

A diagnostic challenge in Midbrain lesions

Nothnagel Syndrome

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CASE HISTORY:

A 38-year-old female with no known comorbidities presented to the OPD with complaints of **blurring of vision** which was sudden in onset and was persisting for 2 days. The patient also reported experiencing **difficulty walking in a straight line**, exhibiting swaying predominantly to the left side. The patient had a history of **head injury** 20 days back.

There was no H/O headache, dizziness, loss of consciousness, ENT bleed, speech abnormalities, dyschromatopsia, impaired coordination, fatigue and generalized weakness.

No H/O ill habits and no relevant family history were present.

GENERAL PHYSICAL EXAMINATION:

The patient is moderately built and nourished. Conscious, cooperative, and well-oriented. No signs of pallor, cyanosis, clubbing, icterus, lymphadenopathy or edema were present. Temperature-98.6°F, Pulse 92 bpm and BP was 118/78 mmHg.

SYSTEMIC EXAMINATION:

Central nervous system:

- Cranial nerve II- **Visual acuity** was decreased (OU-6/24, PH-6/18, and NV-N6). **Reduced field of vision** in the nasal side of the left eye.
- Cranial nerves III, IV, VI- **medial rectus palsy** and **superior rectus palsy** in the Left eye.

- Reflexes: **Jaw jerk**(superficial), **Biceps, Triceps and supinator reflexes** (deep) were found to be **absent** on the right side.
- Gait: **wide base, swaying towards the left**

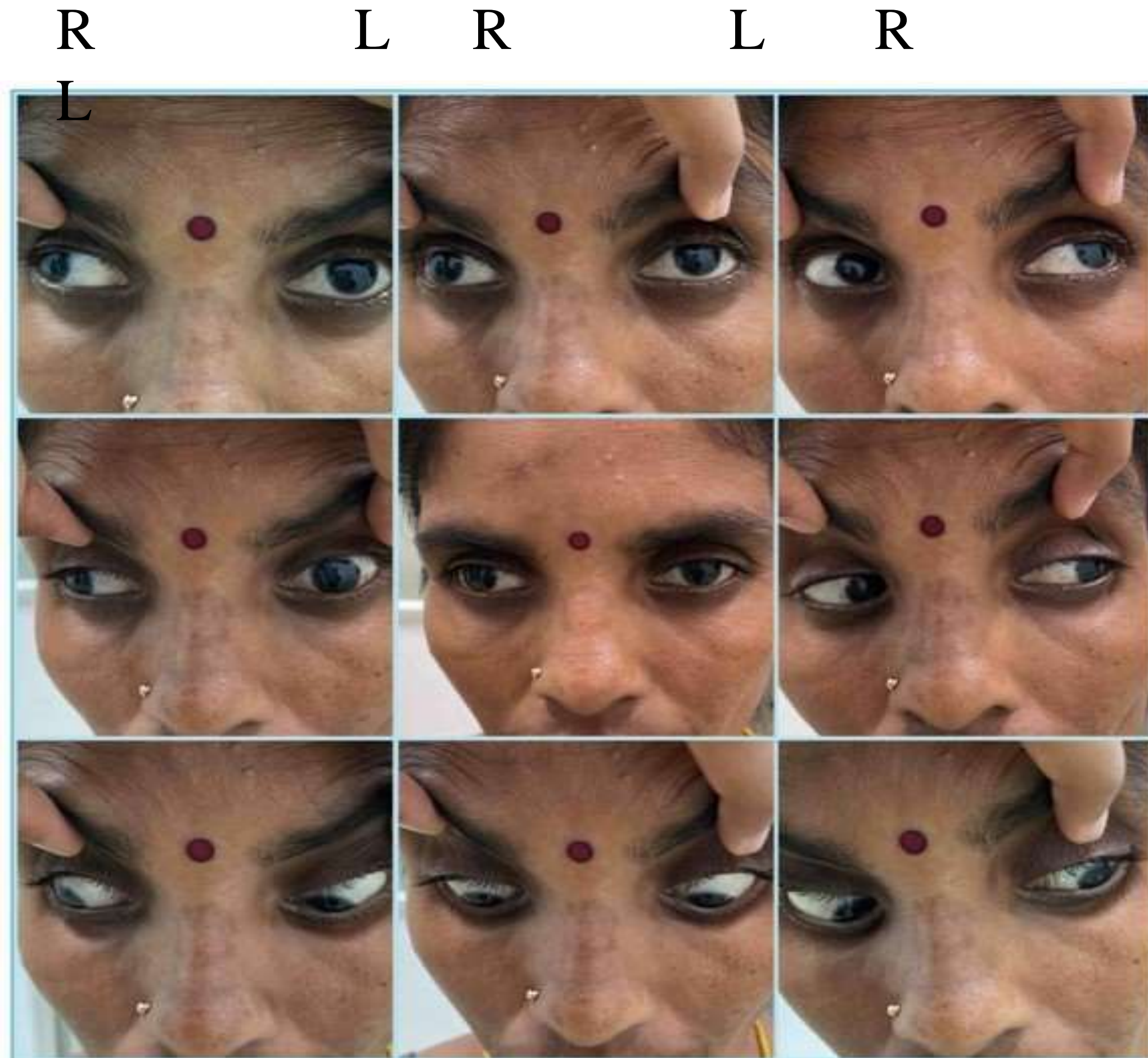
Cardiovascular system: S1 and S2 heard, no murmurs

