

Hypertrophic olivary degeneration

Published on 22.02.2018

DOI: 10.1594/EURORAD/CASE.14830

ISSN: 1563-4086

Section: Neuroradiology

Area of Interest: Neuroradiology brain

Procedure: Education

Imaging Technique: CT

Imaging Technique: MR

Special Focus: Pathology Case Type: Clinical Cases

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Patient: 61 years, male

Clinical History:

One month after having run out of his anti-hypertensive medication, a 61-year-old male patient was diagnosed in the emergency room as having had a stroke. Nine months after the stroke, he developed a synchronous tremor in his palate.

Imaging Findings:

The initial emergency CT examination (day 0) indicated a hyperdense lesion corresponding to an acute haemorrhage in the dorsal pons (Fig 1). An MRI revealed an interval evolution slightly to the right of the dorsal pons. No underlying mass lesion or vascular malformation was noted. Expansile T1 signal loss in the right medullary olive appears to have developed from hypertrophic olivary degeneration. Axial, sagittal, and coronal T1 were recorded on days 0, 30, 120, and 270 (Figs 2, 3, 4), and Coronal FLAIR was recorded on day 270 (Fig 5).

Discussion:

Hypertrophic olivary degeneration (HOD), or inferior olivary hypertrophy, is an uncommon form of transsynaptic degeneration, which occurs when the dentate-rubral olivary pathway, i.e., the Guillain-Mollaret Triangle or GMT, is disrupted [1, 2]. Its pathology includes fibrillary gliosis, olive demyelination, and hypertrophy of those inferior olives.

This lesion's most common aetiologies are a haemorrhagic stroke (ischaemic is less common), brainstem tumours or trauma, metronidazole intoxication, multiple sclerosis, and surgical (or gamma knife) cavernoma brainstem treatment.

Anatomically, the GMAT triangle is made up of three connected nucleus structures: the cerebellum's contralateral dentate nucleus, the inferior olivary nucleus (ION), and the ipsilateral red nucleus (Fig 5). The fibers stemming from the cerebellum's nucleus proceed upward through the dentatorubral tract in the brachium conjunctivum and then across the brain's midline to the contralateral red nucleus which is situated midbrain. Fibers issuing from the red nucleus pass downward to the ipsilateral ION by way of the central tegmental tract. Lesions along this pathway can cause HOD. To complete the triangle, olivodentate fibers proceeding from the cerebellar peduncle pass over to the

ipsilateral ION. This pathway does not play a role in HOD because of the presence of a relay located between the cerebellar cortex and the dentate nucleus along the olivocerebellar tract.

Transmissions through the GMT are responsible for inhibitory control. Hence, disruptions to these GMT transmissions could bring about a loss of inhibitory control. This is the most likely reason that results in tremors such as these: 1) Palatal tremor (PT), which is marked rhythmical contractions of the elevator veli palatine resulting in typically bilateral and symmetric, involuntary soft palate and pharynx movements, which accompany half of all HOD cases [1, 3]. 2) Oculopalatal tremor (OPT), in which PT and pendular nystagmus are both present at the same time. 3) Dentato-rubral tremor (DTR), also called 'holmes tremor', which is a distinctive, low-frequency tremor syndrome which includes resting, intention, and postural tremors.

When viewed in an MRI, HOD has the following characteristics: 1) T2/FLAIR signal intensity and enlarged inferior olive, 2) a persisting hyper-signal of about one month following the ictus, 3) appearance of hypertrophy about 3 – 5 months post-ictus, which usually resolves at 3–4 years post-ictus [1]. In the present case, the patient was diagnosed with HOD 270 days after suffering the dorsal pons haemorrhage.

In general, treatment includes gabapentin, memantine, baclofen, trihexyphenidyl, and Botulinum toxin [1].

