

Case Report: Locked-In Syndrome Due To Basilar Thrombosis

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Abstract: Locked-in syndrome is a rare neurological disorder in which there is complete paralysis of all voluntary muscles except for the ones that control the movements of the eyes.

We reported the case of is a 42 year old gentleman, not known to have any medical illnesses Admitted to ER complain of dizziness and slurred speech for 2 hours, and then he had episodes of jerky movement (seizure like) for few minutes, without loss of consciousness.

Conclusion: This case highlights the challenges in diagnosing locked-in syndrome, emphasizes the key to aid diagnosis as well as the management and prognosis.

Keywords: Locked in syndrome, basilar artery thrombosis, basilar artery thrombosis management, Prognosis.

I. INTRODUCTION

Locked-in syndrome is a rare neurological disorder in which there is complete paralysis of all voluntary muscles except for the ones that control the movements of the eyes. The patient is aware but cannot move or communicate verbally due to complete paralysis of nearly all voluntary muscles in the body except for vertical eye movements and blinking. Locked-in syndrome caused basilar artery thrombosis generally rare cause associated with trauma, vascular, or cardiac malformation. It is caused by damaged to the pons; which is a part of the brainstem that contains nerve fibers that relay information to other areas of the brain. The basilar artery is one the main vessel of the posterior circulation that may produce nonspecific symptoms ranging from headache or vertigo to hemiplegia, coma or cranial nerve palsies and locked-in syndrome. ⁽¹⁾

II. CLINICAL CASE

Finale diagnosis:

Locked in syndrome post basilar artery occlusion.

The patient is a 42 years old gentleman, not known to have any medical illnesses Admitted on 14.feb.2016 as a case of basilar artery occlusion.

Hospital Course:

The patient presented to our ER C/O dizziness and slurred speech for 2 hours, and then he had episodes of jerky movement (seizure like) for few minutes, without loss of consciousness. In the ER, his LOC deteriorated, started to become unresponsive with jerky movements, he was intubated and fully sedated and transferred to ICU and was under their care.

Physical examination:

The patient was in severe distressed with deteriorated conscious, not communicating, only eyes movement and blinking contact, he was not able to speak along with jerky movement of both upper and lower limb.

Glasgow Coma Scale (GCS) was 4 over 15 with unable to maintained airway so tracheostomy tube was inserted

Chest: vesicular breathing, no added sounds

CVS: s1+s2+0, no added sounds

No bedsores

Tracheostomy tube no signs of bleeding or oozing

CT BRAIN ANGIO (Computed tomography angiography (CTA) was done;

The Report: Loss of opacification and filling defect is seen in the basilar artery most likely due to basilar artery thrombosis.

