Case Report

Vertical one and half syndrome due to mid brain hemorrhage - A case report

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Abstract

Supranuclear ocular movements comprise mainly vertical and horizontal movements; horizontal movements are controlled by the centres located in the pons and vertical movements in the midbrain. The classic one and a half syndrome is caused by a unilateral pontine tegmental lesion that includes the paramedian pontine reticular formation and medial longitudinal fasciculus on the same side. Vertical one and a half syndrome (VOHS), as distinct from horizontal, is also known. Vertical one and half syndrome (VOHS) is very rare and is due to midbrain lesions at the level of superior colliculus. It can be due to a number of conditions, but spontaneous non-traumatic, non-hypertensive, midbrain hemorrhage is an uncommon cause of vertical one and half syndrome and is extremely rare. Acute vertical gaze palsy is one of the main clinical manifestations of rostral midbrain, in the vascular territory of the posterior thalamo-subthalamic paramedian artery. This artery lies between the bifurcation of the basilar artery and the origin of the posterior communicating artery. MRI is a sensitive method for the detection of rostral midbrain strokes and contributes to improve clinico-anatomical correlations in vertical gaze palsies. We report a patient with vertical one and half syndrome due to right midbrain hemorrhage.

Key words

Vertical One and Half Syndrome, Mid Brain Hemorrhage.

Introduction

The neural structures and pathways underlying vertical gaze control are located in mesodiencephalic junction (MDJ), rostral mid-brain reticular formation and pretectal area. They include the rostral interstitial nucleus of medial longitudinal fasciculus (riMLF), the interstitial nucleus of Cajal (INC), the nucleus of Darkschewitsch (ND), and the posterior commissure (PC) with its nuclei. The riMLF, the INC, and the PC are neural structures of the

rostral mid-brain reticular formation which have an important role in the control of vertical gaze.

The riMLF is a wing-shape nucleus, lying dorsomedial to the red nucleus and rostral to the oculomotor nuclei. It contains the neural generators for bilateral vertical saccades. Excitatory burst neurons within this nucleus send collaterals to motor neurons supplying yoked muscle pairs of the two eyes. Axon collaterals responsible for upward saccades reach bilaterally the elevator muscles (superior rectus and inferior oblique), crossing within the oculomotor nucleus, whereas collaterals for downward saccades project only to the ipsilateral inferior rectus and superior oblique which act as depressor muscles.

The INC, the neural integrator for vertical, and torsional gaze lie close and caudal to the riMLF. It is responsible for the vertical smooth pursuit and vertical vestibular ocular reflexes. Neurons contained in this nucleus contribute to hold the eyes in eccentric gaze after a vertical saccade and in the eye-head coordination in the roll plane.

The PC contains the projections from INC to the controlateral oculomotor nuclei and the opposite INC. Furthermore, it contains axons responsible for upgaze originating from the nucleus of the PC and projecting to the riMLF, and to the INC.

The final pathways are the motor neurons of the III and IV cranial nerves. The oculomotor nucleus is placed under the superior colliculus beyond which it extends for a short distance into the gray substance in the floor of the III ventricle while the trochlear nucleus is level with the upper part of the inferior colliculus [1].

Vertical one and half syndrome is very rare and is due to midbrain lesions at the level of superior colliculus Acute vertical upward gaze palsy is one of the main clinical manifestations of rostral midbrain, in the vascular territory of the posterior thalamo-subthalamic paramedian artery. The paramedian arteries arise from the P1 segment of the PCA. They are also known as the "mesencephalic artery" or the "posterior thalamosubthalamic paramedian artery." This artery lies between the bifurcation of the basilar artery and the origin of the posterior communicating artery. Diffusion weighted MRI (DWI-MRI) is a sensitive method for the detection of rostral midbrain strokes and contributes to improve clinico-anatomical correlations in vertical upward gaze palsies [2].

Case report

A 50 years old male patient presented with sudden onset of headache since morning and not able to look upwards since morning. He also complained of diploplia on looking upwards. There was no history of blurring of vision, no history of syncopal attacks, no history of loss of consciousness, no history of convulsions, no history of head injury, no history of palpitations or chest pain, no history of orthopnea, platypnea, No history bladder/ bowel incontinence, No history of bleeding or clotting disorder**,** no history of any drug intake. In past history, there was no history of similar complaints in the past, no history of any drug allergies, no history of TB or contact with TB, no history of asthma, epilepsy, CVA, MI, no history of NIDDM, no history of HTN, history of alcoholism+, No history of any Thyroid disorders. Vitals, Temp – Normal, PR =90/min, regular, BP =120/80 mm of Hg, RR=20/min.