Respiratory system: normal bilaterally equal vesicular breath sounds heard

Abdominal system: Soft, non-tender, no organomegaly

Nasal field of vision reduced in left eye due to medial rectus palsy and superior rectus palsy

Visible squint seen when trying to look towards right



INVESTIGATIONS:

Radiological investigations:

MRI Brain:

OBSERVATIONS:

CEREBRAL HEMISPHERE: FLAIR hyperintense focus noted in periaqueductal area on left side of mid brain showing subtle restriction on DWI- Likely acute lacunar infarct.

Rest of the brain parenchyma appears normal.

INTRACRANIAL HEMATOMA: Nil.

THALAMI & BASAL GANGLIA: Normal.

VENTRICLES, CORTICAL SULCI, SYLVIAN FISSURES & BASAL CISTERNS: Normal.

CEREBELLUM: Normal.

SELLA: Normal

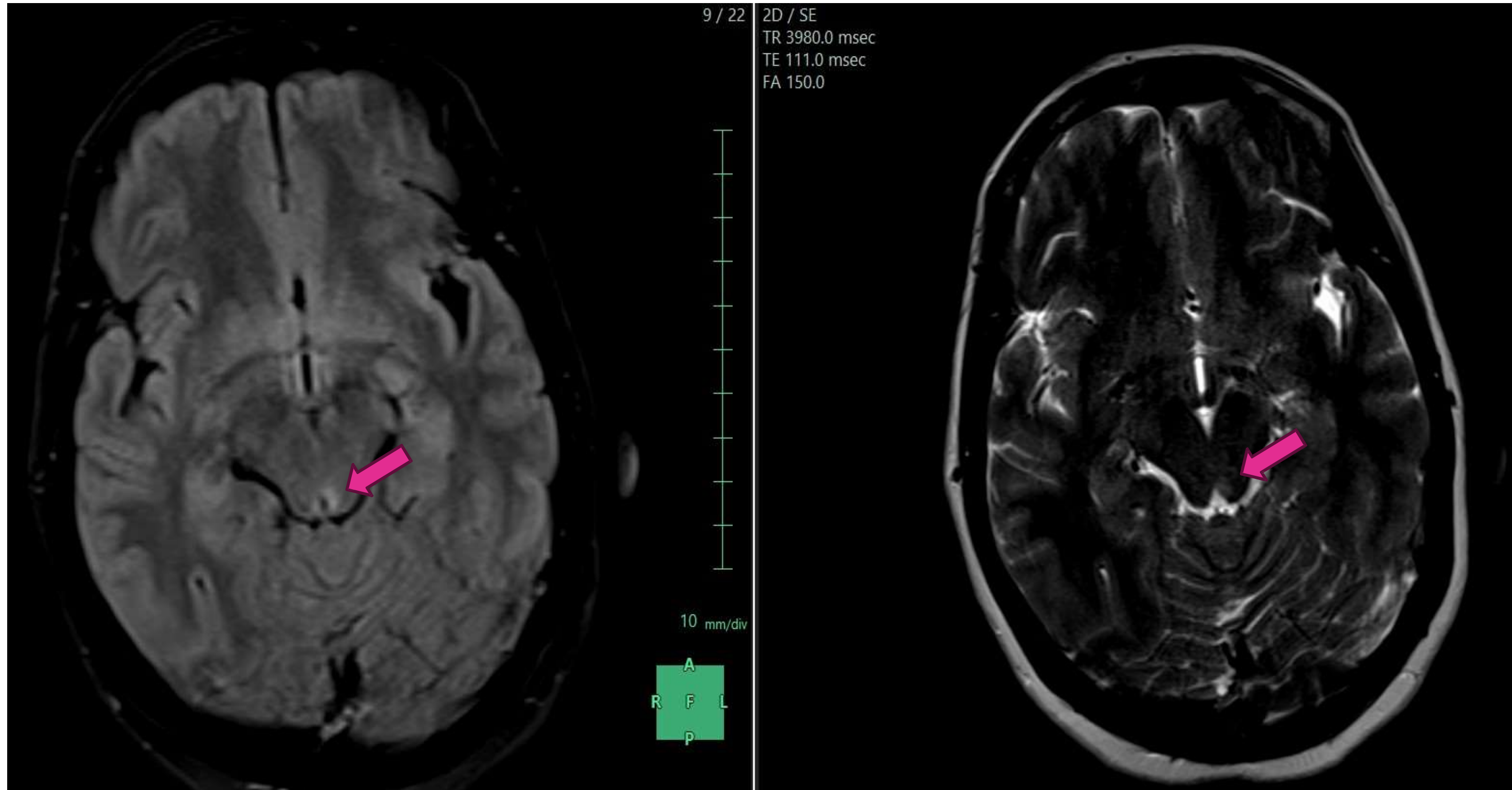
ORBITS: Normal.

