

## CASE REPORTS

# PRESUMPTIVE HUMAN PARALYTIC RABIES- A CASE REPORT

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### Abstract:

*Paralytic rabies is not a common form of human rabies and not frequently observed in clinical medicine. The clinical presentation is different from furious rabies and usually there is lack of hydrophobia and aerophobia which makes the diagnosis less certain. The predominant weakness suggest Guillain Barre syndrome as a close differential diagnosis. Regional distribution of rabies virus antigen in rabies patients whose survival periods are more than 7 days or more and magnetic resonance imaging (MRI) of the CNS indicated subcortical and spinal cord*

*predilection sites are recognized presentation of paralytic rabies. There are clinical, electrophysiological, and pathological indications that peripheral nerve dysfunction is responsible for weakness in paralytic rabies. Even in the absence of clinical weakness, abundant denervation potentials with normal sensory nerve conduction studies and proximal motor latencies suggest anterior horn cell dysfunction. Here is a case report of human paralytic rabies that presented in a Medicine Unit of Dhaka Medical College Hospital (DMCH)*

### Introduction:

Furious rabies is a well known entity with classical clinical presentation with very rapid worsening neurological complications leading to universal fatality. Cardinal features of furious rabies, fluctuating consciousness, hydro- or aerophobia and inspiratory spasms, signs of autonomic dysfunction, were seen in all furious rabies patients. In noncanine rabies endemic areas, such as in North America, where bats are the principal vector of rabies, clinical expression may be variable <sup>1</sup>.

Only one or two classical signs of rabies, or even none, may be seen during clinical course in paralytic rabies. Consciousness are usually well preserved until the preterminal phase. Phobic spasms are reported in half of confirmed paralytic rabies patients in few studies. <sup>2</sup> Weakness is the initial manifestation in paralytic rabies, whereas in furious rabies patient it usually seen before coma. There are some unique clinical features associated with three types of Guillain Barre Syndrome (GBS) and paralytic rabies, but some can overlap and thus may

be indistinguishable clinically <sup>3,4,5</sup>. Here is a case report of a presumptive human rabies that presented in Medicine department of DMCH and created lots of confusion and diagnostic dilemma.

### Case report:

A 35 yrs middle age farmer with permanent residence in Joydevpur (rural area), Gazipur, Dhaka presented in medicine department of DMCH with fever for 15 days, weakness of the both lower limbs for 10 days, restlessness, irrelevant talk followed by drowsiness for 5 days. Fever was high grade, intermittent at onset, later continuous in nature. Five days after the onset of fever the developed lower limb weakness. Weakness gradually evolved into complete paralysis of both lower limbs. He also developed acute retention of urine within this period. He has no H/O haedache, vomiting, convulsion etc.

He had a history of dog bite on 9.09.2011 at 10 am. The bite was severe (Grade III) in left foot and left hand. The bite was unprovocative and attendant complaints of multiple incidences by same dog on that

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day. Initially two geese were bitten by the dog and then the victim was attacked. Subsequently another two persons of same village were bitten by the same dog. The lacerated wound of our index case was treated in local clinic (a small private hospital) by wound care with soap and water. He was advised to take vaccination and treated with antibiotics. On 9.09.2011, he took 1st vaccine dose by Verorab (Vero cell culture vaccine) as per Essen schedule (0,3,7, 14,28). He also took 2<sup>nd</sup>, 3<sup>rd</sup> and 4<sup>th</sup> dose on 12.09.11, 16.09.11 and 23.09.11 through IM route. He was not given Rabies Immunoglobulin (RIG) at any period. On 25.09.11 he developed fever which was high grade in nature initially and then the pattern became intermittent with occasional chills and rigor. There was no focal symptoms and sign initially and there was no travel to any malaria, kala azar or filarial endemic zone. He was prescribed locally with paracetamol and antibiotics without any response.