General examination showed that patient was conscious, coherent, answering questions well, moderately built, moderately nourished, no icterus, cyanosis, clubbing, koilonychias, lymphadenopathy, pedal edema, no neuro cutaneous markers, no peripheral nerve thickening, no trophic ulcers, head and spine was normal, and there was no tenderness. Thyroid was normal.

CNS examination

Intellectual functions were - normal

Cranial nerve examination

Olfactory nerve - able to perceive odor from both the nostrils.

Both eyes were showing Sunset sign (eyes are deviated to lower quadrant with clear sclera seen in upper quadrant), and Colliers sign (lid retraction).

Optic nerve - Visual acuity: Normal, 6/6 in both eyes, Color vision: Normal

Peripheral visual fields by Confrontation test: Normal in all the directions except for lower quadrant, which was difficult due to the palsies present, Pupils: Equal in size, and not reacting to light on both sides, accommodation reflex present on both sides.

Fundus exam was normal in both eyes and there was no papilloedema.

Occulomotor, Trochlear, Abducens Nerves
Examination of conjugate movements -
Horizontal conjugate movements -
Both right lateral and left lateral horizontal movements are normal
Vertical conjugate movements
There was no upward conjugate movement, but downward conjugate movement was normal.

Examination of Individual muscles
3rd CN – Superior Rectii on both sides are paralyzed. Inferior Rectii, Medial Rectii are normal in both the eyes. Right Inferior Oblique was paralyzed, but Left Inferior Oblique was normal.
4th CN – Superior Obliques were normal on both sides.
6th CN – both lateral rectii were normal.
Other cranial nerves were – normal.

Motor system examination was normal, reflexes: superficial reflexes were normal, corneal and conjunctival were normal, abdominals was present, both plantars showed flexor response, deep tendon reflexes were normal on both sides Sensory system: 1. pain, temperature - normal in all limbs, 2. pressure, vibration, joint sensations normal in all the limbs. There were no cerebellar signs. There were no signs of meningeal irritation, skull and spine was normal. Examination of Cardio Vascular System and Respiratory Systems were normal.

Investigations - CBP – Hb-10.4 gm%,TLC-8000, Plt-2, 40,000, N- 74, L- 24, B-0, E-2, M-0, ESR:06 mm, Bleeding Time- 4 mts, Clotting Time- 6 mts, Prothrombin Time- 12 seconds, CUE –Urine Albumin - Nil, Sugar – Nil, HIV – Non reactive, HbsAg – Negative, HBC – Negative, Lipid Profile – NAD, Bl Sugar - 114mg/dl , Bl Urea – 24 mg/dl, S. Creatinine – 0.6 mg/dl, S. Electrolytes – Serum sodium-128 mmol/L, serum potassium 6.9 mmol/L, LFT-Normal, X Ray Chest PA View – NAD, ECG – WNL, USG abdomen – NAD, MRI Brain – Revealed Rt. Midbrain Hemorrhage (**Figure – 1**).

Diagnosis
CVA – Rt. Midbrain hemorrhage in the vascular territory of the posterior thalamo-subthalamic paramedian artery with upward gaze palsy, and bilateral superior rectii palsies, and rt. inferior oblique palsy – vertical one and half syndrome.

Treatment given
- Tab Nimodip 30 mg 2 TID for one month, later on 1 TID
- Inj. Ampicillin 500mg iv QID
- Inj. Amikacin 500 mg iv BD
- Inj. Metrogyl 500 mg iv TID
- Inj. Optineuron 1 amp iv OD
- I.V. Fluids