VISUALISED INTRACRANIAL ARTERIAL FLOW VOIDS: Normal.

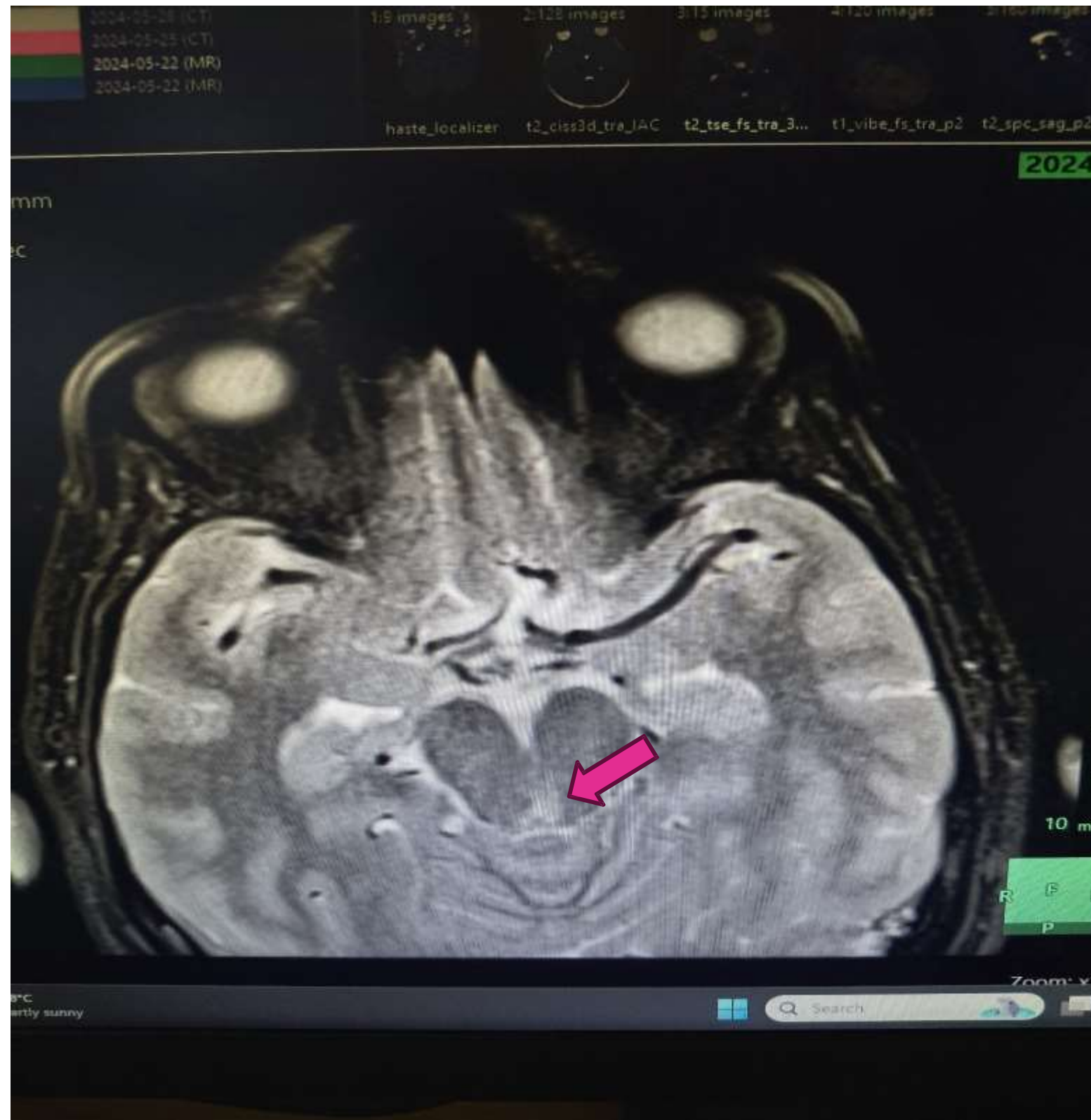
DURAL VENOUS SINUSES FLOW VOIDS: Normal.