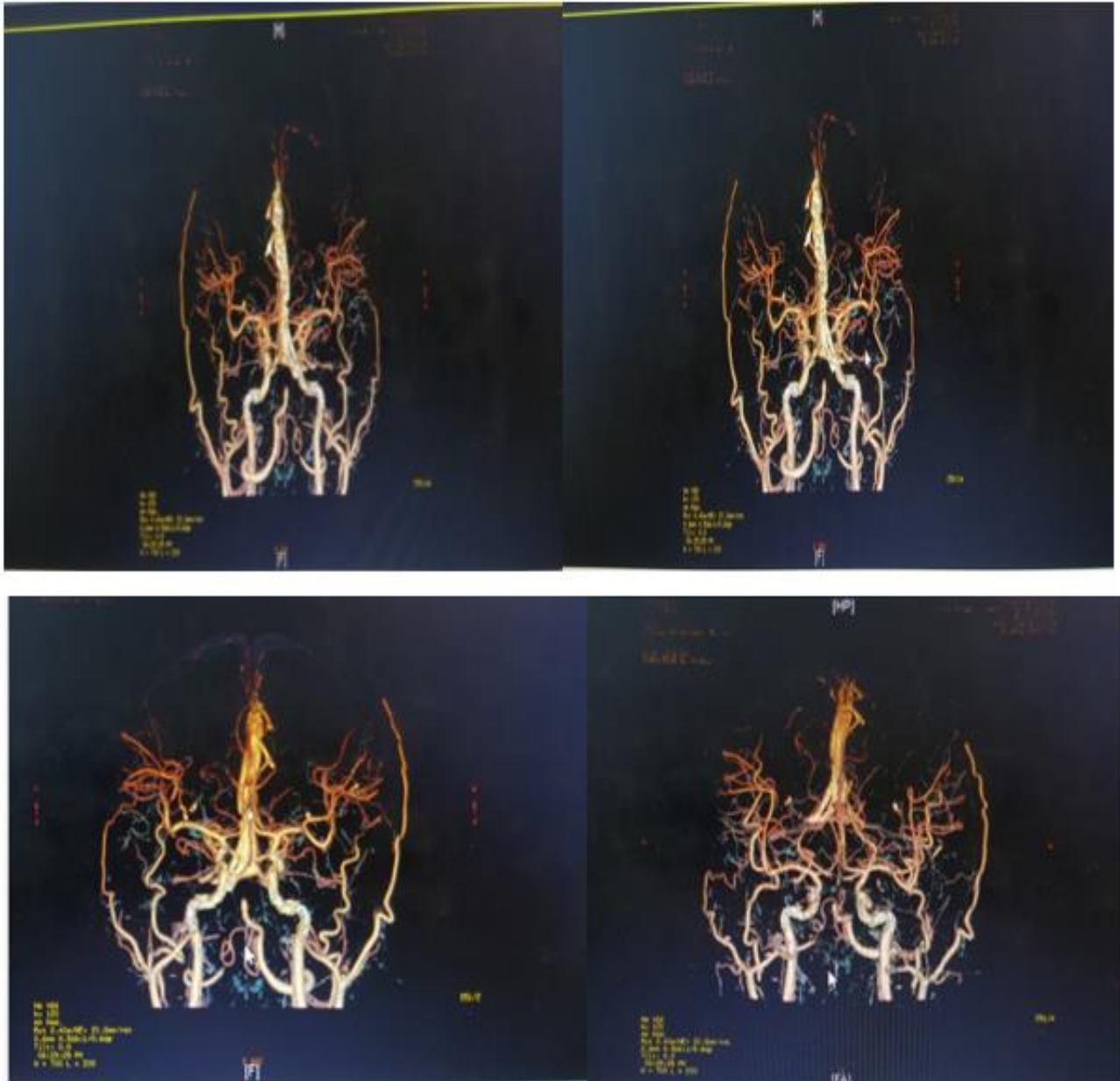


Figure 1: CT BRAIN ANGIO show: a loss of opacification and filling defect is seen in the basilar artery most likely due to basilar artery Thrombosis

On 15.feb.2016, a Basilar Artery thrombectomy was done and sedation was stopped but the patient still unresponsive. Repeated CT showed: III-defined hypodensity in posterior fossa more at left cerebellar hemisphere represent acute infarction. After that he was unresponsive, his GCS 4\15, so tracheostomy was inserted by thoracic surgery.

MRI brain done on February 24; showing: Interval development of hemorrhagic transformation on top of the known extensive posterior circulation infarction at the brain stem and bilateral cerebellar hemispheres.