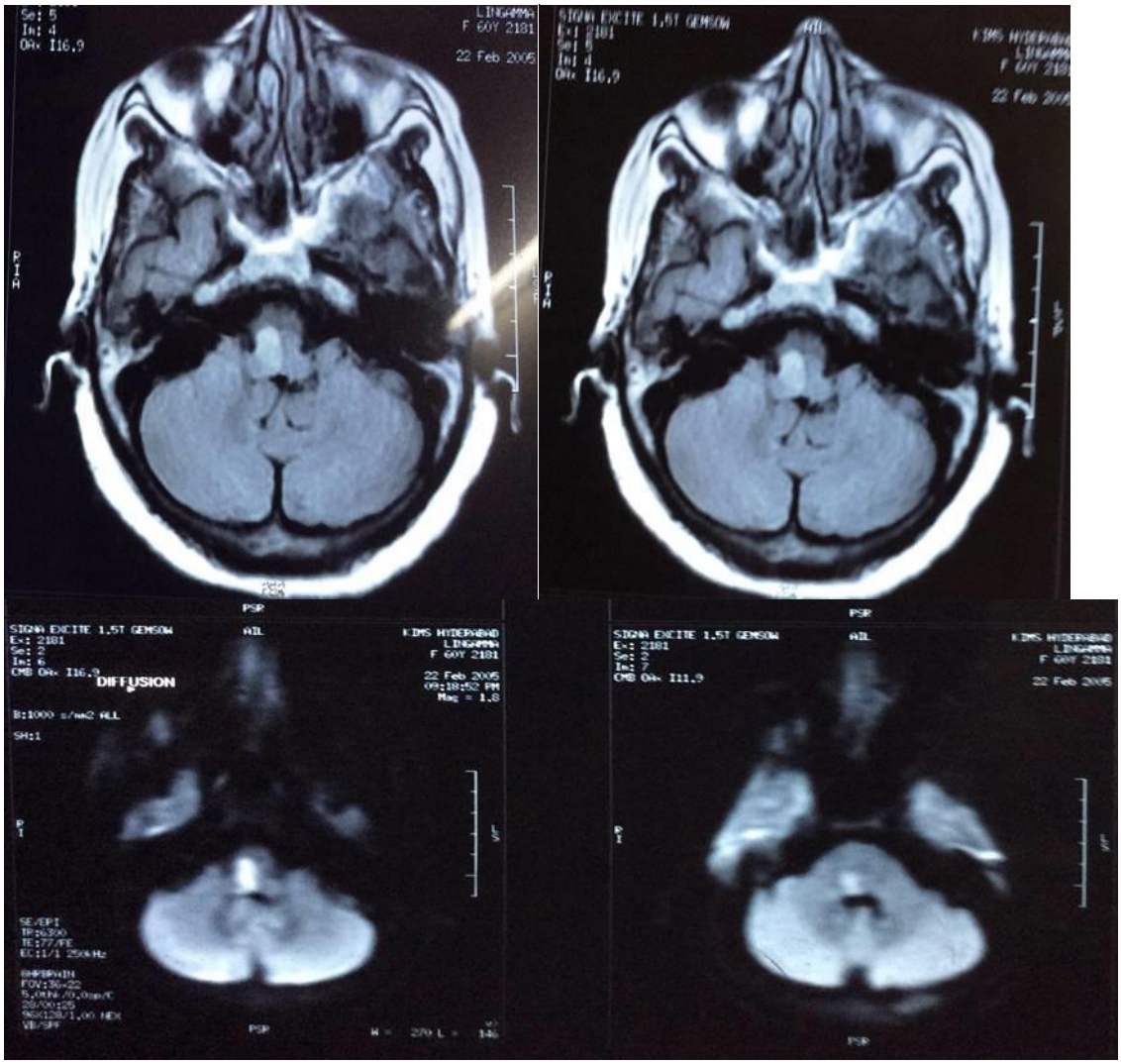
Patient completely recovered when reviewed after one month of treatment.

Discussion

The subnucleus subserving the superior rectus is located in the caudal two thirds of the oculomotor nucleus on the contralateral side. Because decussation of fibres to the superior rectus takes place within the oculomotor nuclear complex, the lesions affecting the nucleus may simultaneously involve ipsilateral superior rectus subnucleus, as well as the crossing fibres, resulting in bilateral superior rectus palsy [3].

Vertical saccades are generated by burst neurons lying in the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF). Each burst neuron projects to motoneurons in a manner such that the eyes are tightly coordinated (yoked) during vertical saccades (**Figure – 2**).

Figure – 1: MRI Brain – Revealed Rt. Midbrain Hemorrhage.