SUPERFICIAL SOFT TISSUE/SCALP: Normal.

IMPRESSION: Acute lacunar infarct in periaqueductal area on left side of mid brain.



Arrows indicating the hyperintense foci



Arrow indicating FLAIR hyperintense foci
in midbrain

CT Angiography: Normal

CSF analysis: Normal

CBC: Normochromic normocytic anaemia with
Vitamin B12 deficiency

MANAGEMENT:

Inj. METHYL PREDNISOLONE 1g at morning for 6 days

Tab.ECOSPIRIN 75mg at night for 5 days

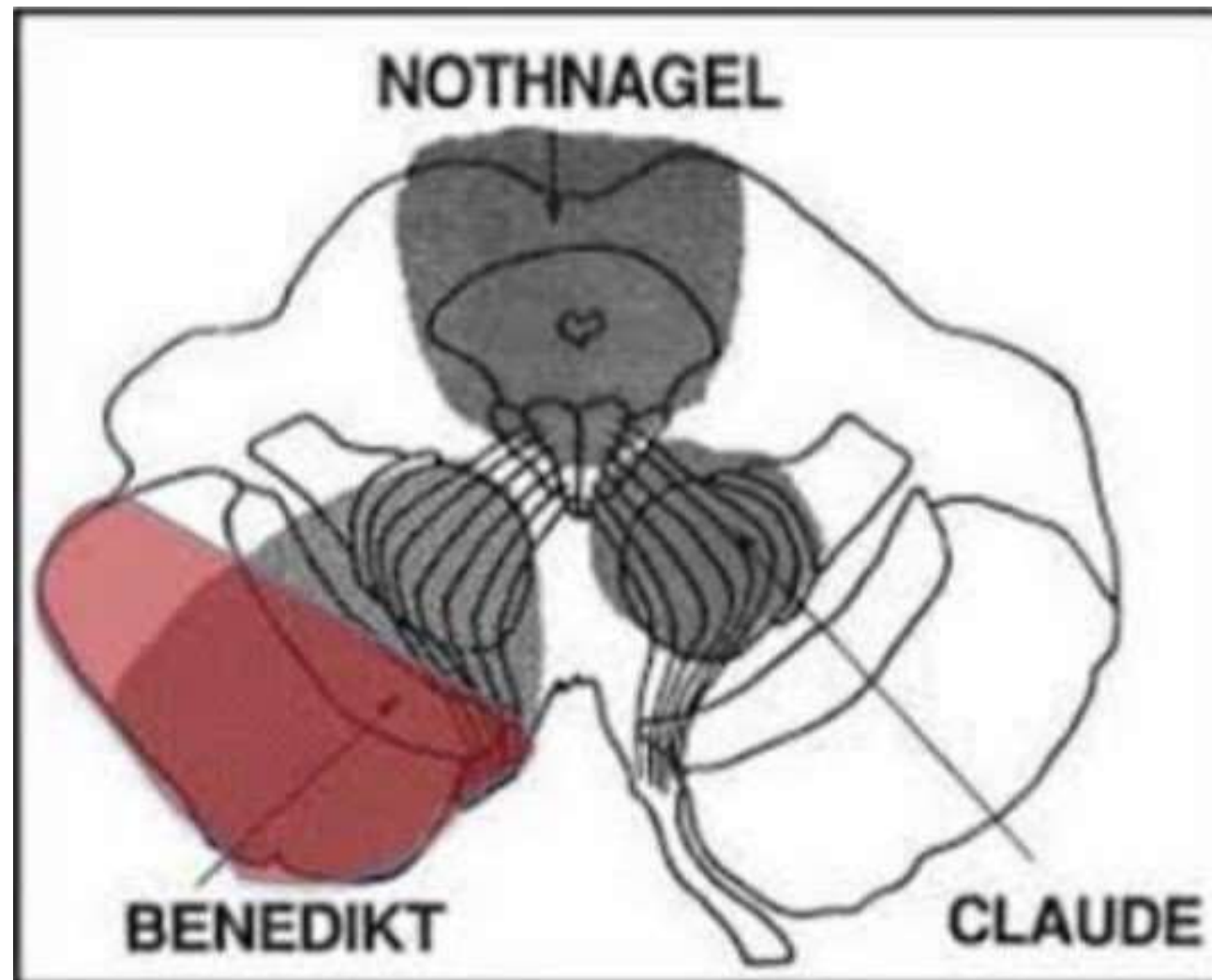
Inj.OPTINEURON at afternoon for 4 days

DISCUSSION:

- The association of unilateral oculomotor palsy of midbrain origin with contralateral ataxia is referred to as the Nothnagel Syndrome.¹
- Nothnagel attributed this clinical syndrome to lesions of the colliculi and compression of the third cranial nerve nuclei.²
- The ataxic component of Nothnagel syndrome is explained by a dorsal midbrain abnormality of either neoplastic or vascular origin, involving the superior cerebellar peduncles.²

- The aetiology of Nothnagel syndrome is attributed to vascular events such as stroke and haemorrhages, neoplasms, oculomotor nerve compression and infarcts³ other potential causes such as demyelinating diseases should also be considered.
- It remains a diagnostic challenge due to its complex presentation and its potential for developing complications. In this case report we highlight the importance of identifying the etiological factor causing the syndrome as early as possible for swift management.

DIFFERENTIAL DIAGNOSIS



Weber's syndrome- Cerebellar peduncle injury
Ipsilateral oculomotor nerve palsy
Contralateral hemiparesis

Benedikt's syndrome- Red nucleus injury
Ipsilateral oculomotor nerve palsy
Contralateral tremors, Ataxia, Athetosis

