

A Rare Case of Dumb Rabies Mimicking Guillain Barre Syndrome

D. Raj Mohan Reddy^{1*}, Riya V Raju², Mohammed Kaif³ and Shaik Dastageer Faizur Rahman⁴

¹Department of General Medicine, Osmania General Hospital, Hyderabad, Telangana, India

²Maharajahs Institute of Medical Sciences, Vizianagaram, Andhra Pradesh, India

³Mallareddy Medical College & Hospital, Rangareddy, Telangana, India

⁴Kakatiya Medical College, Warangal, Telangana, India.

Abstract

Introduction: Rabies is a zoonotic disease that spread through animal bites (dog bites in tropical countries). Rabies can be presented in two forms: (i) Encephalitic and (ii) Paralytic rabies. Encephalitic (Furious) rabies is seen in 80% of cases whereas Paralytic (dumb) rabies in 20% of cases. Encephalitic rabies presents with cardinal symptoms like hydrophobia and aerophobia whereas paralytic rabies presents with acute flaccid paralysis resembling Guillain barre syndrome (GBS). Multiorgan dysfunction occurs in both forms but occurs late in paralytic rabies.

Case representation: A 37-year-old male presented with complaints of fever, weakness of both upper and lower limb weakness, neck drops, and hoarseness of voice from 4 days with a history of category III dog bite 30 days back for which he took tetanus toxoid, rabies immunoglobulins and took 5 doses of ARV (anti-rabies vaccine) as prophylaxis. There was no history of hydrophobia or aerophobia. At the time of presentation, his vitals were stable. On examination there was bilateral lower motor facial nerve palsy along with bulbar palsy, hypotonia in both upper and lower limbs, reduced power in all limbs, absent deep tendon reflexes, and plantar reflex showing mute response on both sides. Routine blood tests showed neutrophilic leucocytosis and moderate hyponatremia with serum sodium - 126 meq/L. Nerve conduction studies were inconclusive. MRI brain showed bilateral thalamic hyperintensities. CSF analysis is unremarkable. A provisional diagnosis of GBS was made and IVIG with the dose of 2 g/kg over 5 days was started. Later in the hospital, he was intubated in view of respiratory distress and tachypnea. The patient developed generalized tonic clonic seizures later in the course of the hospital. The patient's GCS gradually deteriorated and went into cardiac arrest. CPR was done and resuscitated. After two days, the patient went into cardiac arrest and could not be revived despite resuscitative efforts. The patient succumbed on the eighth day of admission.

Conclusion: Paralytic rabies is an uncommon presentation of rabies and is more common in rabid bat bites observed in Western countries. It is uncommon in dog bite which is rampant in Asian countries. Moreover, cases of dumb rabies presenting as acute flaccid paralysis resembling GBS pose diagnostic challenges. Hence, a history of dog bites and post-exposure prophylaxis should be sought [2]. The presence of fever and constitutional symptoms should lead to a search for an alternate diagnosis [3]. In Western countries, even the absence of a bite should raise the suspicion of rabies. Paralytic rabies is a common presentation in patients presenting with adequate post-exposure prophylaxis. Even though rare, vaccine-induced GBS should be taken into consideration for diagnosis [2].

Keywords: Paralytic Rabies; Acute Flaccid Paralysis; Dog Bite

***Correspondence to:** D. Raj Mohan Reddy, Department of General Medicine, Osmania General Hospital, Hyderabad, Telangana, India; E-mail: rdrajmohan@gmail.com

Citation: Reddy RM, Raju RV, Kaif M, Rahman SDF (2023) A Rare Case of Dumb Rabies Mimicking Guillain Barre Syndrome. *Neurol Sci Neurosurg*, Volume 4:1. 133. DOI: <https://doi.org/10.47275/2692-093X-133>

Received: May 16, 2023; **Accepted:** June 28, 2023; **Published:** June 30, 2023

Case Report

A 37-year-old male from south India presented with complaints of fever, and weakness of lower limbs followed by upper limbs for 4 days. His weakness in limbs was sudden in onset and rapidly progressive. He also complained of difficulty swallowing both solids and liquids. The patient was unable to produce a voice and drooping of the neck was present. History of nasal regurgitation while attempts of feeding were present. He had been suffering from fever from the day of onset of weakness which was low grade associated with an evening rise in temperature.

