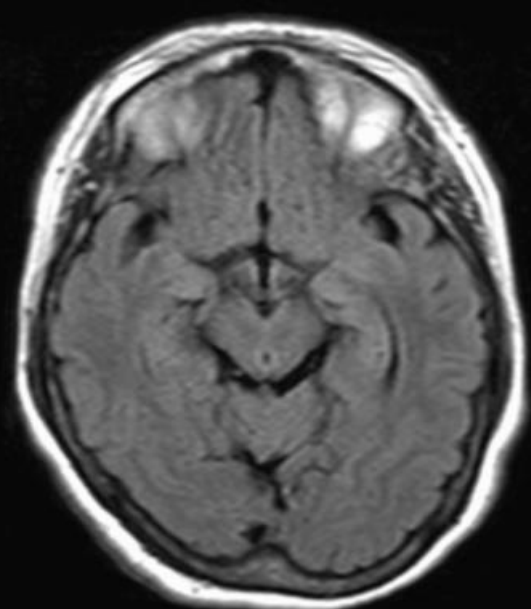
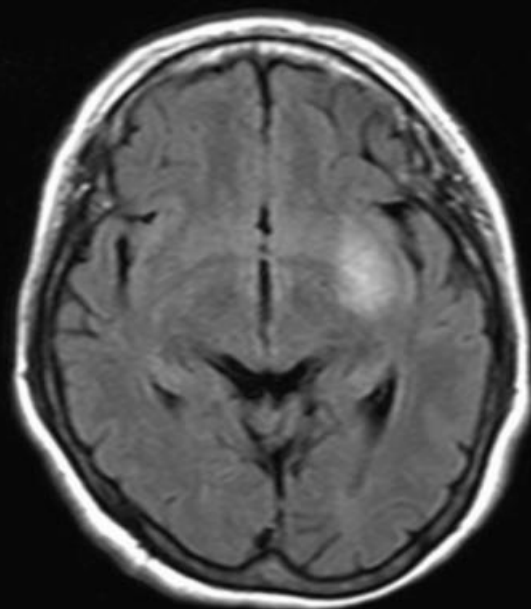
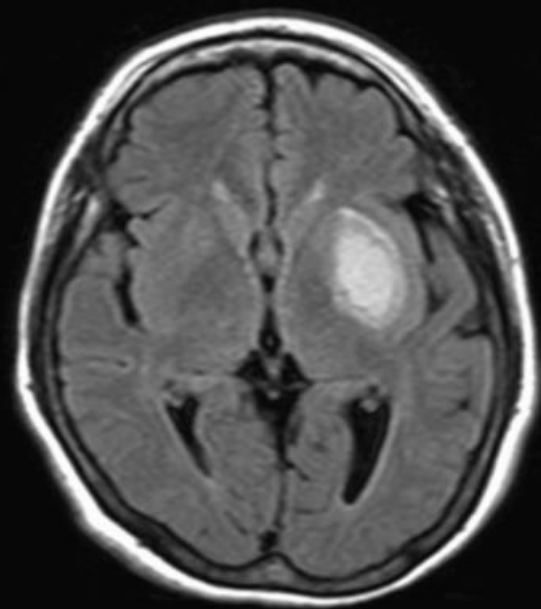


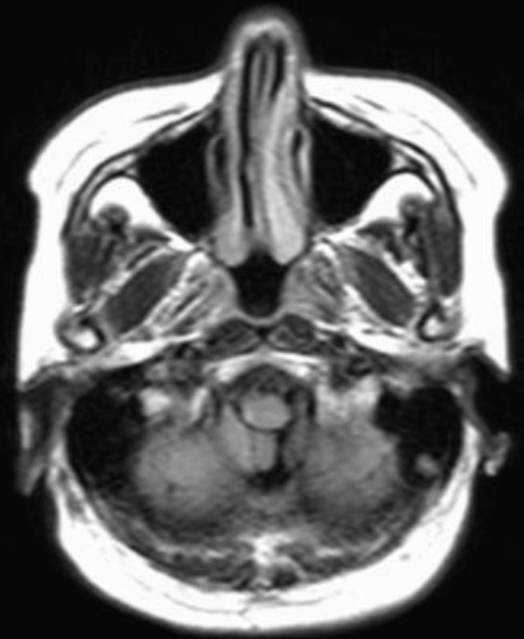
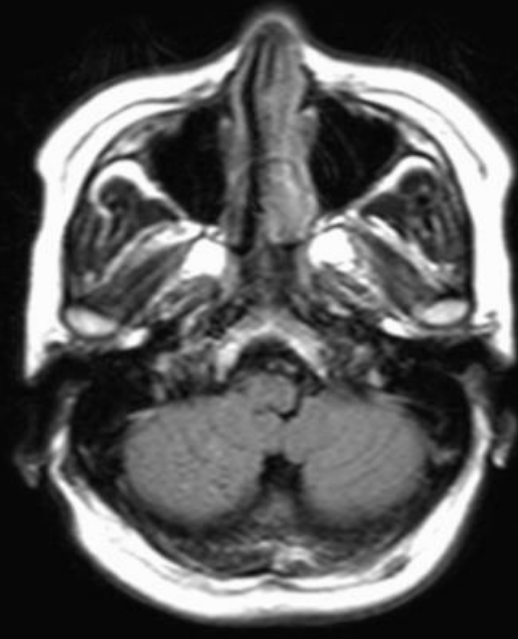
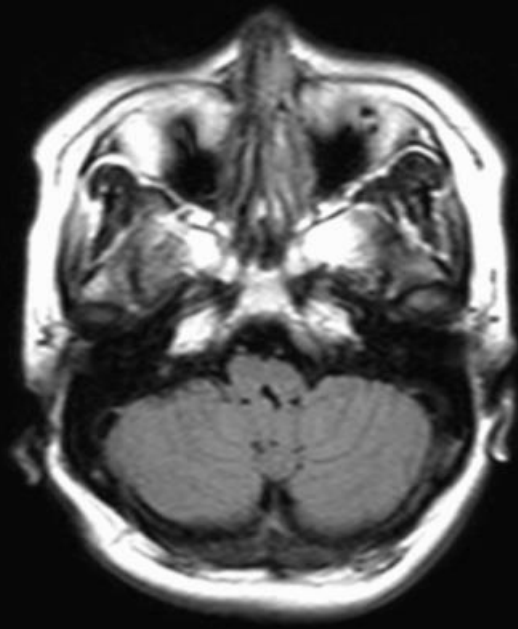
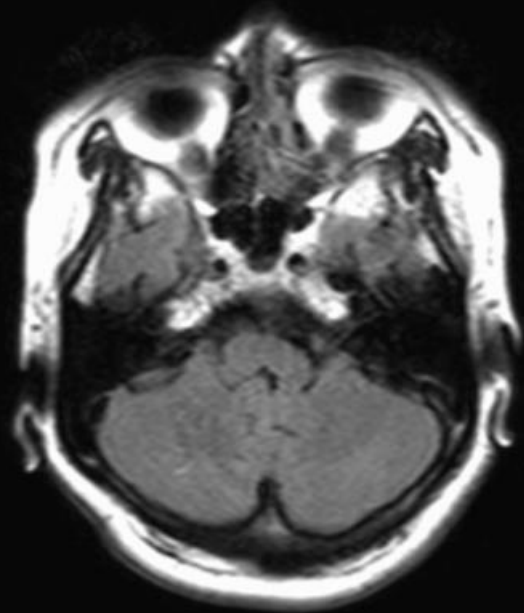