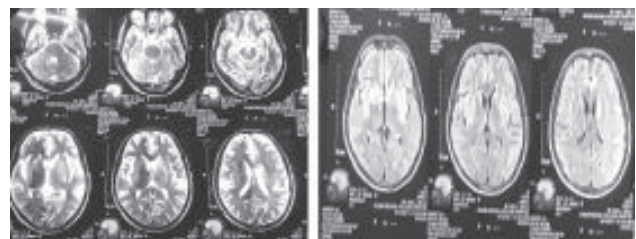


**Fig.-1:** Dog bite at above left ankle joint and left index finger

On 29.09.2011 he felt prostration and mild weakness of lower limbs. He was able to move or maintain activity of daily living on his own and that's why he took no advice initially. But on 5.10.2011 he felt extreme weakness and started incoherent talk. The patient was brought to Medicine outdoor of DMCH but unfortunately he was not advised for admission. Rather few tests were advised and he was treated with anti ulcerant and vitamins. He went home and started irrelevant talk with restlessness and it was followed in next morning with more weakness of lower limbs and partial weakness of upper limbs. His blood sample was taken and tested for S. creatinine and electrolytes. His S. creatinine was 1.4 mg/dl, Na was 123 mmol/l and rest are normal. On 7.10.2011 he became unconscious and developed complete paralysis. He was rushed to DMCH and admitted in a Medicine unit. On examination he was toxic, not anaemic, nonicteric, body temperature of 101<sup>o</sup>F, pulse 100/min, BP-110/80 mHg, unconscious GCS(11), no sign of meningism, flaccid quadriplegia. No cranial

neuropathy, No hydrophobia or aerophobia. Rest of the examination were unremarkable. His local injury was completely healed. But there were multiple scar marks in left leg and left hand.

Investigation showed – Hb-10.8 g/dl, ESR 33 mm, Platelet 150000/L, Leukocyte count 6900/L, Neutrophil 90%, Lymphocyte 6%, SGPT 48 u/l, S. Creatinine 1.1 mg/dl, S. Na<sup>+</sup> 136 mmol/l, S. K<sup>+</sup> 4.4 mmol/l, CSF study: Protein 131 mg/dl, Sugar 116 mg/dl, WBC 4/mm<sup>3</sup>, Lymphocytes 90%, Neutrophils 10%, on Gram's stain and ZN stain no organism seen and VDRL negative. NCV study showed demyelination and remyelination and axonal injury in peripheral nerves. PCR of CSF could not be done due to lack of facility. MRI revealed Hyperdensity in subcortical territory and brain stem. Serum antibody test (IFA) for rabies, REEFIT test in CSF and virus neutralizing antibody (VNA) could not be done due to lack of facility. Viral RNA detection through neck skin and saliva was attempted but could not be done due to his progressive deteriorating clinical course. His status remained same for next 72 hours. A CSF examination was done and serum was taken for freezing. No test for detection of antibody against rabies antigen was done as there is no facility to do the serological or CSF test for rabies in Bangladesh. He was treated with Methyl prednisolone (1 gm daily for 5 days) with Inj Ceftriaxone and metronidazole and inj Omeprazole. On 9.10.2011 at night, patient's condition deteriorated and dyspnoea started. On 10.10.2011 the patient's GCS became 3 and there was huge frothing of saliva from mouth. His vitals were still stable but dyspnoea was worsening. The patient developed decerebrate posturing followed by central pontine hyperventilation. He was diagnosed as a case of Paralytic Rabies with Respiratory paralysis.



**Fig.-2:** MRI revealed hyperdensity in subcortical and brainstem

Consultation with rabies expertise was sought home and abroad. There was constant hyper secretion of saliva throughout the course of his illness. There was refusal of admission of the case in ICU facility due to unavailability of bed. There was no improvement in respiratory failure through the

conservative management by respiratory stimulant. There was no hydrophobia or aerophobia during course of illness but penile tumescence was observed during hospital stay. Milwaukee protocol was not tried in this case as the case was paralytic rabies rather than furious one. Seven days after admission into DMCH he succumbed to death due to respiratory failure. Autopsy was not done due to lack of agreement by attendant.