Differential Diagnosis List: Hypertrophic olivary degeneration, Demyelinating disorders, Astrocytoma, Infections

Final Diagnosis: Hypertrophic olivary degeneration

References:

- Tilikete C, Desestret V. (2017) Hypertrophic Olivary Degeneration and Palatal or Oculopalatal Tremor. *Front Neurol* Jun 29;8:302 (PMID: [28706504](#))
- Sabat S, Mannering N, Agarwal A. (2016) Hypertrophic olivary degeneration: A clinico-radiologic study. *J Neurol Sci* Nov 15;370:180-186 (PMID: [27772756](#))
- Konno T, Broderick DF, Tacik P, Caviness JN, Wszolek ZK. (2016) Hypertrophic olivary degeneration: A clinico-radiologic study. *Parkinsonism Relat Disord* Jul;28:36-40. (PMID: [27132500](#))

Figure 1

a



Description: Emergency CT shows hyperdense lesion corresponding to an acute haemorrhage at the dorsal pons. **Origin:** Augusta University

b



Description: Emergency CT shows hyperdense lesion corresponding to an acute haemorrhage at the dorsal pons. **Origin:** Augusta University

c



Description: Emergency CT shows hyperdense lesion corresponding to an acute haemorrhage at the dorsal pons. **Origin:** Augusta University

d



Description: Emergency CT shows hyperdense lesion corresponding to an acute haemorrhage at the dorsal pons. **Origin:** Augusta University

Figure 2

a



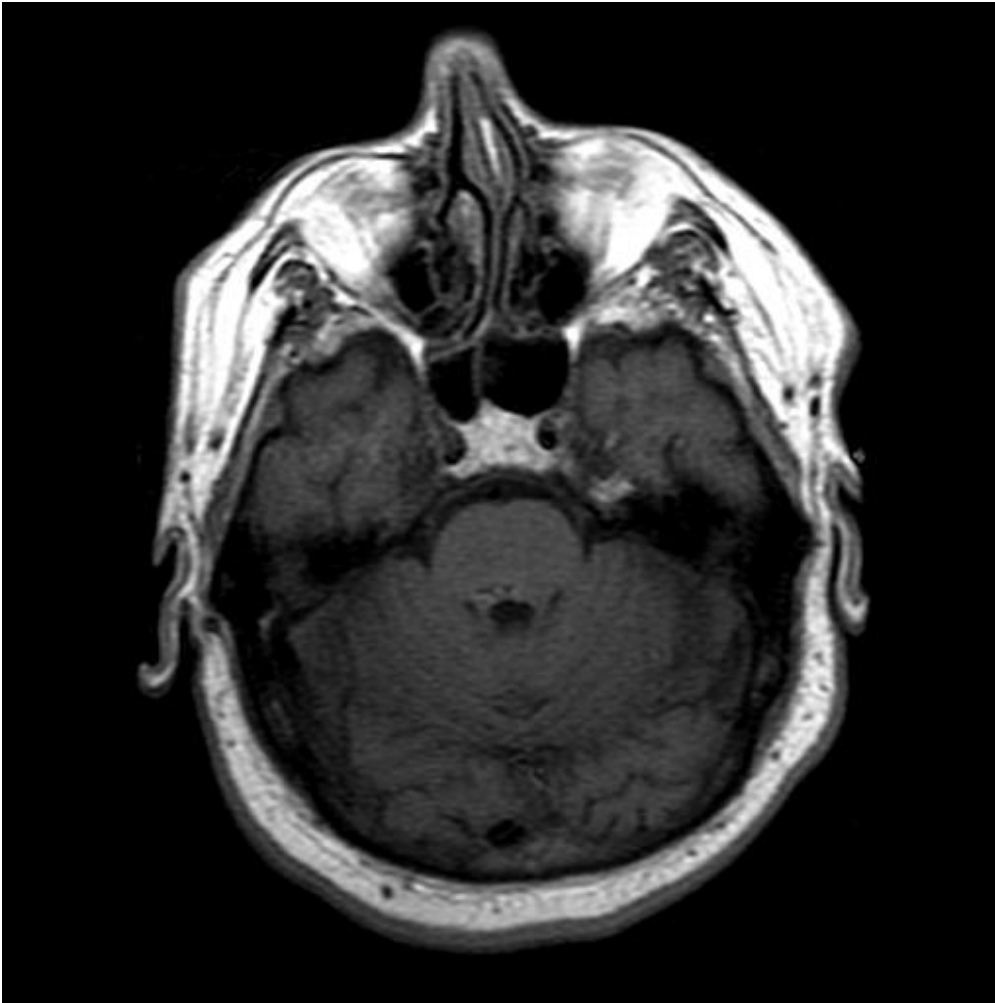
Description: Day 0. Interval evolution of haemorrhage of the dorsal pons, a slightly eccentric to the right. **Origin:** Augusta University