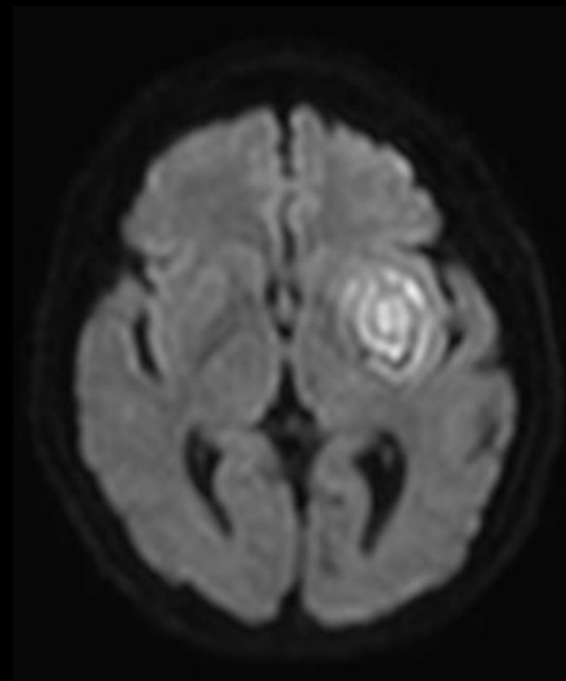
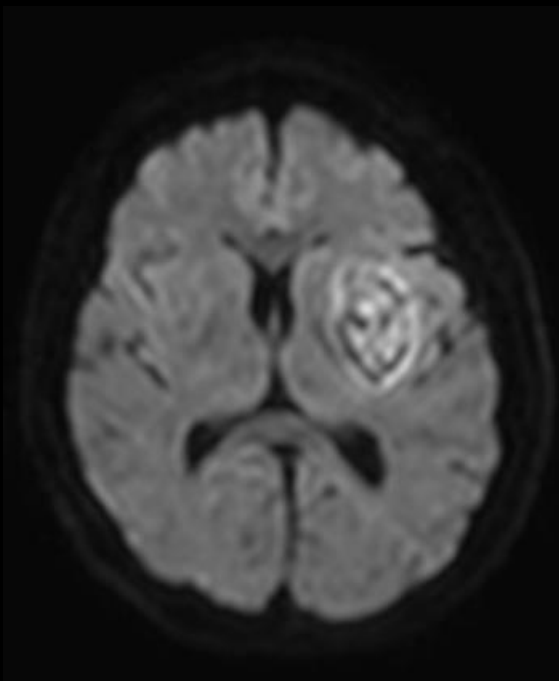
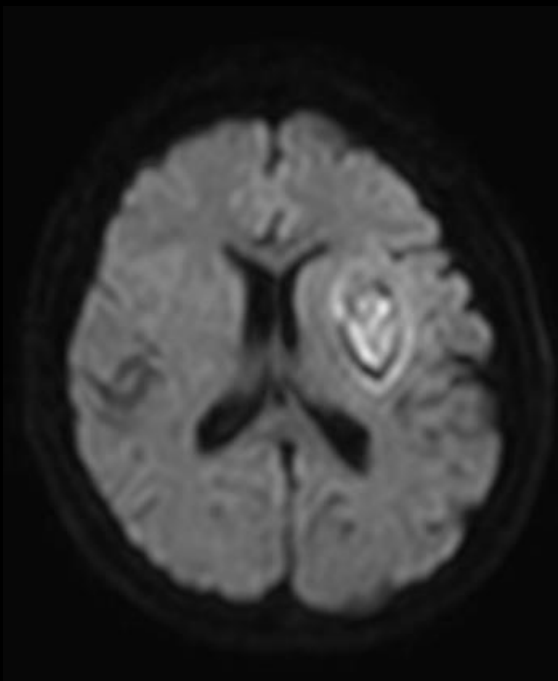
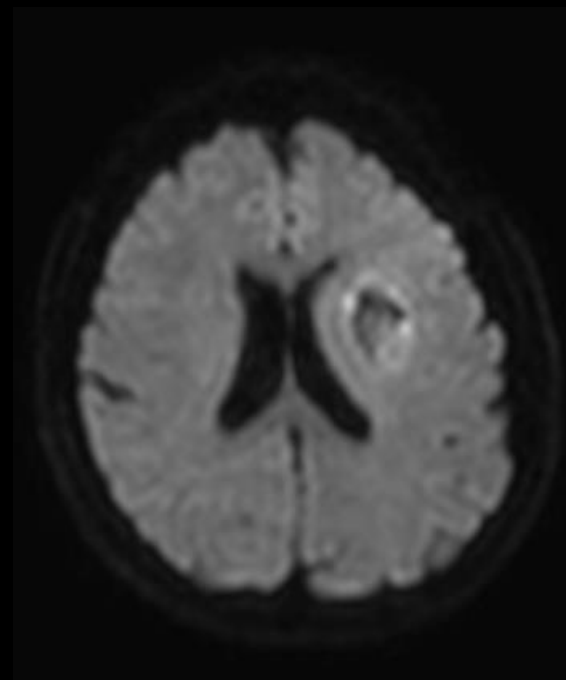
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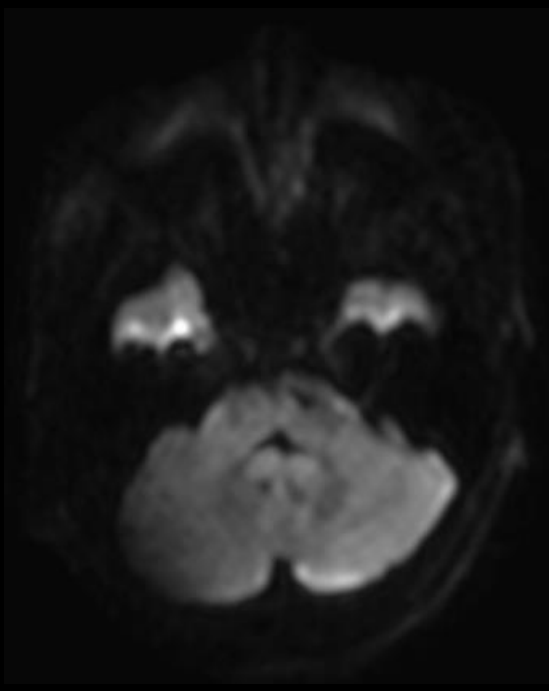
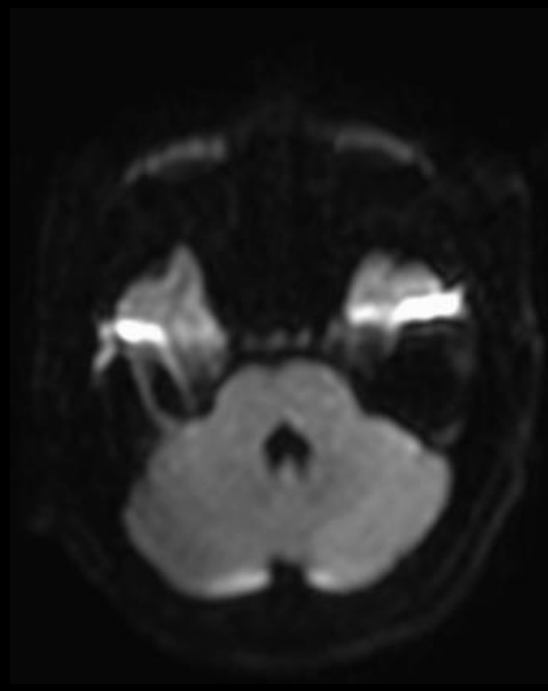
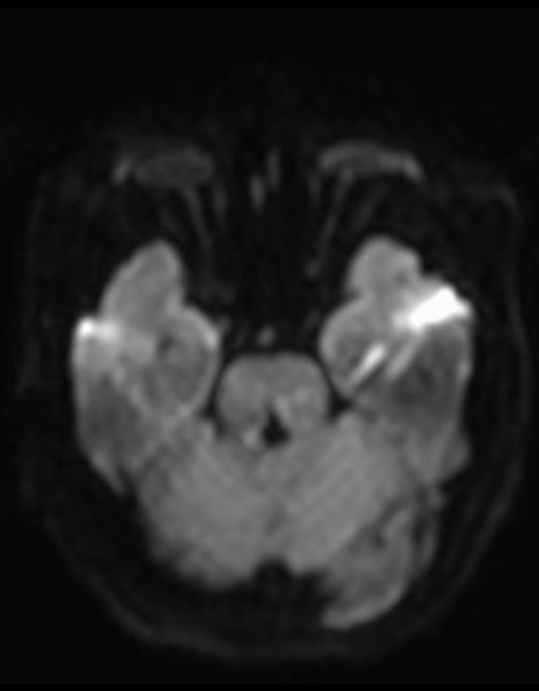
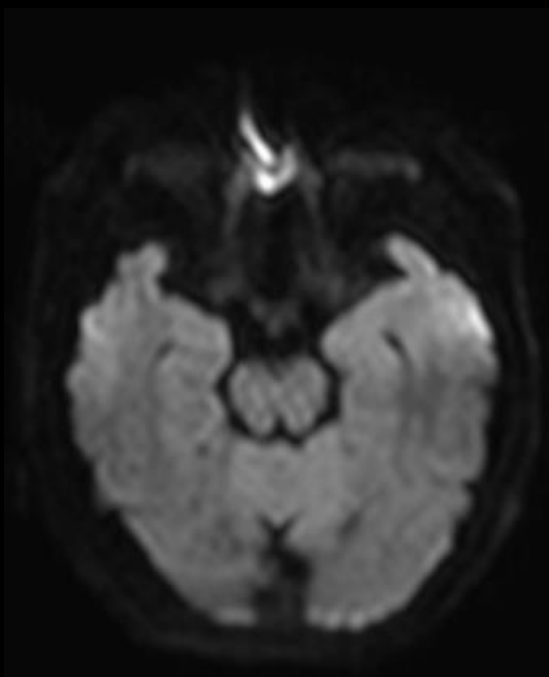
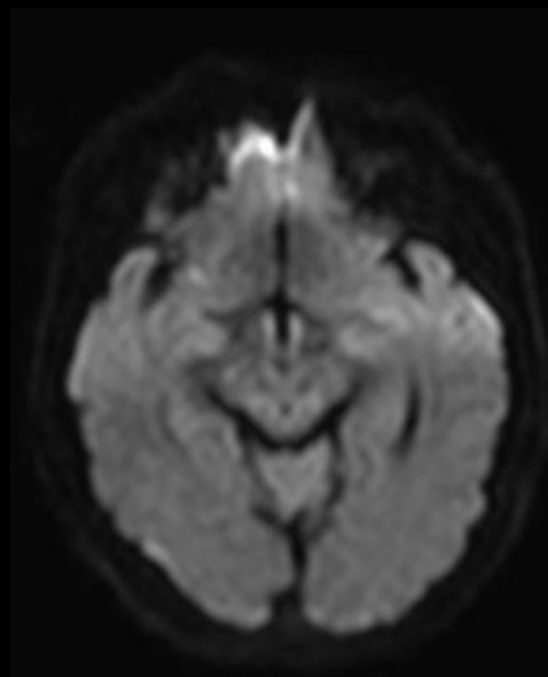
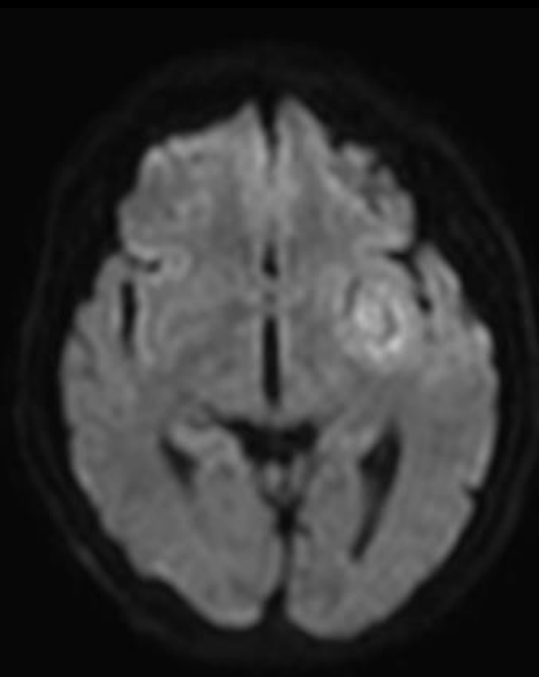


