

## Boerhaave's Syndrome (Ruptured Oesophagus) in a Case of Rabies

C T Oon

3 Mt Elizabeth #12-09, Mt Elizabeth Medical Centre, Singapore 228510  
C T Oon, MBBChir, DTM&H, FRACP

### ABSTRACT

A 46-year-old Caucasian man was admitted with a history of dog-bite in a foreign country six months previously. He presented with multisystem complaints, died suddenly soon after admission and the only significant finding at post-mortem was a ruptured oesophagus. Immunological tests confirmed rabies.

Keywords: dog bite, sudden death, ruptured oesophagus, polymerase chain reaction, rabies

### INTRODUCTION

Rabies has plagued mankind for thousands of years and has been known in ancient Egypt and China from the fifth century<sup>(1)</sup>. The disease is caused by the Rhabdovirus of the genus Lyssavirus spread from animals or bats to humans by saliva. Due to strict animal quarantine control in Singapore, which includes rabies vaccination for dogs on arrival, rabies has not been brought into Singapore by animals<sup>(2)</sup>. Rabies causes an encephalomyelitis that is usually fatal with more than 35,000 deaths occurring each year in Asia. India has 30,000 deaths yearly, China 200 deaths in 1995, mainly in the South-Eastern part and Indonesia saw 36 deaths in the same year<sup>(3)</sup>. In Vietnam, 409,131 patients were treated for possible rabies exposure in 1995 with 375 deaths. From 1985 - 1995 in Thailand, post-exposure vaccinations increased from 77,000 to 150,000 of which half were in children under 12 years of age<sup>(4,5)</sup>.

In 1998, 362 Filipinos died of rabies and 10,000 dogs are thought to be infected every year<sup>(6)</sup>. This disease also occurs in Bangladesh, Nepal, Pakistan, Sri Lanka and others in this region. Rabies symptoms appears within two months after the bite of a rabid animal in 50% of the cases. In children, a quicker development of the disease occurs compared to adults<sup>(7)</sup>. The onset has varied from four days to nineteen years, but most present within a year<sup>(8,9)</sup>. The last rabies case in Singapore was in 1953 (K T Goh personal communication).

### CASE REPORT

A 46-year-old Caucasian patient was admitted to Mt Elizabeth Hospital on 23 May 1999. He worked in Shanghai for eighteen months as a manager of a mining firm. About six months prior to the date of admission, he was bitten on the right hand by his own dog which he obtained from outside Shanghai five weeks earlier. The dog which also bit other members of his family died, but was not examined after death. Everyone had rabies vaccination except the patient and a pregnant daughter. On 16 May 1999, he felt tired and his right hand ached, had mild hyperaesthesia, which was attributed to playing darts. Over the next two days, he had a fever and sore throat which made it difficult for him to swallow and eat. His throat was found to be inflamed with aphthous ulcers and he was given Rocephin with effect. At this stage, he was extremely hyperexcitable, kept walking up and down the hospital ward and refused to sleep. The patient also developed a fear of water and drinking, had significant hypersialorrhoea and severe nausea. In the past, he had no illnesses or allergies, and he is a non smoker. He has been married for 25 years, and this was the first time he has worked in a foreign country. As his condition did not improve, his company decided to send him to Singapore for further treatment.

Upon admission, he was flushed, able to walk and had some chest tightness. Initially, he was afebrile, but his temperature went up to 38.2-C subsequently and he had episodes of sweating. His BP was recorded at 160/90 and pulse 110/min. He appeared lucid but showed some emotional lability. Clinical examination of the nervous system was essentially normal. His throat was still mildly inflamed, but heart, lungs and abdomen were normal, and he had occasional hiccups with urinary incontinence and vomiting. Hb 17.8 gms, WBC 22,300, platelet 262,000, urea 29 mg/dL, creatinine 1.1 mg/dL, potassium 2.7 mmol/L, sodium 137 mmol/L, chloride 105 mmol/L, calcium 9.2 mg/dL and glucose 140 mg/dL were recorded. Chest X-ray did not show any obvious abnormality. ECG showed normal rhythm and lumbar puncture was not done.