b



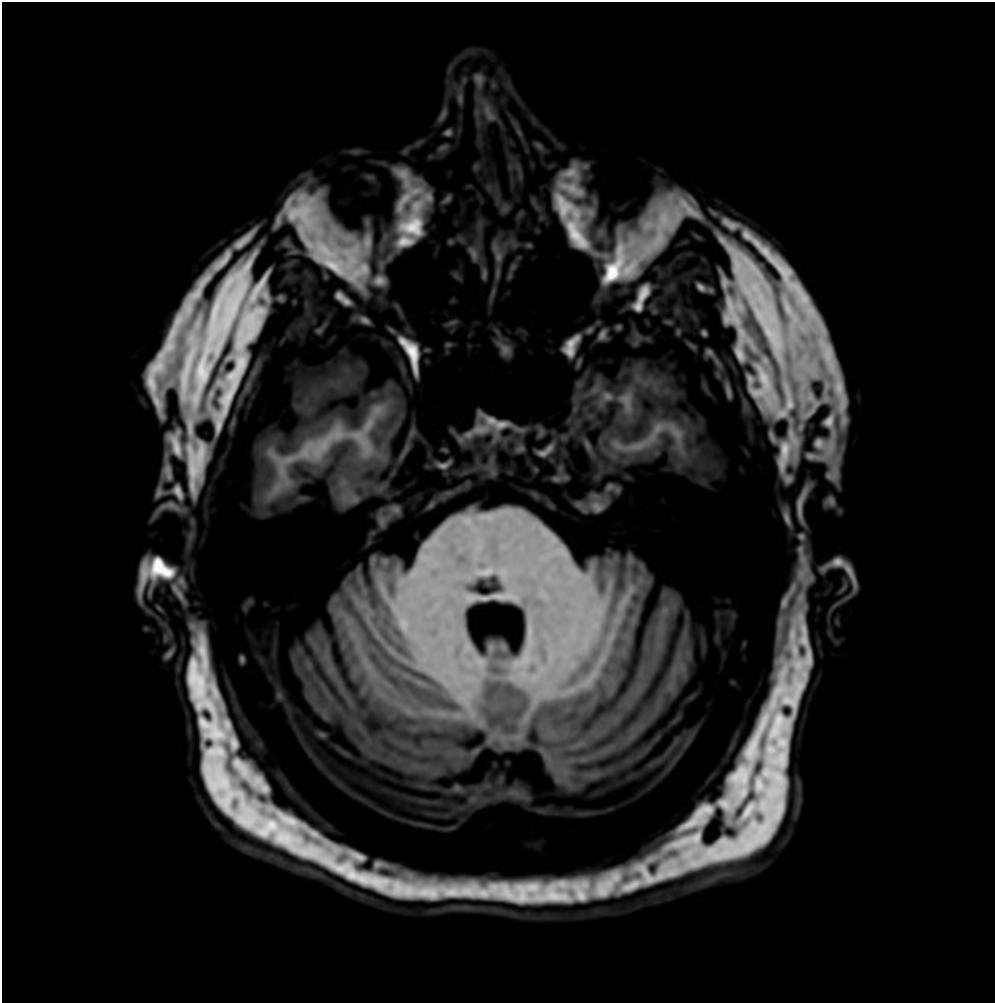
Description: Day 30. Interval evolution of haemorrhage of the dorsal pons, a slightly eccentric to the right. **Origin:** Augusta University

c



Description: Day 120. Interval evolution of haemorrhage of the dorsal pons, a slightly eccentric to the right. **Origin:** Augusta University

d



Description: Day 270. Interval evolution of haemorrhage of the dorsal pons, a slightly eccentric to the right. **Origin:** Augusta University