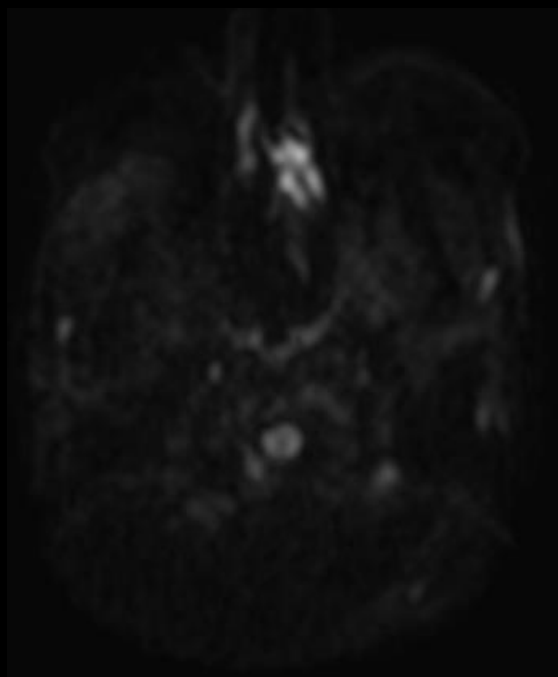
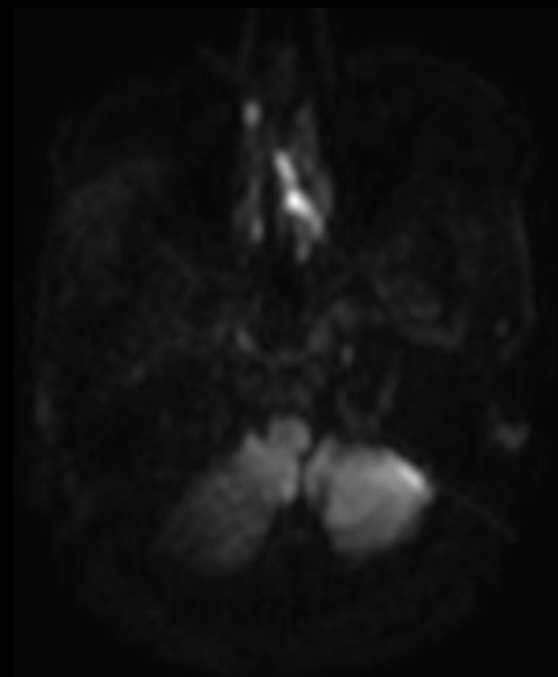
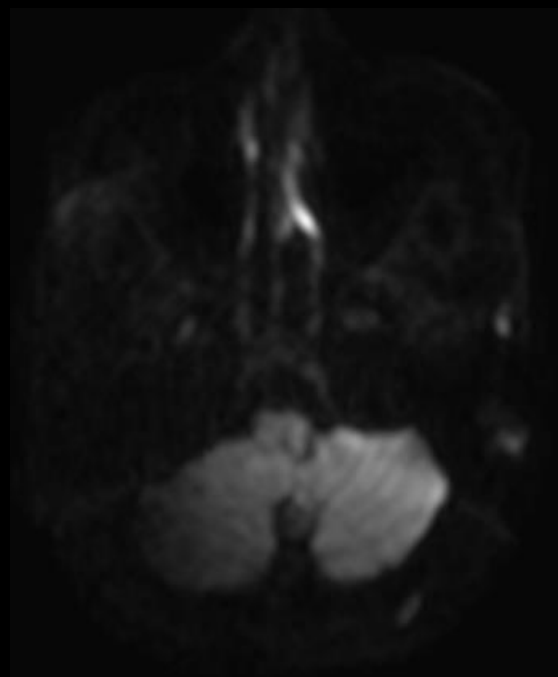
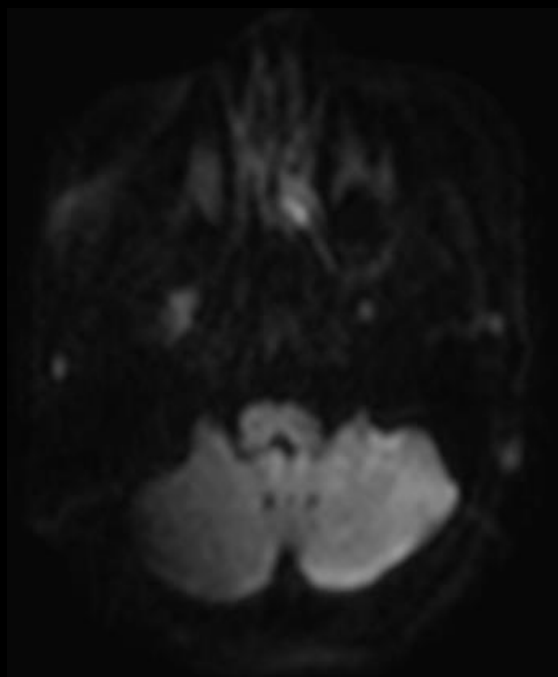