Saccadic innervation from riMLF is unilateral to depressor muscles but bilateral to elevator muscles, with axons crossing within the oculomotor nucleus. Thus, riMLF lesions cause conjugate saccadic palsies that are usually either complete or selectively downward. Each riMLF contains burst neurons for both up and down saccades, but only for ipsilateral torsional saccades. Therefore, unilateral riMLF lesions can be detected at the bedside if torsional quick phases are absent during ipsidirectional head rotations in roll. The interstitial nucleus of Cajal (INC) is important for holding the eye in eccentric gaze after a vertical saccade and coordinating eye-head movements in roll. Bilateral INC lesions limit the range of vertical gaze. The posterior commissure (PC) is the route by which INC projects to ocular motoneurons. Inactivation of PC causes vertical gaze-evoked nystagmus, but destructive lesions cause a more profound defect of vertical gaze, probably due to involvement of the nucleus of the PC. Vestibular signals originating from each of the vertical labyrinthine canals ascend to the midbrain through several distinct pathways; normal vestibular function is best tested by rotating the patient's head in the planes of these canals (**Figure – 3**) [1, 4].

Figure – 2: Anatomic schemes for the synthesis of upward, downward, and torsional eye movements. From the vertical semicircular canals, primary afferents on the vestibular nerve (vn) synapse in the vestibular nuclei (VN), and ascend in the medial longitudinal fasciculus (MLF) and brachium conjunctivum (BC) to contact neurons in the trochlear nucleus (CN IV), oculomotor nucleus (CN III), and the interstitial nucleus of Cajal (INC). (For clarity, only excitatory vestibular projections are shown.) The rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF), which lies in the prerubral fields, contains saccadic burst neurons. It receives an inhibitory input from omnipause neurons of the nucleus raphe interpositus (rip), which lie in the pons [4].

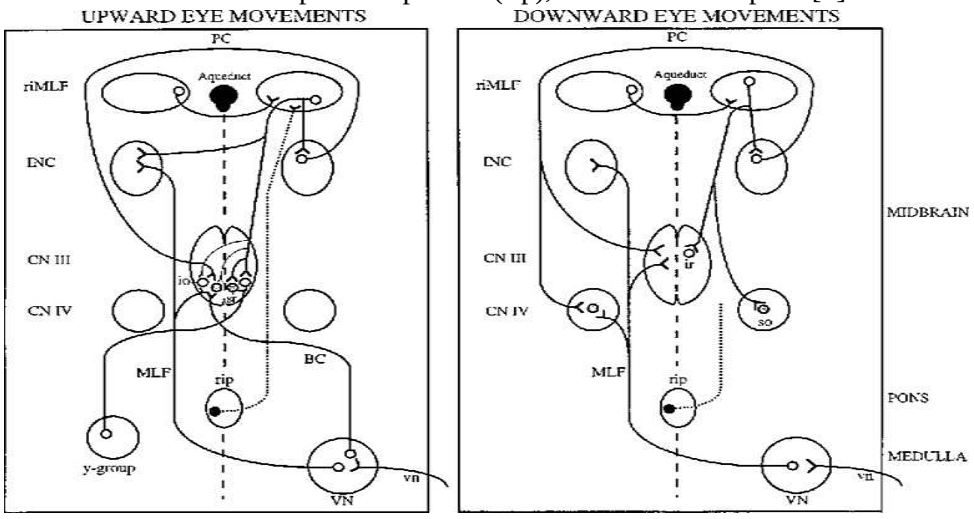


Figure – 3: Sagittal section of monkey brainstem showing location of rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF), interstitial nucleus of Cajal (INC), posterior commissure (PC), and other structures important for the control of vertical gaze. Shaded areas indicate the mesencephalic reticular formation (MRF), paramedian pontine reticular formation (PPRF), and medullary reticular formation (Med RF). Asterisks indicate the location of cell groups of the paramedian tracts, which project to the cerebellum. III - oculomotor nucleus; IV - trochlear nucleus; VI - abducens nucleus; CG - central gray; MB - mammillary body; MT -mammillothalamic tract; N III - rootlets of the oculomotor nerve; N IV - trochlear nerve; N VI - rootlets of the abducens nerve; ND - nucleus of Darkschewitsch; NRTP - nucleus reticularis tegmenti pontis [4].