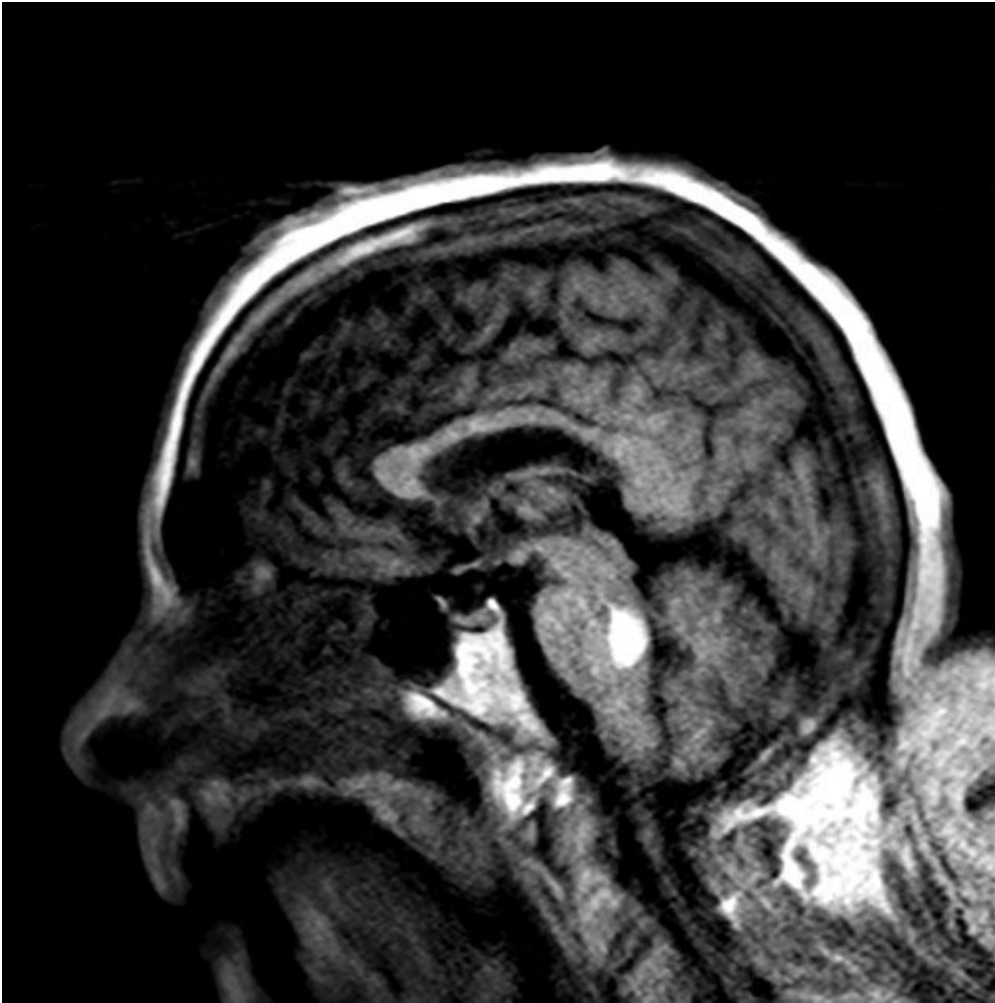
Figure 3

a



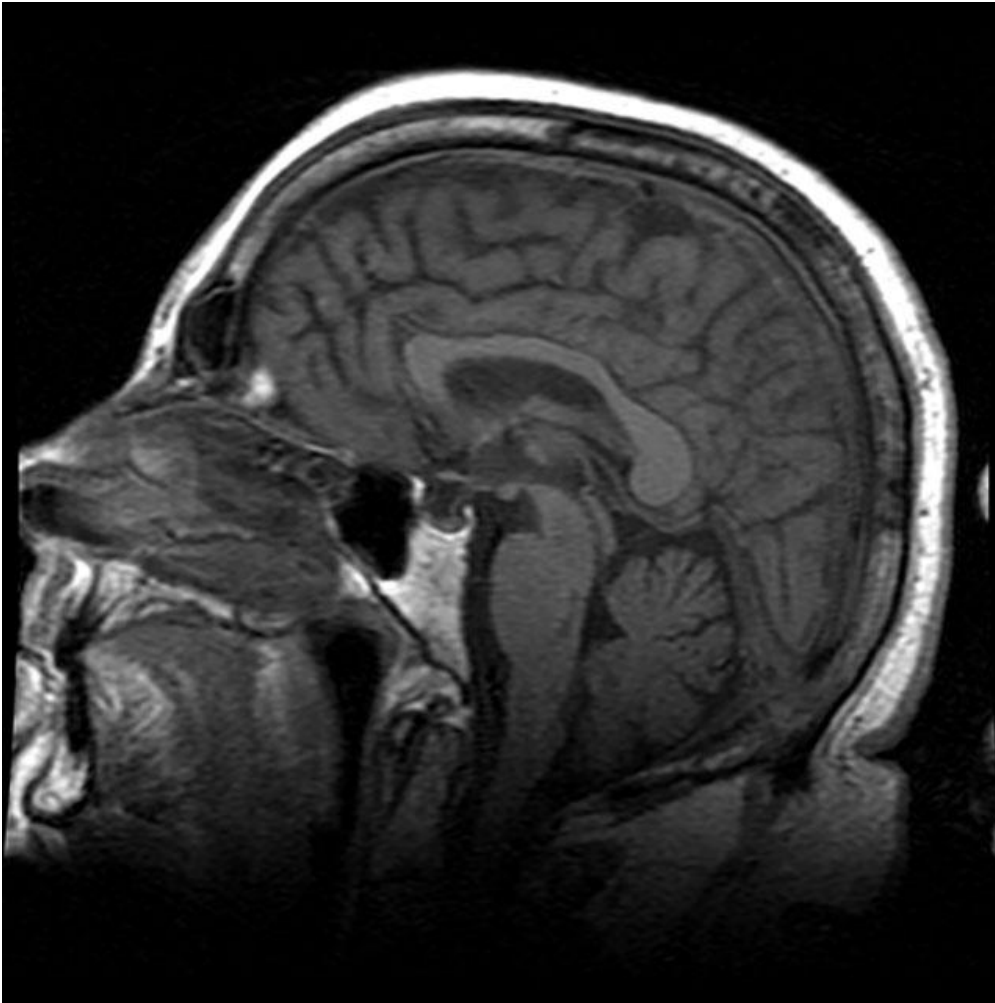
Description: Day 0. Interval evolution of haemorrhage of the dorsal pons. **Origin:** Augusta University

b



Description: Day 30. Interval evolution of haemorrhage of the dorsal pons. **Origin:** Augusta University

c



Description: Day 120. Interval evolution of haemorrhage of the dorsal pons. **Origin:** Augusta University