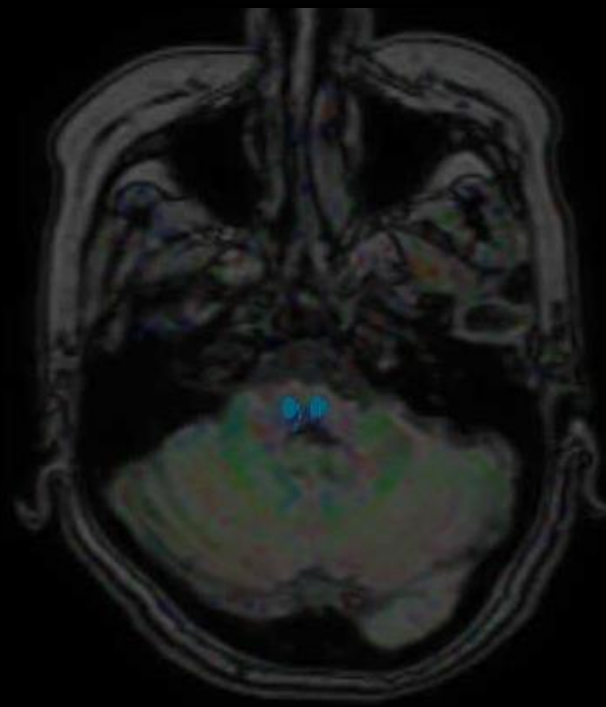
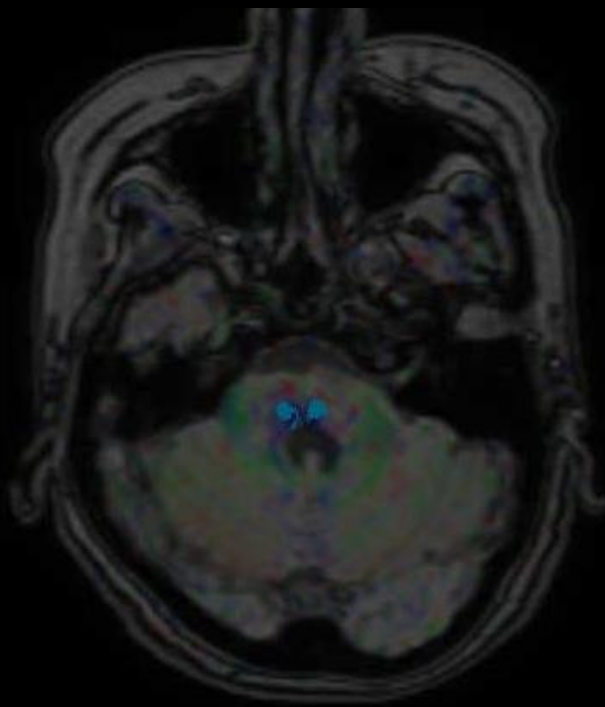
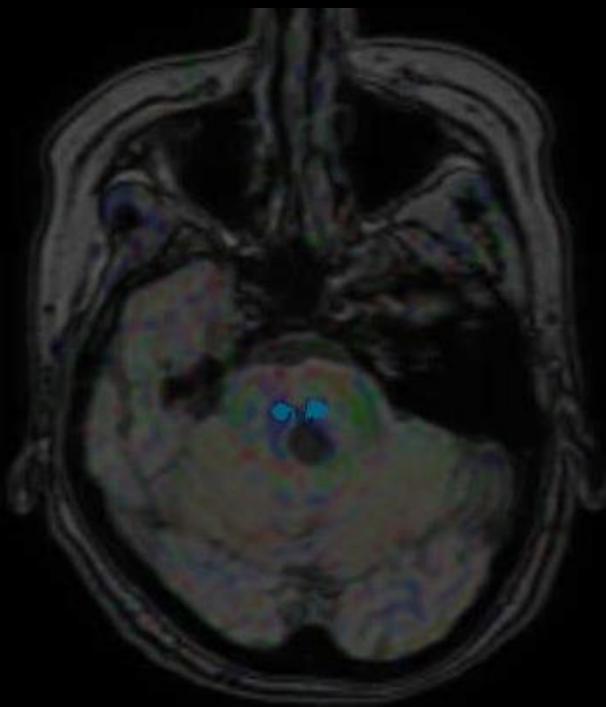
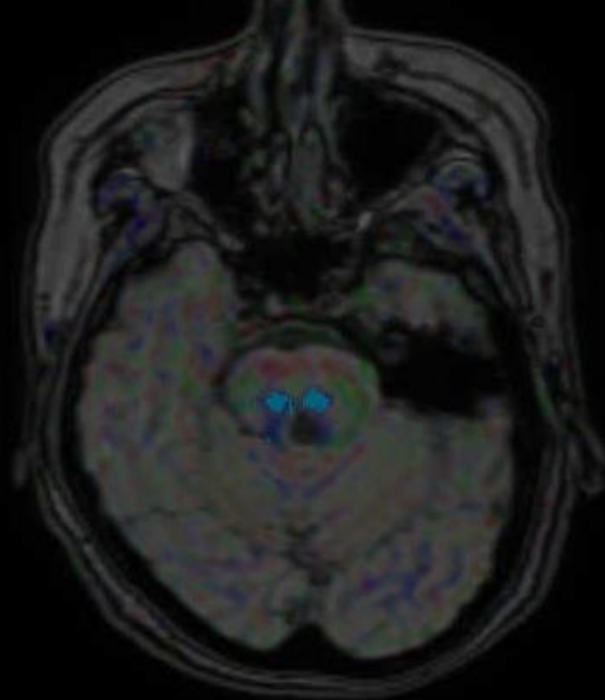
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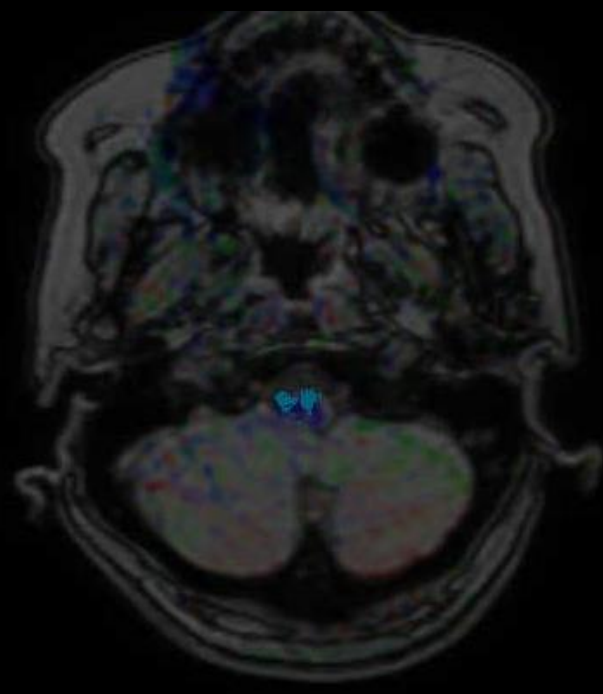