There was no history of paraesthesia, muscle pains, and sensory loss. No history of bowel and bladder incontinence and no other autonomic symptoms like sweating, palpitations, syncope, and dry skin. History related to other cranial nerve dysfunction like loss of smell, blurring of vision, diplopia, difficulty in chewing, and hearing loss is not present.

No H/o hydrophobia and aerophobia.

The patient was bitten by a stray dog on his right calf with category III exposure and the status of the dog post-bite is unknown. He took tetanus toxoid and five doses of anti-rabies vaccine, and he took equine rabies immunoglobulins for rabies along with routine wound care. He suffered from oral carcinoma four years back for which he took chemotherapy and radiotherapy. He was not diabetic, or hypertensive, and no other comorbidities were present. He was not on any medication.

At the time of admission, the patient was conscious and was unable to produce a voice, obeying oral commands. His vitals at the time of admission, Temperature - 99.20 °F, Pulse rate - 94/min, Blood pressure - 130/80 mmHg in the supine position, lungs were clear and there were no additional heart sounds.

On cranial nerve examination, bilateral lower motor neuron type of facial palsy was present and gag reflex was absent on both sides;



palatal symmetry was maintained. Another cranial nerve examination was normal. Hypotonia on both upper and lower limbs was present. No wasting of muscles was observed. The power of the upper limbs at all joints was 1/5 and the lower limb was 0/5. Deep tendon reflexes (biceps, supinator, triceps, knee, and ankle) are absent. Plantar reflexes on both sides were mute. No sensory and cerebellar disturbances were observed. No heart rate variability or postural variations in blood pressure was observed. Other system examinations were normal. His single breath count at the time of admission is 20.

A provisional diagnosis of GBS was made and was started on Intravenous immunoglobulin 2 g/kg over 5 days. On the third day in the hospital, he was intubated in view of respiratory distress and was put on ventilatory support. The next day he developed one episode of seizure generalized tonic clonic seizure. His sensorium deteriorates day by day and two days later went into cardiac arrest. Cardiopulmonary resuscitation was done and a return of spontaneous circulation was attained.

Investigations

Complete blood picture showed 13,800/ μ l, 14.5 g/dl, platelets - 2,50,000/ μ l, arterial blood gas analysis before intubation showed pH - 7.118, pCO₂ - 70.5, pO₂ - 92.3, HCO₃⁻ - 18 meq/L. ESR - 25 mm, CRP - 17.5 ng/dl. Biochemical profile: serum urea - 38.5 mg/dl, serum creatinine - 0.71, total bilirubin - 0.69, direct bilirubin - 0.29, alanine transaminase - 14 u/L, albumin - 4.5 g/dl, serum sodium - 126 meq/L, serum potassium - 4.5 meq/L, serum chloride - 91 meq/L, phosphate - 3.4 mg/dl. Viral markers are negative for HIV, HBV, and HCV. A basic laboratory workup showed raised leucocyte count and hyponatremia. The patient also developed respiratory acidosis on the second day of admission in view of which the patient was intubated and put on ventilatory support.

Nerve conduction studies showed normal distal latencies, and normal conduction velocity apart from reduced SNAPs in the lower limb. Thus, were inconclusive.

CSF analysis shows proteins - 32 mg/dl, and glucose - 62 mg/dl, with 7 cells and all cells showing lymphocytes. A possible diagnosis of paralytic rabies is made based on clinical history, examination, and MRI findings (Figure 1). A notification to higher authorities was sent regarding the same. The patient's GCS has not improved, and the patient's parents have been explained about high chances of mortality and ventilator dependence is explained.

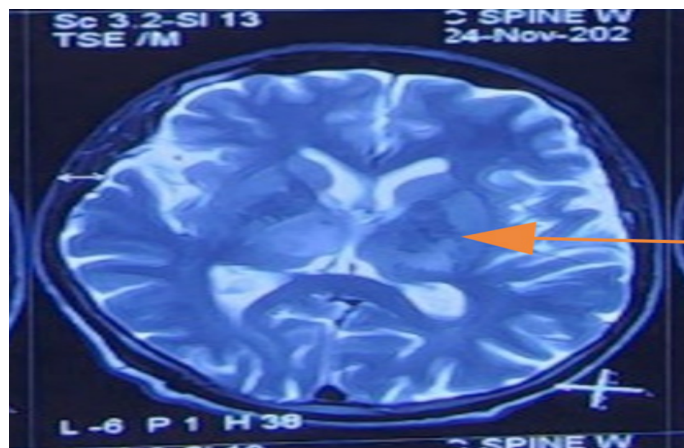


Figure 1: Axial MRI brain T2/FLAIR sequence showing hyperintensities in bilateral thalami, tectum, substantia nigra, and dorsal pons.