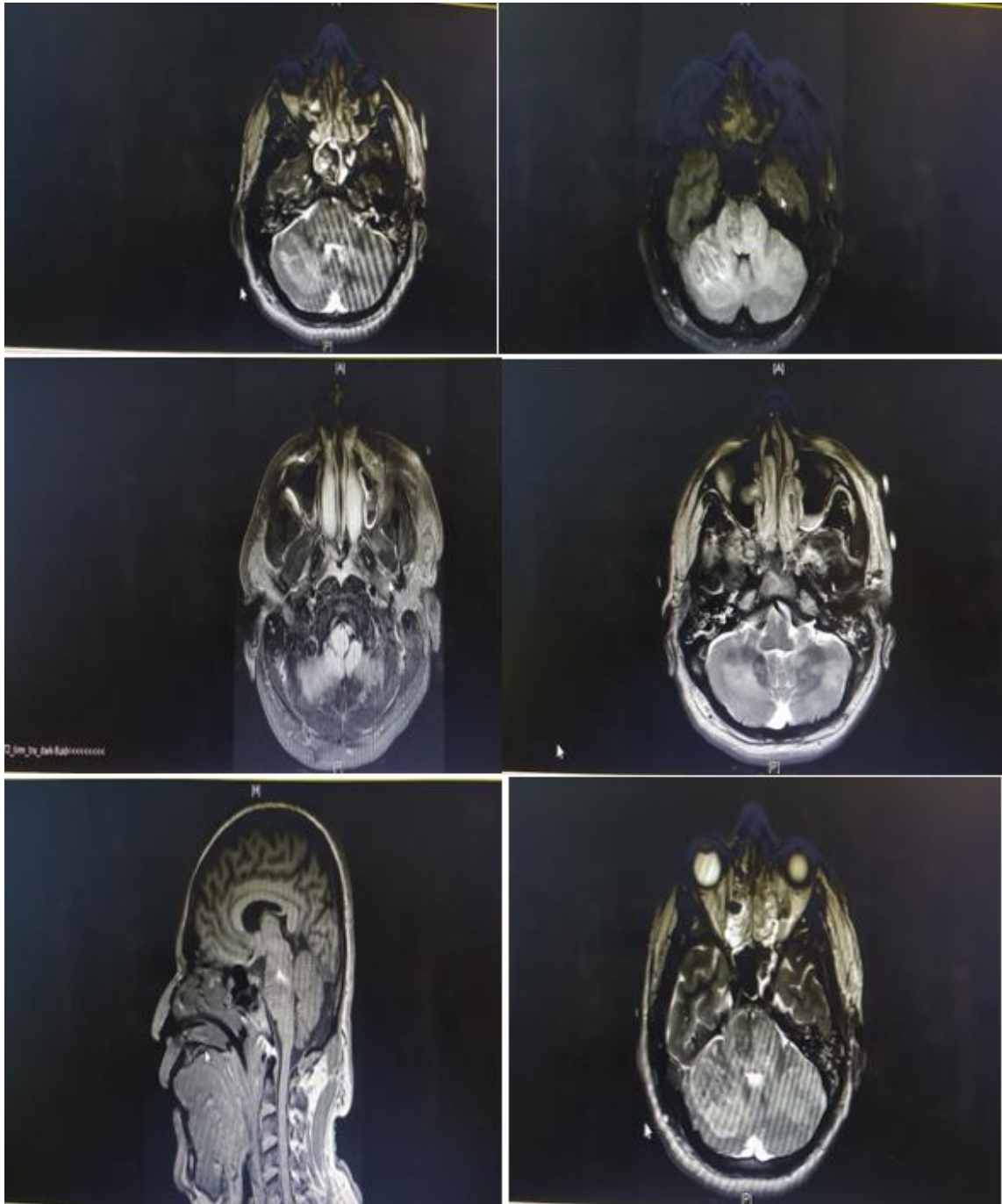


Figure 2: MRI brain images show: Interval development of hemorrhagic transformation on top of the known extensive posterior circulation infarction at the brain stem and bilateral cerebellar hemispheres.

He was then stable, and shifted to the ward on tracheostomy tube.

The patient was seen by **swallowing assessment**, who advised for long term feeding through PEG tube. **PEG tube** was inserted on the 26 th of JULY 2016. He is tolerating feeding through PEG tube. He is on adult feeding formula neutrnl

In the ward he developed patient had multiple infections in his course of admission.

Latest infection was aspiration pneumonia on 16/7/2016 evident on CXR showing right middle zone opacity He was afebrile, no leukocytosis. He completed a course of tazocin® (Piperacillin/tazobactam) 4.5 Gram IV Q6h. and completed another course of Tazovin® 4.g Gram IV Q6h.

The patient has long-standing **tachycardia**. Cardio team were consulted to rule out other causes of tachycardia. In addition, we need to rule out subclinical seizure.

Moreover, we prescribed analgesia if he is in pain.

He had flickering of both eyes (abnormal eye movements), primary team administered midazolam and it resolved completely (? seizure).

Latest CT BRAIN of 16JUL-2016: CLINICAL DATA: SEUZIRE. TECHNIQUE: NON-ENHANCED CT BRAIN. FINDINGS: Comparison was made with previous similar study dated 16th of Feb 2016. The patient is known case of basilar artery occlusion leading to extensive infarction at the brain stem predominantly in the pons as well as bilateral cerebellar hemispheres. There is no acute territorial intracranial hemorrhage. There is again noted well-defined hypodensity in posterior fossa more at left cerebellar hemisphere with effacement of the cortical sulci. Areas of hypodensity are seen in the pons. The ventricles are normal in size and position. No mass effect or midline shift.

The rest gray/ white matter differentiation is grossly maintained.