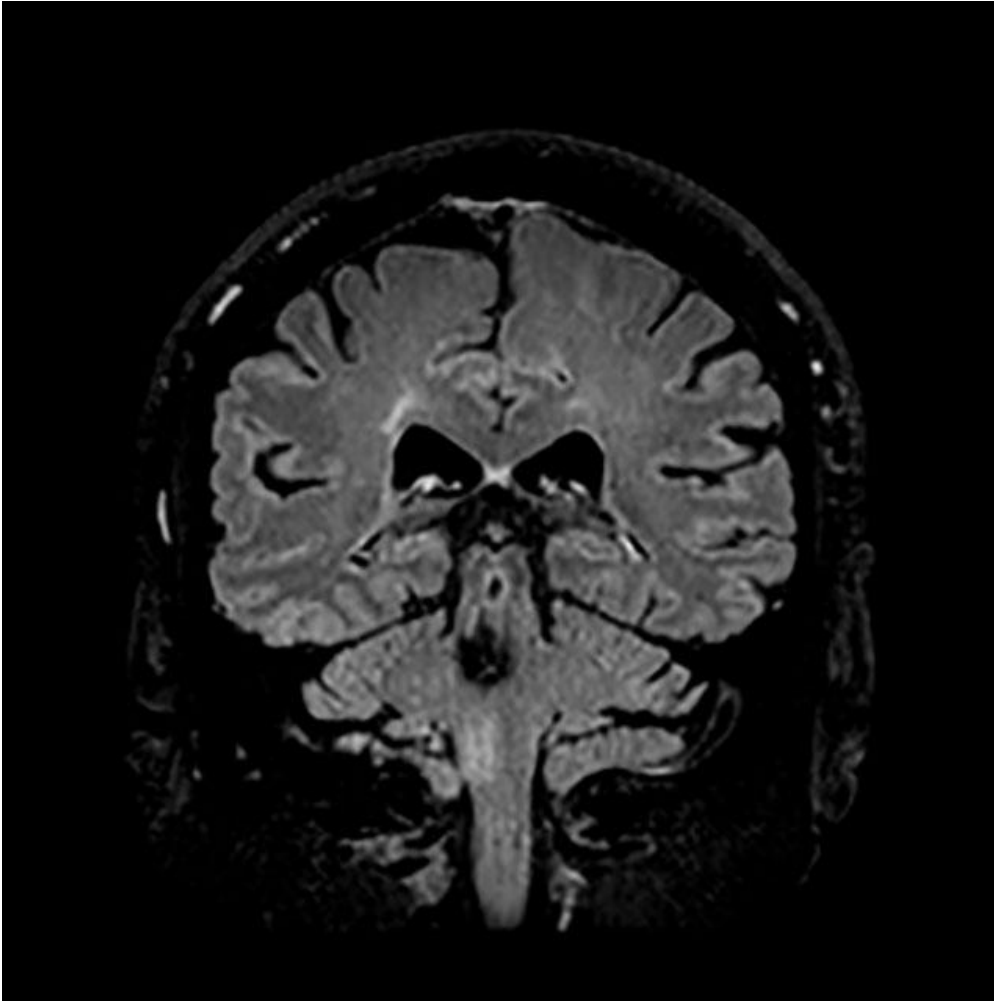
d



Description: Day 270. Interval evolution of haemorrhage of the dorsal pons. **Origin:** Augusta University