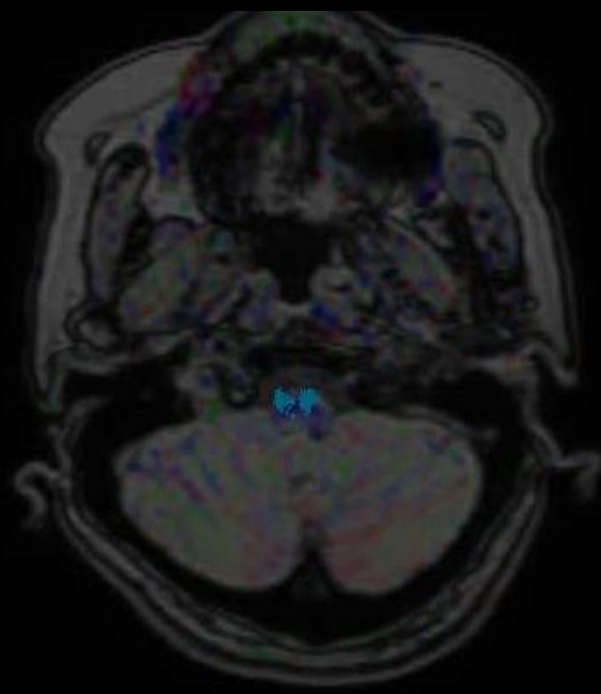
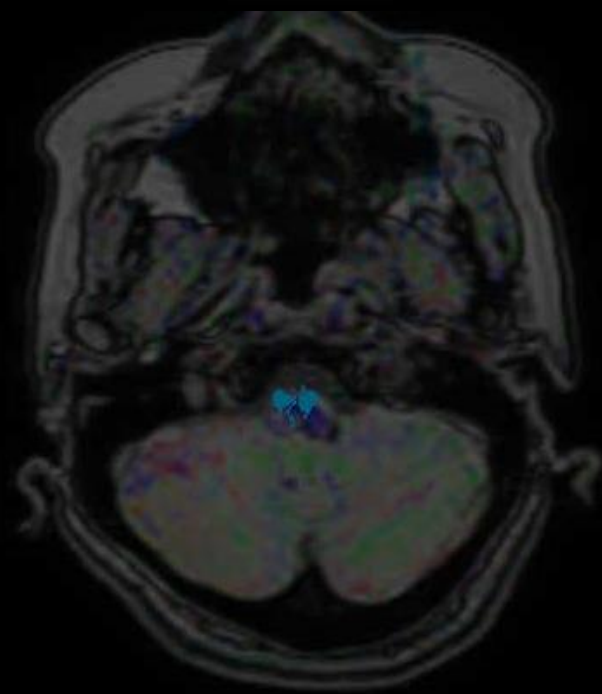
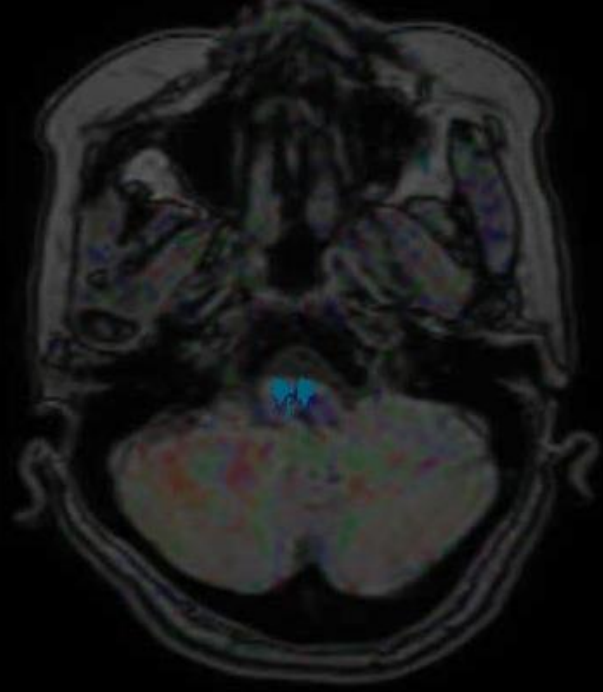
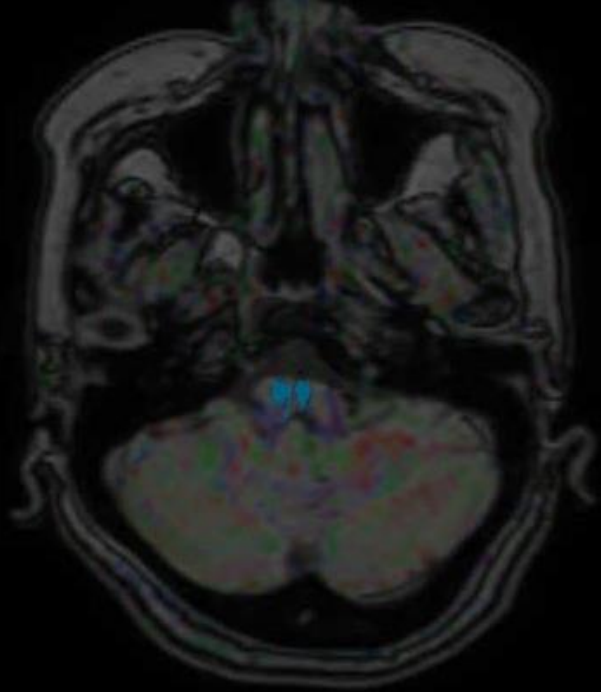
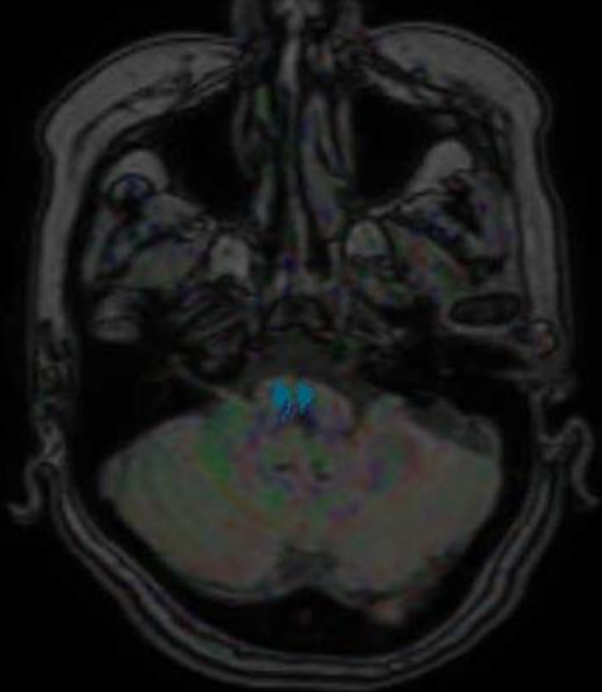


