

report of atrial flutter following device closure in an infant which was successfully ablated. In conclusion, caution has to be exercised in closing the atrial septal defects with a device in infants although it has been reported to be a safe alternative(6).

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## Rabies Encephalitis

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*A 12-year-old boy presented with fever and sore throat of 6 days duration followed by vomiting and altered sensorium. He had received 4 doses of antirabies vaccine following a dog bite 4 weeks back. Rabies immunoglobulin was not given. History of hydrophobia and aerophobia were strikingly absent. The possibilities of rabies encephalitis and vaccine induced acute disseminated encephalomyelitis (ADEM) were considered. MRI brain showed exclusive grey matter changes characteristic of rabies. The diagnosis was further confirmed by serological tests.*

**Key words:** *Encephalitis, Hydrophobia, Rabies.*

Rabies should be considered in the differential diagnosis of any case of encephalitis, especially when there is a history of animal bite, signs of excitation or phobic symptoms. But atypical presentations can pose significant problems in the

diagnosis, especially when the patient is comatose and the pathognomonic signs are lacking. We describe a case of encephalitic rabies that presented without the classical symptoms of rabies like hydrophobia or aerophobia. Rabies occurring after postexposure antirabies vaccination creates a diagnostic dilemma when vaccine induced ADEM is a distinct possibility.

#### Case Report

A 12-year-old male child was brought to the Emergency room with vomiting and drowsiness of one day. He also had a history of fever and sore throat of 6 days.

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On examination, he was febrile (temp:101°F) and drowsy. Vital signs were stable. Pupils were equal and reactive bilaterally. Motor system examination revealed hypertonia of all 4 limbs with Grade IV power. All the DTR were brisk bilaterally. Plantar responses were equivocal. Neck rigidity was present. Fundus was normal. There were no cerebellar signs. Other systems were within normal limits.

On further questioning, the parents revealed that 4 weeks ago the boy was bitten on the hand, by a neighbour's unvaccinated dog. He was given 4 doses of chick embryo cell antirabies vaccine, starting from the second day of the bite. Rabies immunoglobulin was not given. He had complained of sore throat, there was no typical history of hydrophobia or behavioral changes.

A provisional diagnosis of rabies encephalitis was made, even though the classical symptoms of rabies were absent. Failure of administration of rabies immunoglobulin after the bite favored this diagnosis. The possibility of vaccine induced ADEM, though rare with the modern cell culture vaccines, was also considered.

Laboratory investigations revealed neutrophilic leucocytosis, normal blood glucose, electrolytes, liver and renal function tests. CSF analysis showed a normal cell count, biochemistry and culture was sterile. CSF analysis for Japanese B antibody was negative. EEG showed generalized slow wave-forms. CT scan head with contrast was unremarkable. MRI of the brain done subsequently showed bilateral hyperintensities in the basal ganglia, midbrain, pons, thalamus and the hippocampus on the T2 weighted and Fluid attenuation inversion recovery (FLAIR) images.

On tenth day of illness, CSF tested was strongly positive for rabies antigen and immune complexes. An ELISA technique using plates coated with monoclonal antibody to rabies N protein was used for antigen detection. Neutralizing antibodies to rabies virus were detected in both serum and CSF by the standard Rapid Fluorescent Focus Inhibition Test (RFFIT).

Within 24 hours of admission, the patient became comatose and was electively ventilated. He

developed generalized hypotonia, areflexia and quadriparesis. On the third day of hospitalization, profuse hypersalivation was noted in the ventilated patient. His condition deteriorated and he succumbed to death on day 16 of hospitalisation.

## Discussion

There are two forms of human rabies, the well-known encephalitic (furious) form and the paralytic (dumb) form(1). The encephalitic form starts with fever, malaise, pharyngitis and paraesthesia at the site of the bite followed by the classical neurologic symptoms of hydrophobia, aerophobia, agitation, hypersalivation and seizures. This is followed by paralysis and coma. Death is usually due to respiratory failure. Paralytic rabies accounts for 20% of rabies. Hydrophobia and aerophobia are present only in half of these patients and it closely resembles Guillian Barre Syndrome(2).

The diagnosis of rabies encephalitis is usually unmistakable and is based on the unique clinical symptoms. Hydrophobia, which is the most characteristic and widely known feature of rabies, was not present in our case. In a study by Chabra, 5% of patients with rabies did not have hydrophobia(3). In an earlier study, Krishna and Dutta reported 24.4% of patients with neurological symptoms without hydrophobia(4). Such non-classical presentations may require laboratory investigations and neuroimaging to arrive at a diagnosis.

The presence of rabies antigen and antibodies in the CSF, as well as serum antibodies confirmed the diagnosis of rabies. There are reports of viral antigen, antibody and immune complexes coexisting and being detected in the same sample in certain CNS infections(5). Vaccination usually does not induce antibodies in the CSF(6). The normal CSF picture also helped to exclude ADEM. MRI brain showed predominant gray matter involvement, which suggested rabies; ADEM is characterized by predominant white matter involvement(2). Lack of administration of Rabies immunoglobulin in spite of a WHO Category III exposure may also account for the occurrence of rabies in our patient.

In summary, rabies can present with a wide variety of clinical symptoms, ranging from

hyperexcitation and phobic spasms to coma and flaccid paralysis. Hydrophobia alone should not be taken as a pathognomonic feature to diagnose rabies. MRI of the brain shows distinct grey matter abnormalities in rabies and is a useful tool when the diagnosis is in doubt. Rabies immunoglobulin should be administered to all cases of Category III exposures, in addition to vaccination.

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