He was rehydrated and given Rocephin. A few hours later, while still appearing quite well, he suddenly collapsed and resuscitation was unsuccessful. At that time, it was thought that he had a massive coronary thrombosis. Post-mortem examination showed mild cardiomegaly and atheroma. The brain was normal with mild cerebral oedema and the lower oesophagus showed 3 transmural tears of 3 cms, 2 cms and 3 cms (Boerhaave's Syndrome) with severe mediastinitis.

Tests on the brain tissue showed:

1. Positive Direct Fluorescent Antibody test for intraneurone rabies antigen.
2. PCR (Polymerase Chain Reaction) positive for rabies.
3. Nucleotide sequence analysis 100% homologous with rabies sample from Anhui Province, P R China (collected in 1998 by Arthur King at the Central Veterinary Laboratory, Weybridge, U K).

The result is consistent with a dog bite in China (CDC Atlanta).

### DISCUSSION

The difficulty of diagnosing rabies in Singapore is its uncommon occurrence. The differential diagnosis to be considered are rabies phobia, drug intoxication, viral and Jap B encephalitis, tetanus and fever of unknown origin. The initial prodromal symptoms of the patient were non-specific and

moreover, he was mobile and lucid. Suggestive rabies symptoms were hydrophobia, hiccups from diaphragmatic spasms, sweating, urinary incontinence and the sensory disturbance of the bitten hand, suggesting neurological disturbance. Terminal cardiac arrest occurred in this patient which is the usual ending in one third of cases<sup>(10)</sup>. The suspicion of this disease initially is essentially clinical. Antibody test is not useful at the beginning as results are only positive after the first week or later<sup>(11)</sup>. The patient died in the first week and his test was negative (WHO Rabies Research Centre, Bangkok). There is no specific medication for rabies and antiviral treatment is ineffective<sup>(12)</sup>. Treatment is symptomatic and supportive, but death invariably occurs. Survivors are usually those who contracted the disease but had been vaccinated previously, they however, had residual deficits<sup>(13)</sup>. The ruptured oesophagus (Boerhaave's syndrome) was probably caused by vomiting that was not apparent on admission. The chest X-ray was normal and no surgical emphysema or pleural effusion was present. The high white cell count was partly contributed by the mediastinitis.

Transplacental rabies infection is known in animals, but not in humans. Many exposed pregnant women were given post exposure treatment (PET) and have had healthy newborns<sup>(14)</sup>. As the pregnant woman bitten by the same dog could be incubating the disease, vaccination and rabies immunoglobulin (RIG) was given one month after delivery of the child and six months later. No treatment was indicated for this newborn and as far as is known, both are well.

Human to human transmission via infected secretion has never been documented virologically nor has viraemia been detected<sup>(15)</sup>. Many infections have occurred from grafting infected cornea<sup>(16)</sup>, and two non-laboratory confirmed cases were described from a human bite and kiss in Ethiopia<sup>(17)</sup>. Transmission of rabies virus by inhalation and causing disease occurred in two persons in infested caves and from unrecognised bat bites which are often hardly visible<sup>(18,19)</sup>. Exposure is defined as penetration of the skin by teeth, contact with contaminated saliva on mucus membrane, cornea, conjunctiva, open wounds, scratches on the skin and needle stick injuries. Nursing and medical staff in contact with the patient are given PET for reassurance against this deadly infection. Besides it is also good public health practice to have nurses immunised against this disease in preparation for future cases. The Human Diploid Cell Vaccine (HDCV) was used and no undue reactions occurred except for minor muscle ache and itching over the injected site.

## CONCLUSIONS

Singapore has many foreign nationals from countries with rabies working here. Should they die suddenly with encephalitis, have abnormal neurological signs or if they give a history of a dog bite, (less commonly a cat bite) investigations should be done to exclude rabies.

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