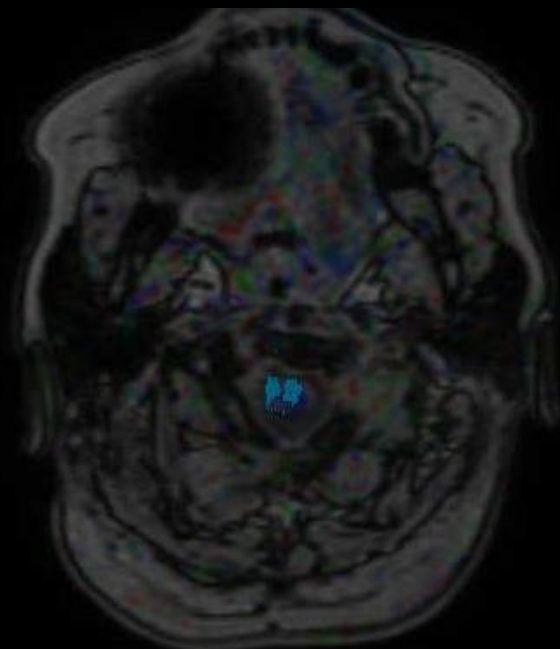
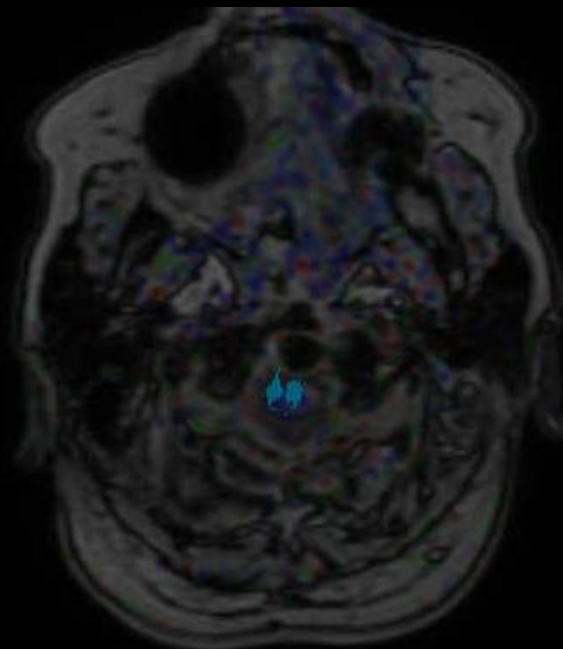
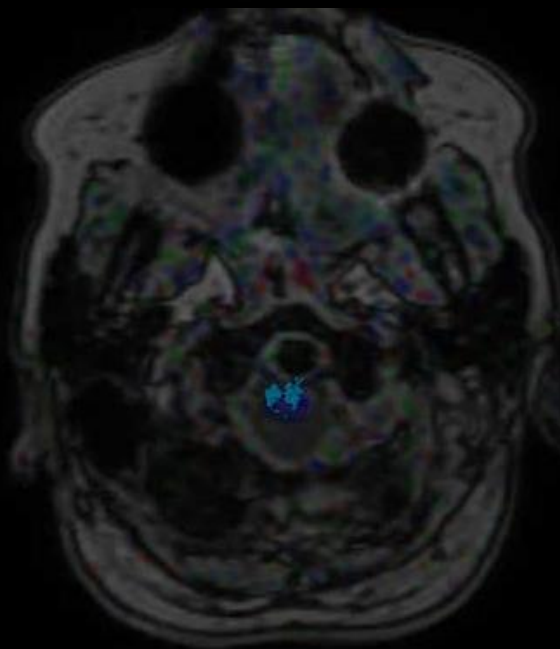
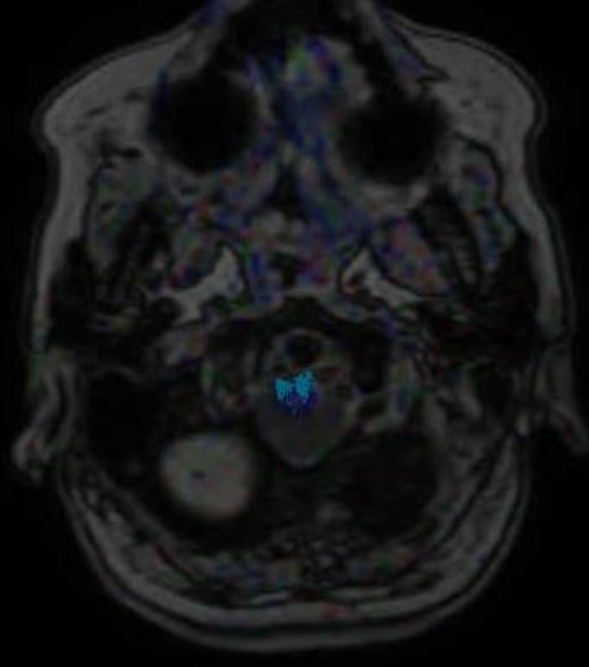
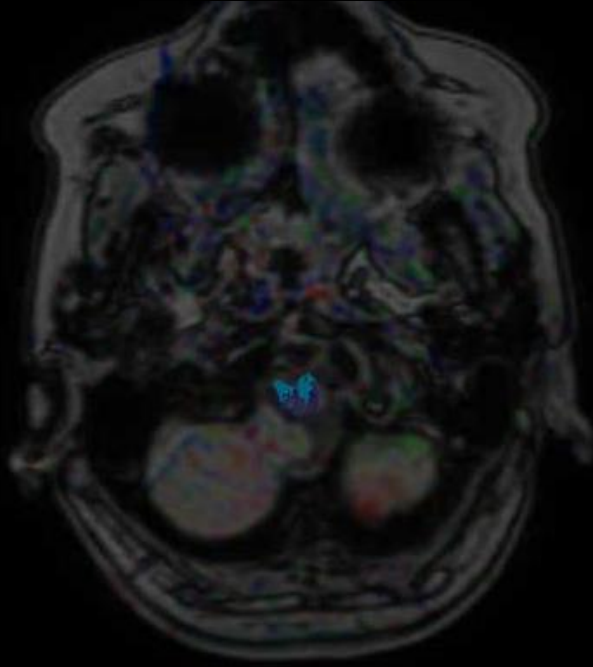
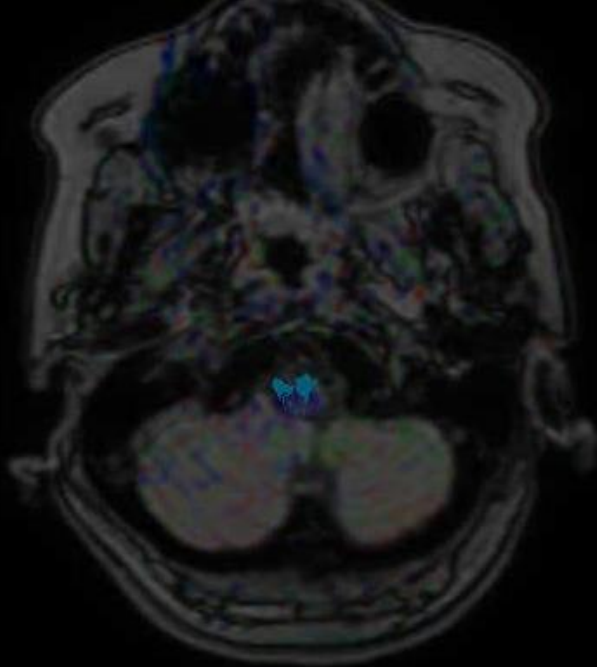