Claude's syndrome- Red nucleus and Superior cerebellar peduncle injury
Ipsilateral oculomotor nerve palsy
Movement disorders and ataxia

Nothnagel syndrome- Superior cerebellar peduncle injury
Ipsilateral oculomotor nerve palsy
Contralateral cerebellar ataxia
swaying towards injured side

CONCLUSION:

- Nothnagel syndrome, is characterized by a triad of oculomotor impairment, gait ataxia, and cerebellar related signs.
- The CT angiography excluded significant vascular abnormalities and the normal CSF analysis ruled out other conditions such as meningitis or vestibular neuritis.
- Additionally, the patient's biochemical profile revealed decreased serum Vitamin B12 levels, which, while not directly linked to the primary diagnosis, provided an additional layer of diagnostic complexity indicating nerve impairment.

- The management regimen, including high-dose methylprednisolone, antiplatelet therapy, and vitamin supplementation, reflects a targeted approach addressing inflammation, preventing further vascular incidents, and correcting potential nutritional deficiencies.
- This case highlights the significant intersection of acute midbrain infarction and Nothnagel syndrome, underscoring the importance of integrating clinical and radiological findings to manage the condition ensuring the multifaceted nature of the patient.

REFERENCES:

1. Derakhshan I, Sabouri-Deylami M, Kaufman B. Bilateral Nothnagel syndrome. Clinical and roentgenological observations. *Stroke*. 1980 Mar;11(2):177-9.
2. Triarhou LC, Manto M. Nothnagel Syndrome. *The Cerebellum*. 2023 Aug;22(4):487-505.
3. Sharma R, Gaillard F, Thurston M, Nothnagel syndrome. Reference article, Radiopaedia.org <https://doi.org/10.53347/rID-54279>

THANK YOU