After the resuscitation attempts, the patient was on mechanical ventilation, and inotrope support in view of shock. In subsequent days, the patient's consciousness deteriorated, and became fully comatose. The patient's pupils are fully dilated and not reacting to light. The patient succumbed on the eighth day of his admission.

Discussion

Rabies is an acute infectious process that is rapidly progressive involving CNS [4]. Rabies virus is an RNA virus belonging to the family Rhabdoviridae and genus *Lyssavirus* [5].

Pathogenesis: Rabies is transmitted to humans through animal bites. The incubation period ranges from 20 - 150 days. The virus resides in the inoculation site for most time of the incubation. The virus travels retrogradely to axons by passing through the neuromuscular junction and then to the dorsal nucleus and anterior motor neuron to rapidly ascent to the brain. From CNS, the virus transmits through centrifugal spread along nerves to salivary glands, skin, cornea, and other organs [1].

Pathological signs include mild inflammation with monocyte infiltration of the meninges and perivascular infiltration. Negri bodies that contain eosinophilic inclusion bodies containing viral proteins and RNA are rarely seen [1].

Clinical manifestations of rabies start with prodromal features which include malaise, fever, headache, nausea, vomiting, and anxiety. Pain and paraesthesia overexposure site is the earliest specific feature.

The clinical presentation occurs in two forms: (i) Encephalitic rabies (furious rabies) and (ii) Paralytic rabies (dumb rabies). Encephalitic rabies is seen in 80% and classical features include hydrophobia and aerophobia which occur due to early brain stem involvement [1]. Brain stem involvement leads to the loss of neurons that inhibit inspiratory neurons near the nucleus ambiguus leading to exaggerated reflexes. The above mechanism leads to hypersalivation and pharyngeal dysfunction [1]. Paralytic rabies also known as dumb rabies seen in 20% of patients. Dumb rabies presents with acute flaccid paralysis resembling GBS. Typical encephalitic rabies symptoms like hydrophobia and aerophobia both are usually absent [1]. Patients with paralytic rabies survive a little longer but eventually develop multiple organ dysfunction. Paralytic rabies is easily mistaken for GBS [1,6].

In the above patient, the presence of fever and constitutional symptoms along with the history of dog bite raised the suspicion of paralytic rabies. The presence of fever and constitutional symptoms at the time of onset in GBS is very rare and the presence of the above symptoms should always lead to the evaluation of an alternative diagnosis.

Other features of rabies include cardiac/respiratory failure, disturbances of water balance like SIADH or diabetes insipidus, ARDS, cardiac arrhythmia, and GI hemorrhage. In the above patient, reduced serum sodium levels are present reflecting SIADH [3].

Laboratory investigations that aid in the diagnosis of rabies include CSF analysis. CSF analysis shows mild pleocytosis and mildly elevated protein levels. The presence of rabies virus-specific neutralizing antibodies in cerebrospinal fluid in unimmunized patients is diagnostic [2]. Neutralizing antibodies develop late in the course of disease in serum. MRI brain shows nonspecific findings like T2 signal hyperintensities in the brain stem or cortical grey matter areas. In the above patient, thalamic hyperintensities in T2 weighted images are observed [7].



In the suspected case of rabies, the presence of the virus is confirmed by either rabies-specific virus antibodies, RT-PCR amplification, or direct fluorescent antibody testing. Samples used are Cerebrospinal fluid, serum, saliva, and Skin biopsy to detect the virus at nerve endings of the base of hair follicles, and brain tissue (difficult to perform in living patients). The presence of rabies-specific antibodies in CSF is diagnostic of rabies encephalitis regardless of immunization status. Serum antibodies against the rabies virus develop late in the course of disease in previously unimmunized patients [1].

The differential diagnosis for Encephalitic rabies includes viral encephalitis, autoimmune encephalitis, and post-infectious encephalomyelitis. Rabies may be unusually present with neuropsychiatric symptoms and may be misdiagnosed as a psychiatric disorder [8]. There is no established treatment for rabies. Aggressive management with supportive care has been reported to increase survival in some patients [1].