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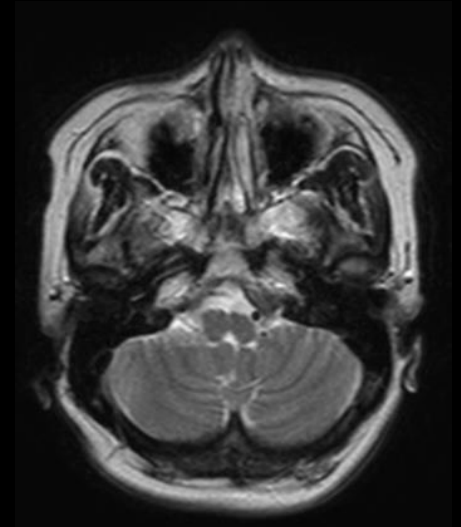
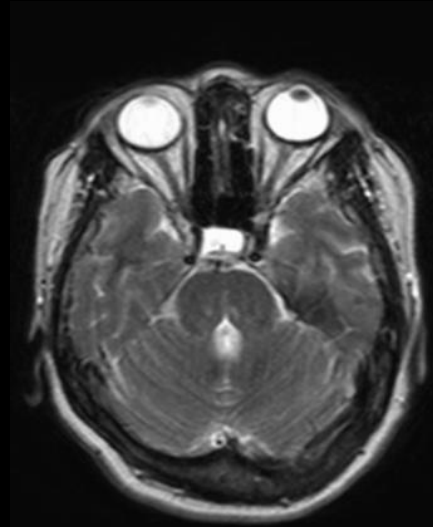
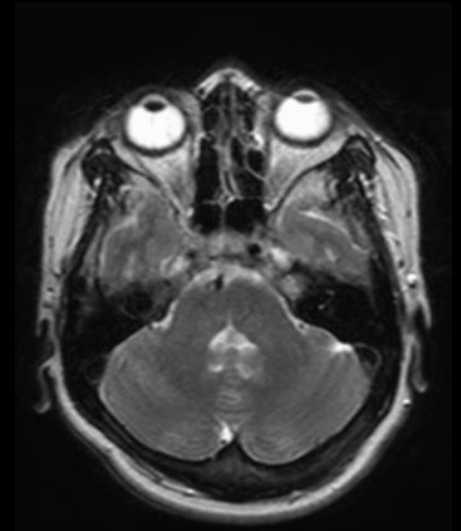
・左被殻出血