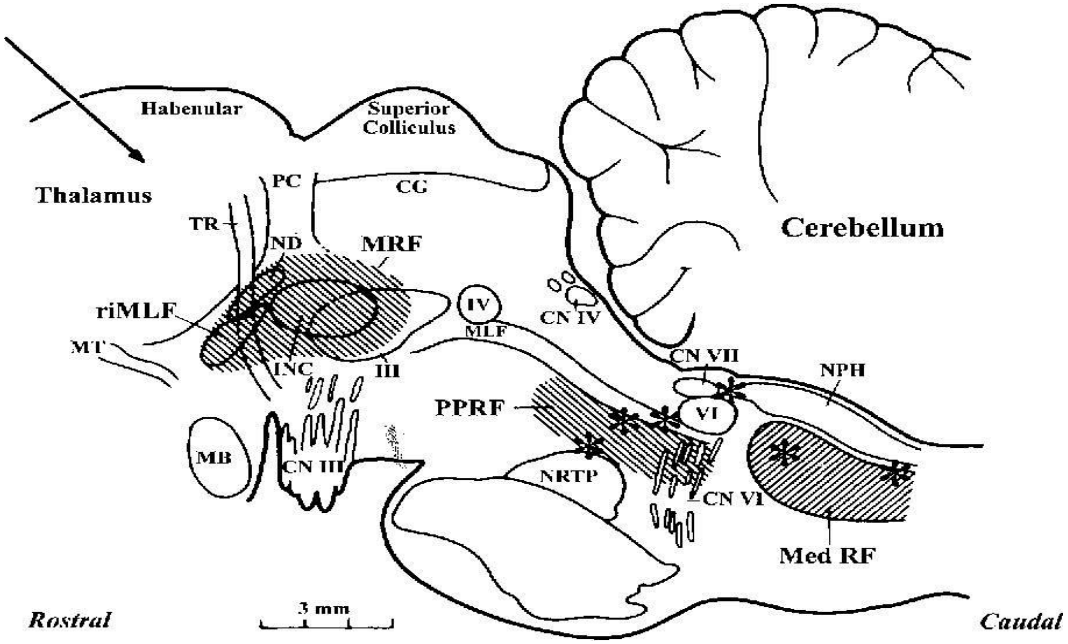


Figure – 4: Midbrain blood supply (https://memorangapp.com.)

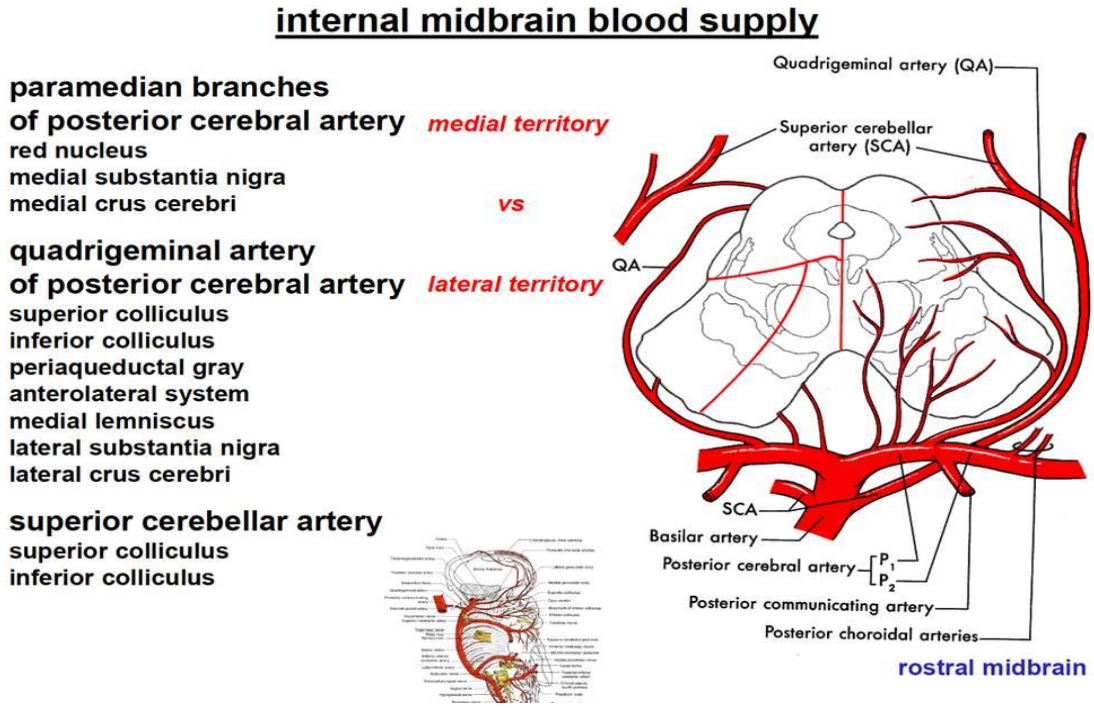


Figure – 5: Vertical eye movements (h): riMLF controls contralateral upward and ipsilateral downward gaze [15].