Post exposure prophylaxis for is required for patients with an animal bite which could be rabid. As part of post-exposure prophylaxis along with wound care, Rabies immunoglobulins (RIG) should be given. Both Equine and Human Rabies immunoglobulins can be used [9]. The dose is 20 IU/Kg for humans and 40 IU/kg for equines and should be infiltrated at the wound site and the remaining should be injected at a distant IM site. Anti-rabies vaccine should be given in 4-day courses at 0, 3, 7, and 14. The fifth dose which is given on the 28th day is no longer recommended [9].

In the above case, the patient has developed rabies despite adequate post-exposure prophylaxis. The cause could be due to inoculation may have a large number of free nerve endings and high viral load. Other causes of vaccine failure include missing unidentified wound sites and failure of post-exposure prophylaxis according to WHO protocols. The last reason would be unlikely in the above scenario [10].

Conclusion

Paralytic rabies is an uncommon presentation of rabies among both encephalitic and paralytic forms. Paralytic rabies is more commonly seen in rabid bat bites observed in Western countries and uncommon among dog bite victims (common in Asian countries). Moreover,

cases of dumb rabies presenting as acute flaccid paralysis resembling GBS pose diagnostic challenges. Hence, a history of dog bites and post-exposure prophylaxis should be sought. Presence of fever and constitutional symptoms in patients of acute flaccid paralysis patients should lead to a search for alternate diagnoses other than GBS. In Western countries, even the absence of a bite should raise the suspicion of rabies. Paralytic rabies is seen in patients even with adequate post-exposure prophylaxis. Even though rare, vaccine-induced GBS should be taken into consideration for diagnosis. Paralytic rabies also progresses to multiorgan dysfunction and occurs late when compared to encephalitic rabies. Unfortunately, supportive care is the only line of management.

References

1. Fooks AR, Jackson AC (2020) Rabies: scientific basis of the disease and its management. Academic Press.
2. Wilson JM, Hettiarachchi J, Wijesuriya LM (1975) Presenting features and diagnosis of rabies. *Lancet* 306: 1139-1140. [https://doi.org/10.1016/s0140-6736\(75\)91021-1](https://doi.org/10.1016/s0140-6736(75)91021-1)
3. Wakerley BR, Yuki N (2015) Mimics and chameleons in Guillain-Barré and Miller Fisher syndromes. *Pract Neurol* 15: 90-99. <https://doi.org/10.1136/practneurol-2014-000937>
4. Warrell MJ, Warrell DA (2004) Rabies and other lyssavirus diseases. *Lancet* 363: 959-969. [https://doi.org/10.1016/S0140-6736\(04\)15792-9](https://doi.org/10.1016/S0140-6736(04)15792-9)
5. Hemachudha T, Phanthumchinda K, Phanuphak P, Manutsathit S (1987) Myoedema as a clinical sign in paralytic rabies. *Lancet* 329: 1210. [https://doi.org/10.1016/s0140-6736\(87\)92186-6](https://doi.org/10.1016/s0140-6736(87)92186-6)
6. Feder HM, Petersen BW, Robertson KL, Rupprecht CE (2012) Rabies: still a uniformly fatal disease? Historical occurrence, epidemiological trends, and paradigm shifts. *Curr Infect Dis Rep* 14: 408-422. <https://doi.org/10.1007/s11908-012-0268-2>
7. Kalita J, Bhoi SK, Bastia JK, Lashkar S, Mahadevan A, et al. (2014) Paralytic rabies: MRI findings and review of literature. *Neurol India* 62: 662-664. <https://doi.org/10.4103/0028-3886.149394>
8. Gadre G, Satishchandra P, Mahadevan A, Suja MS, Madhusudana SN, et al. (2010) Rabies viral encephalitis: clinical determinants in diagnosis with special reference to paralytic form. *J Neurol Neurosurg Psychiatry* 81: 812-820. <https://doi.org/10.1136/jnnp.2009.185504>
9. WHO Expert Consultation on Rabies: Third Report. [<https://apps.who.int/iris/handle/10665/272364>] [Accessed June 29, 2023]
10. Wilde H (2007) Failures of post-exposure rabies prophylaxis. *Vaccine* 25: 7605-7609. <https://doi.org/10.1016/j.vaccine.2007.08.054>