・脳幹の形態異常

橋正中背側の陥凹

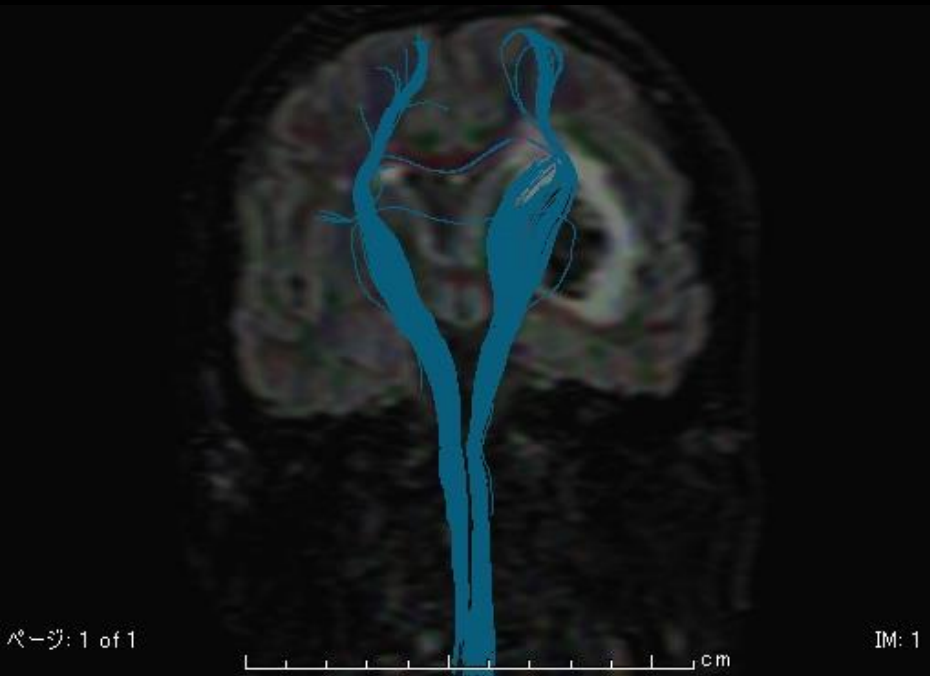
顔面神経丘の欠損

蝶形の延髄

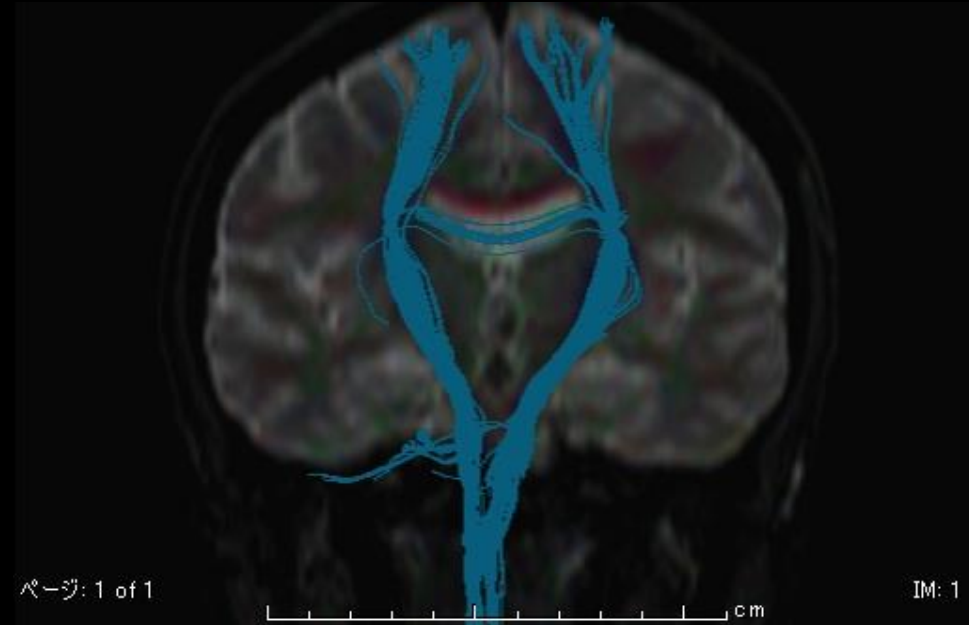


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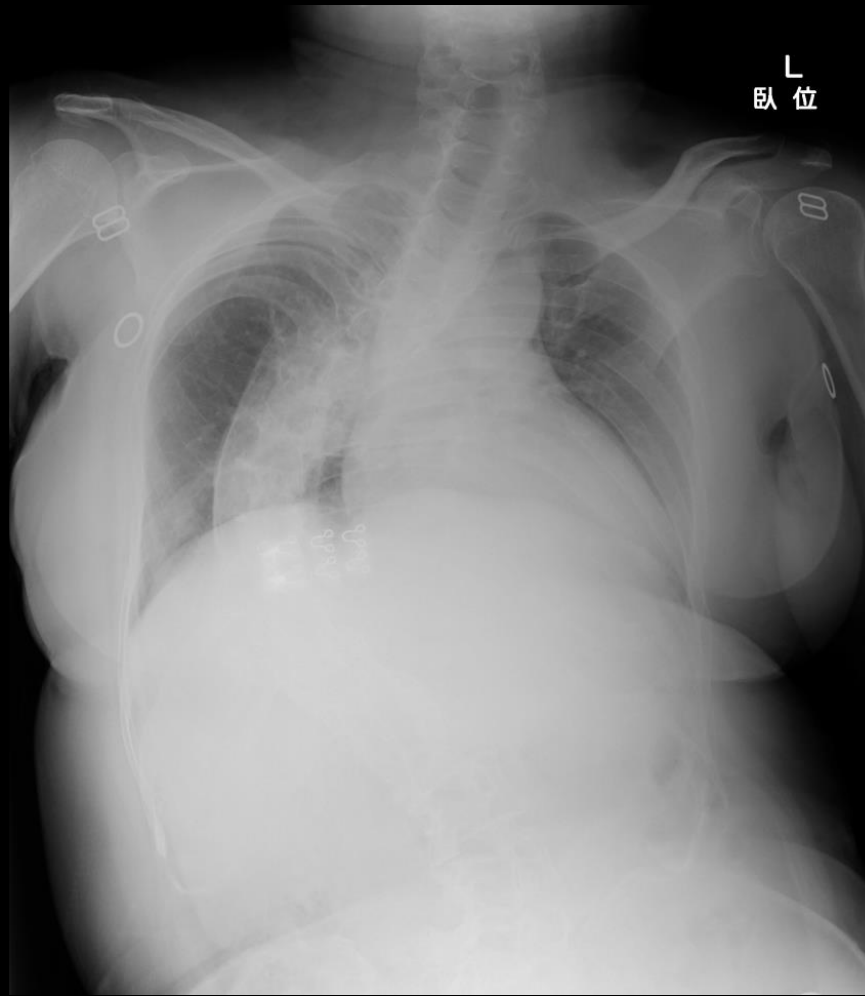
- ・錐体交叉の欠如



本症例



正常



L
臥位

↑ ↑ ↑
肺野

診断

HGPPS:

Horizontal Gaze Palsy with

Progressive Scoliosis

+ left puteminal hemorrhage

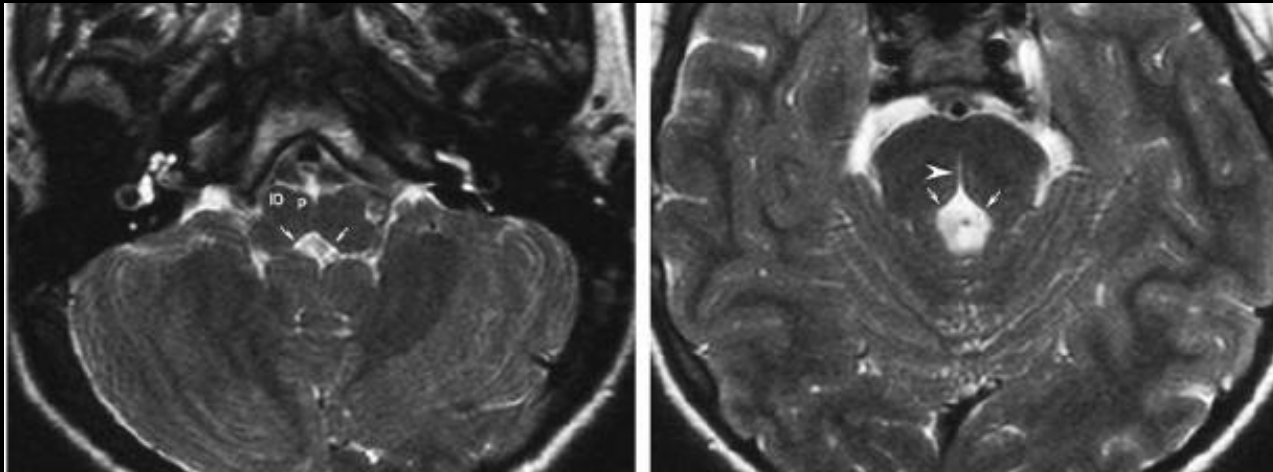
HGPPS

- ・ROBO3の変異による脳幹の形成異常と神経交叉の不良
- ・先天性の水平注視麻痺と進行性の側弯を特徴とする
- ・皮質脊髄路、脊髄後索の経路が交叉しない

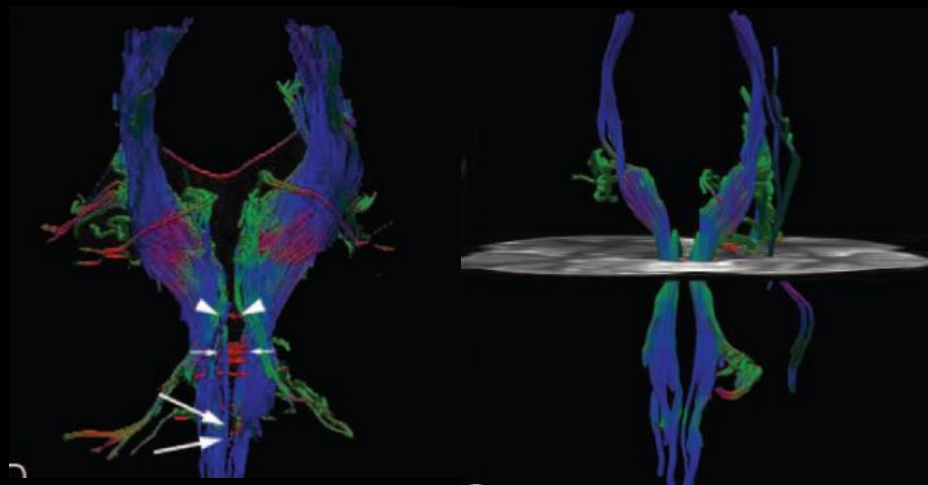
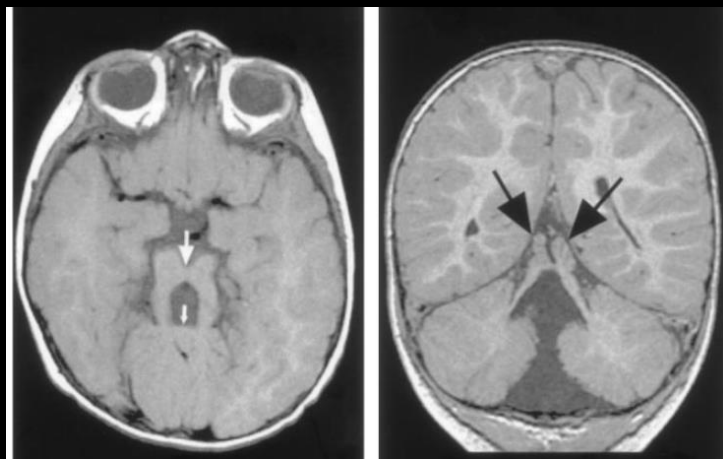
(脊髄視床路は正常に交叉)

画像所見

- ・顔面神経丘の低形成
- ・橋正中背側の陥凹（split pons sign）
- ・テント形の第四脳室
- ・薄束核、楔状束核の後方隆起の欠損
- ・錐体に比べ下オリーブ核が突出して見える



- Joubert症候群も錐体交叉の欠損が見られる



normal

Joubert syndrome