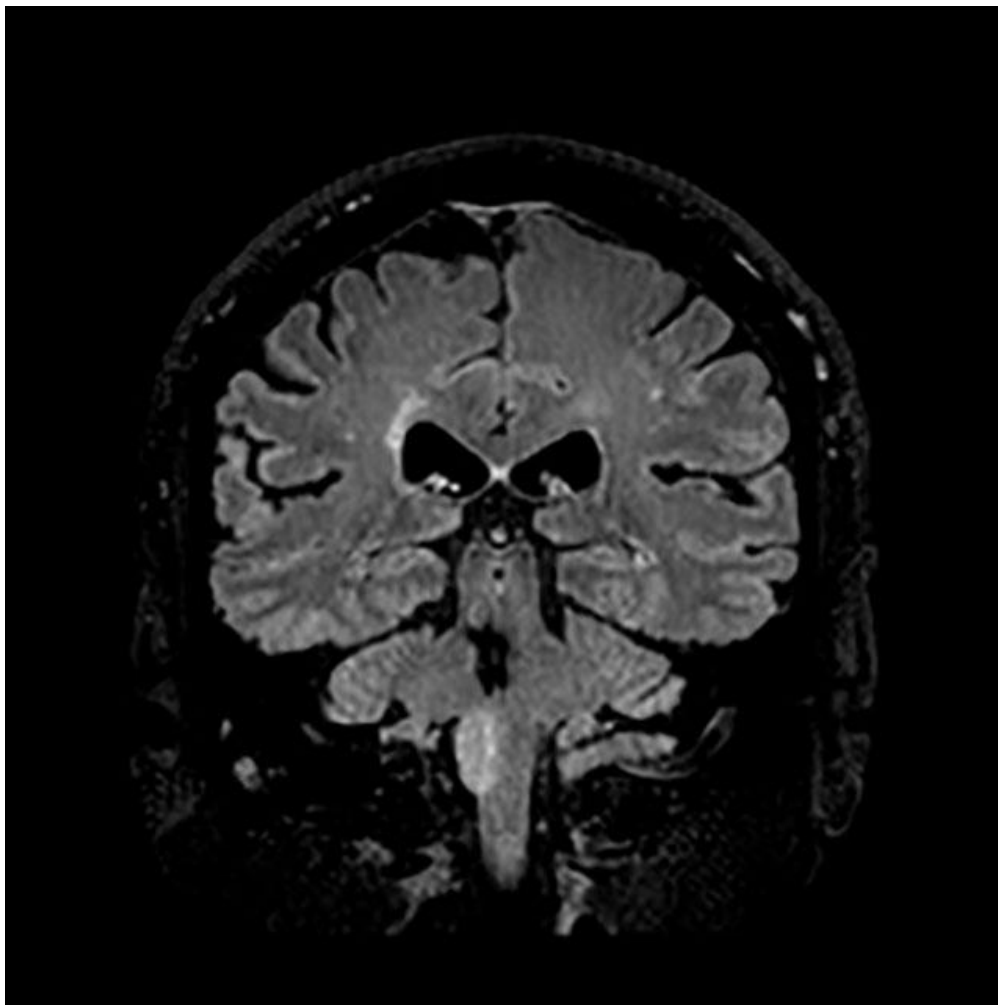
Figure 4

a



Description: Day 270. Interval development of an expansile lesion of the right medullary olive. **Origin:** Augusta University