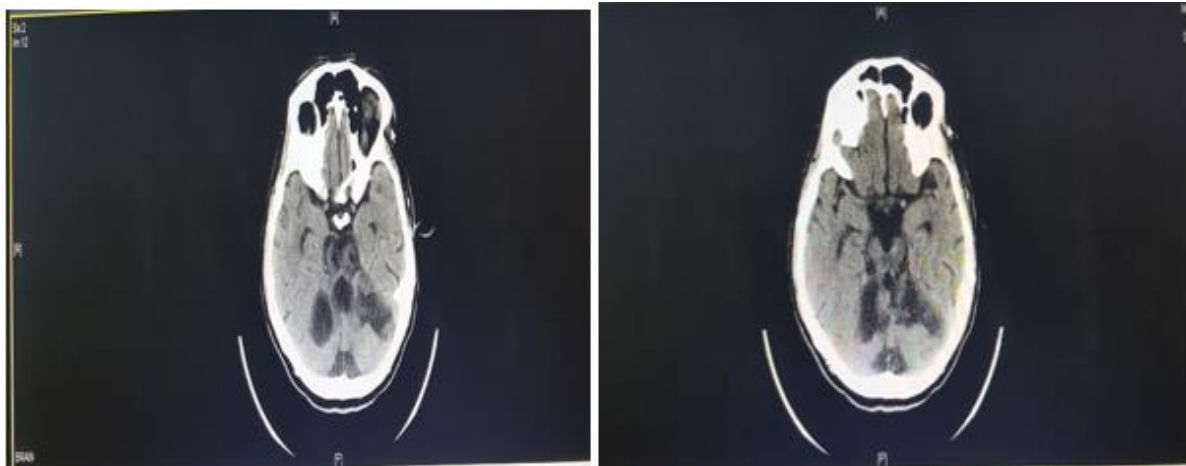


Figure 3: NON ENHANCED CT BRAIN showed: There is again noted well-defined hypodensity in posterior fossa more at left cerebellar hemisphere with effacement of the cortical sulci.

EEG 18-JUL-2016 showed slowing only.

Patient discharged on:

- Acetylcysteine 20% inhalation solution 200 mg + Sodium Chloride 0.9% 3 mL
- Aspirin, 81 mg, 1 tab, Oral, Daily
- Enoxaparin, 40 mg, 0.4 mL, Subcutaneous, q24hr
- Propranolol, 20 mg, 2 tab(s), Oral, BID
- Levetiracetam, 500 mg, 1 tab(s), Oral, q12hr
- Simvastatin, 20 mg, 1 tab(s), Oral, HS

Provided with: adult diaper, under pad, gauze, thermovent for tracheostomy and suction tube, adult feeding for formula (Neutrin 1).

Recommendation: the team recommended that the patient has a good nursing care and to be follow up with neurology for seizure.

III. DISCUSSION

In this case, the patient developed locked in syndrome due the basilar artery thrombosis leading to complete absence of voluntary movement with cognitive function intact.

Locked-in syndrome caused basilar artery thrombosis most commonly related to trauma, vascular, or cardiac malformation, as the basilar remain a major supply of posterior circulation; patient with basilar artery thrombosis commonly presented with sudden and dramatic neurological impairment mostly sudden vertigo, dysarthria, headache and motor deficits such as quadriparesis ,hemiparesis and altered consciousness. [1].

As causes, the most common risk factor is hypertension, hypertension is found to be the reason of 70% of cases. It followed by diabetes mellitus, peripheral vascular disease, coronary artery disease, cigarette smoking, and hyperlipidemia.

As diagnosis of locked-in syndrome is usually made clinically. A variety of tests may be performed to rule out other conditions. Diagnostic approach CT brain none enhanced is one of first test to be done as in this case as may show hyperdense basilar sign of acute presentation, however it is less sensitive. While enhanced CT show filling defect. CT angiography is the gold standard and helpful in identifying occluded and dolichoectatic vessels. In addition, angiogram should be used as tool helpful in assisting treatment decision making.

MRI is more sensitive than CT scan; showing helpful findings include lesions that suggest micro bleeds, tumors, vertebral/basilar dissections and vertebral/basilar dolichoectasia,. A magnetic resonance angiogram (MRA) as approved can identify vertebral or basilar occlusion as it have sensitivity of as high as 97% and a specificity of as high as 98%.

Recanalization of acute basilar artery occlusion led to reduction in mortality by 2-fold and reduction in the risk of death by 1.5-fold. In the absence of clear evidence, treating patients with basilar artery thrombosis in the context of a clinical trial seems most reasonable. Treatment approach of patient of basilar thrombosis is primary fall upon maintaining hemodynamic stability of vital sign, the assessment and management of the airway is the most vital point of patient management at early stage, given the fact of the frequent impairment of consciousness and lower cranial nerves involvement in patients with brainstem ischemia. As the endotracheal intubation is recommended in most patients to keep their airway clear and patent while maintaining normal ventilation. [7].

Today, clinical neurologists would agree that thrombolytic therapy is an efficient strategy for acute basilar artery occlusion. As most Patients with a disease course of more than 3 hours and up to 12 hours should be considered for intra-arterial thrombolysis, provided that ischemic changes are not present on the CT scan.

Following that, the using anti coagulation is to get the benefits of preventing the recurrence emboli or the extension of thrombosis along with antiplatelet.

In addition, the most important thing, the prognosis of patient with basilar artery thrombosis it is generally poor, with the mortality rate reported at greater than 70%. However, the prognosis can be depends on multiple factors and causes, including and not limited to the following: Decreased level of consciousness, tetraplegia, dysarthria, pupillary abnormalities, bulbar symptoms, diplopia, embolism related to cardiac cause and bilateral cerebellar lesions. [2].

IV. CONCLUSION

Locked in syndrome due the basilar artery thrombosis is rare condition and the management of such case is related to the condition and associated cause affecting both prognosis along with treatment outcome ,as locked in patient unable to communicating both verbally and voluntary with fluctuating motor and sensory symptom making management more complicated

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