**Fig.-3:** Paralytic rabies with decerebrate posture.

#### Discussion:

Diagnosis of Rabies on clinical ground alone is difficult and unreliable except when a specific clinical sign of hydrophobia or aerophobia is present. Some patient present with a paralytic or Guillian Barre like syndrome (GBS) or other atypical clinical features<sup>1,6</sup>. Classical signs in furious rabies is of brain involvement include spasms in response to tactile, auditory, visual or olfactory stimuli (e.g. hydrophobia or aerophobia) alternating with periods of lucidity, agitation, confusion and signs of autonomic dysfunction.<sup>1,6,7</sup>. These spasms occur at sometime in almost all rabid patients in whom excitation is prominent. However, spontaneous inspiratory spasms usually occur continuously until death and their presence often facilitates clinical diagnosis.

Excitation is less evident in paralytic Rabies, and phobic spasms appear in only 50% of these patients<sup>1</sup>. During the early stages of paralytic rabies, notable signs include myoedema at percussion sites, usually in the region of chest, deltoid muscle and thigh, and piloerection can be seen. Atypical or non-classic rabies is being increasingly recognized and maybe responsible for under-reporting. In our case the patient presented with fever and progressive paralysis followed by unconsciousness. Initially it was like a GBS presentation although high fever is unlikely to be present in classical GBS while later the loss of

consciousness can also provoke a diagnosis of viral encephalitis. Although the percussion myoedema was not classically present in this case, piloerection and penile tumescence was seen constantly and there was huge frothiness from mouth as the disease progresses. There was no improvement of consciousness observed in this case and as the time goes there was involvement of respiratory function which leads to fatality within ten days.

Diagnosis of rabies depends on High index of clinical suspicion with history of animal exposure and occasionally laboratory tests including Postmortem diagnosis Fluorescent antibody techniques RT-PCR (Reverse Transcription - Polymerase Chain Reaction) and MRI of brain or Antemortem diagnosis by nuchal skin biopsy.<sup>8</sup> In this fatal case, we could not arrange postmortem diagnosis due to lack of permission from patients attendant. In MRI the common finding is clearly demonstrated anterior horn cell involvement in paralytic patient associated with flaviviruses, poliovirus, and West Nile virus<sup>9,10,11,12</sup>.

In human rabies of both forms, spinal cord gray matter and anterior horn cell were involved in

axial gradient-echo T2-weighted images; however, lateral and posterior columns were also affected<sup>13</sup>. In MRI of brain in our case we found hyperintense shadows in T2 image in subcortical and brain stem area which is consistent findings but not specific for rabies.

The antemortem confirmation is even more difficult in Bangladesh due to lack of facilities to do PCR or VNA( virus neutralizing antibody). The nape of neck was targeted to do biopsy but due to progressive deterioration we could not arrange that. The test is confirmatory but lack of sensitivity and even within 3 days the test may turn negative. Although we collected the CSF to do REEFIT test but the laboratories are not well acquitted with the process. In Bangladesh it is now crucial to have these test facilities available as the number of dog bite is enormous in rural setting and every year 2000 cases are having rabies<sup>14</sup>. The tertiary care hospital should be well equipped to confirm the diagnosis of rabies and if necessary for the paralytic one.

The management of rabies patient is supportive one as the fatality is around 100 percent. There has been only 6 cases of furious rabies treated by a specific protocol (Milwaukee protocol) and thereby have recovered completely or partially. In paralytic cases, this protocol is hypothetically not indicated and the expertise opinion was against our index case. So the treatment provided was just supportive and unfortunate to see the patient died with rapidity.

Paralytic rabies is not a common disease and the high index of suspicion is crucial. The GBS and variants and viral encephalitis is the differentials on consideration list. The rapidity of course and fatality within 11 days and flaccid paralysis with unconsciousness on the background of CAT III injury by dog could make the diagnosis of paralytic rabies in our case. This case is the eye opener for clinician who are dealing often with dog bite cases.

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**Conflicts of interest:** None

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