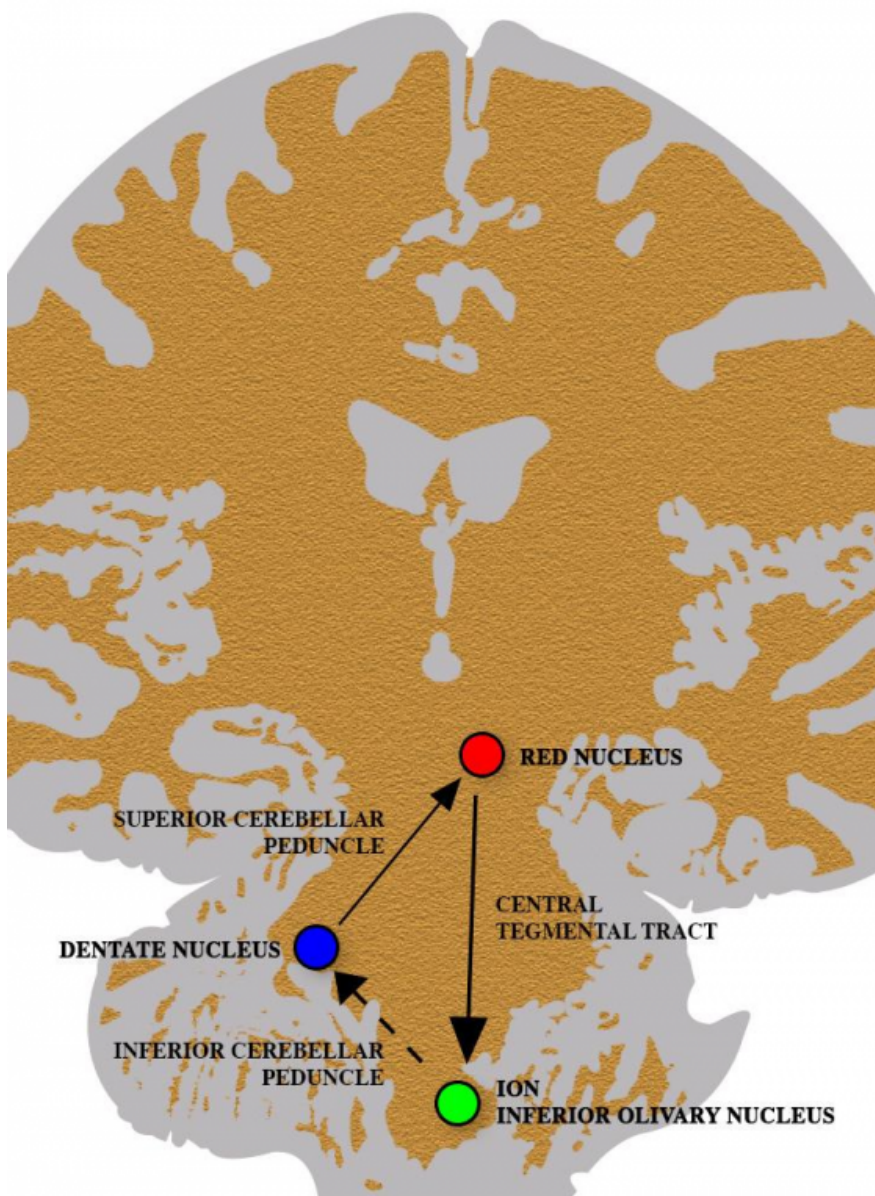
b



Description: Day 270. Interval development of an expansile lesion of the right medullary olive. **Origin:** Augusta University