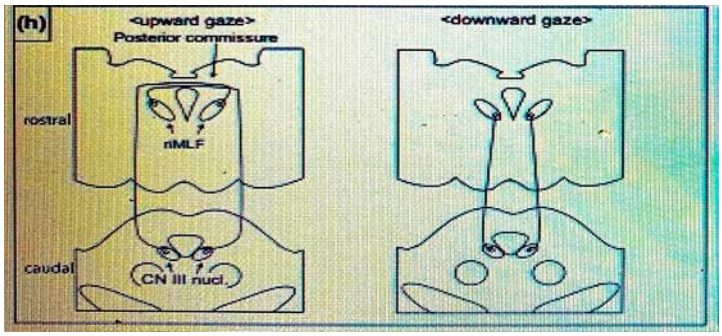
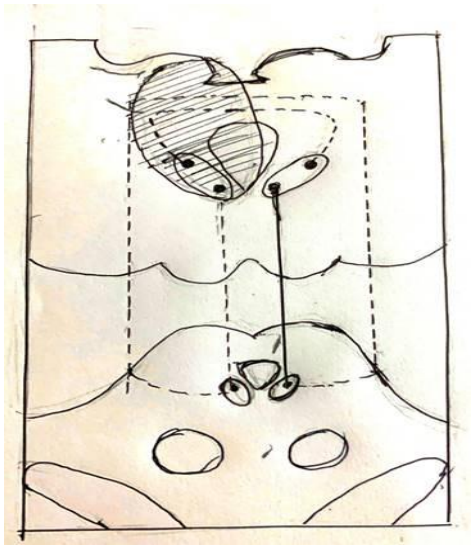


Figure – 6: The present case - Classical VOHS involves bilateral upward and ipsilateral downward gaze (dotted lines) by a mid brain lesion (Right eclipse).



The vertical gaze palsies have been attributed to associated lesions of vertical eye movement control centers in the rostral midbrain. The frequent coexistence of both midbrain and paramedian thalamic lesion is related to their vascular supply; a single vessel arising near the top of the basilar may branch to supply both the paramedian region of the thalamus and the rostral medial mesencephalon. Alternatively, isolated medial thalamic or midbrain lesions can occur in individuals in whom the paramedian peduncular arteries arise separately from the paramedian thalamic vessels (**Figure - 4**). In patients with lesions of the midbrain/thalamic junction, clinical features can be correlated with lesion location with the use of MRI [5].

Etiology

A lesion of the vertical gaze pathways can occur due to number of causes, which are very rare: Tumor- pineal germinoma or teratoma, Hydrocephalus- aqueductal stenosis leading to dilation of third ventricle and enlargement of the suprapineal recess with pressure on the posterior commissure, Vascular- midbrain and/or thalamic hemorrhage or infarction, subdural hematoma, aneurysm. The posterior thalamo-subthalamic paramedian artery is a single perforating artery that supplies both riMLF, Metabolic- Niemann-Pick Type C, Gaucher disease, Tay-Sachs disease, maple syrup urine disease, Wilson disease, kernicterus, Drug-induced- barbiturates, carbamazepine, neuroleptic agents, Degenerative- progressive supranuclear palsy (usually downward gaze palsy), Huntington disease, cortical basal degeneration, diffuse Lewy body disease, parkinsonism, hereditary spastic ataxia, Infectious – Whipple disease, encephalitis, syphilis, tuberculosis, Miscellaneous- multiple sclerosis, hypoxia, trauma, mesencephalic clefts, benign transient form of child hood [6, 7].

There are very few case reports regarding vertical gaze palsies, more so with vertical one and half syndrome (VOHS) [8, 9, 10, 11, 12, 13, 14, 15]. In the present case, the patient presented with vertical one and half syndrome (VOHS), due to rt. midbrain hemorrhage in the vascular territory of the posterior thalamo-subthalamic paramedian artery with upward gaze palsy, and bilateral superior rectii palsies, and rt. inferior oblique palsy (**Figure – 5, 6**).

Conclusion

A patient with hemorrhage in midbrain on the right side suffered a conjugate upgaze palsy associated with a monocular paresis of downward gaze in the ispsilateral eye, the Vertical One-and-a-Half Syndrome (VOHS). Conjugate horizontal gaze were normal. A complex disturbance of vertical gaze is due to a unilateral midbrain hemorrhage. Patient was kept on nimodipine. He made remarkable improvement.

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