Figure 5

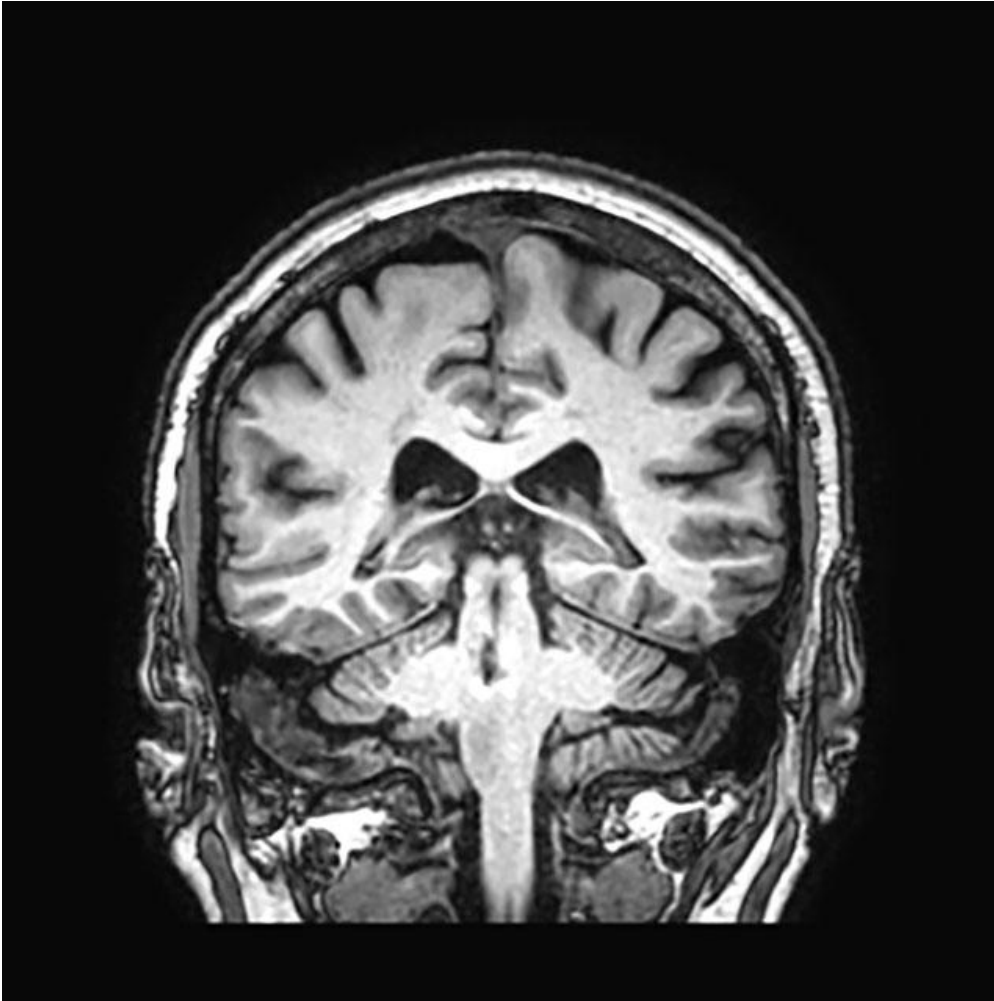
a



Description: The triangle of Guillain and Mollaret, also known as dentato-rubro-olivary pathway. **Origin:** Augusta University

Figure 6

a



Description: Day 270. Interval development of an expansile lesion of the right medullary olive. **Origin:** Augusta University

b



Description: Day 270. Interval development of an expansile lesion of the right medullary olive. **Origin:** Augusta University