

Clinical Case Report

Rabies in a blind patient: Confusion after corneal transplantation

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ABSTRACT

Two blind persons received corneal transplants from a single donor who showed no signs of rabies before he died. One of the recipients, a young girl, died 16 days later of rabies and the other recipient survived. We discuss the possible mode of transmission of rabies to the first recipient and the management of the second recipient.

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INTRODUCTION

Rabies is usually transmitted following exposure to the saliva of an infected animal through bites or licks on abraded skin. Organ transplantation, including corneal transplantation, has been shown to transmit rabies from an infected donor to the recipient.^{1,2} If the donor died of an illness which was clinically suspected to be rabies, it is easy to establish the mode of transmission. However, if the cause of the donor's death is not clear, it can be difficult to establish transmission. We present a case of rabies in the recipient of a corneal transplant in whom the source of the rabies infection was uncertain.

THE CASE

A 10-year-old girl was admitted to the ophthalmology department of a hospital in Bengaluru in early 2011. She was diagnosed to have bilateral congenital hereditary endothelial dystrophy. Since a donor cornea had been obtained, she underwent penetrating keratoplasty of the right eye in early 2011. The postoperative period was uneventful and the graft took well.

She remained well for the next 2 weeks and then developed headache which was not associated with fever or convulsions. She was referred to the paediatrics department of the same hospital. On examination, she was incoherent, aggressive and irritable with excessive salivation. She had delusions and subsequently developed hydrophobia and aerophobia. She was unable to stand due to hypotonia. There was no neck rigidity. Bilateral plantar

reflexes were flexor. Examination of the respiratory system, the cardiovascular system and the abdomen was normal.

She died a few days later—16 days after keratoplasty. The paediatrician made a diagnosis of rabies and attributed it to the corneal transplant. An autopsy was done and the whole-brain specimen was sent to the department of neurovirology, WHO Collaborating Centre for Rabies, National Institute of Mental Health and Neurosciences (NIMHANS), Bengaluru for rabies virus/antigen detection. The test was strongly positive by direct rapid immunohistochemical test (dRIT; Fig. 1)³ and fluorescent antibody test (FAT),⁴ confirming the diagnosis of rabies.

Source of the corneal graft

A few days before the keratoplasty, bilateral corneal grafts (following enucleation) were obtained from a 26-year-old man who had died due to a suspected myocardial infarction at a district hospital 200 km from Bengaluru. The patient had complained of chest pain. At the time of admission, there were no symptoms of rabies and a history of dog bite had not been enquired about. The diagnosis of myocardial infarction was made on the basis of the history of chest pain and no investigations or autopsy had been done. These details were confirmed telephonically from the treating physician, the patient's relatives and the ophthalmologists who removed the eye balls. The corneal grafts were transferred to an eye bank and from there they were shifted a day later to the hospital in Bengaluru.

As the first recipient developed rabies following corneal transplantation, the corneal graft was suspected to be the source of infection. However, the corneal graft came from a person who had not died of any neurological disease and the cornea was obtained in a district hospital by a qualified ophthalmologist. The recipient could have died of rabies following exposure to a rabid animal in the past. Being blind, she may not have noticed exposure to a rabid animal. However, the parents did not recall her having

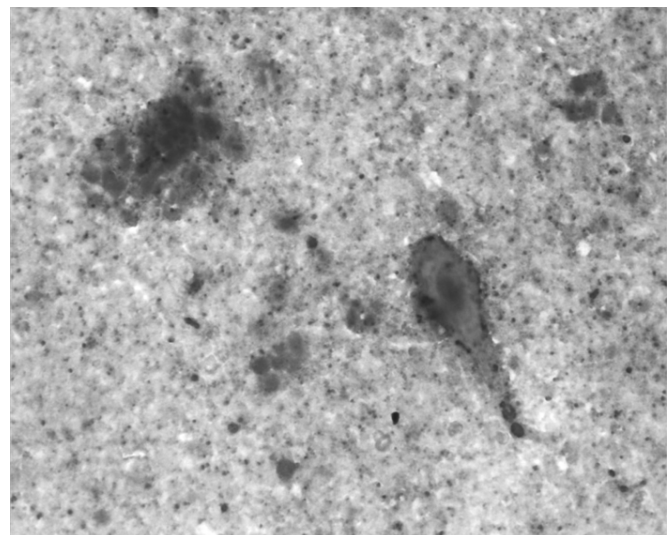


FIG 1. A photomicrograph of the brain of the patient tested by dRIT. Note the dark brown particles present in the neurons and outside (x400)

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been bitten by a dog or by any other animal. Therefore, the source of infection in this recipient remained unclear.

The second recipient

The recipient of the other cornea from the same donor was a 55-year-old woman who was admitted to the same hospital in Bengaluru in early 2011 and underwent corneal transplant in the right eye. As the other recipient had died of rabies following corneal transplantation, this recipient was given rabies post-exposure prophylaxis with 1 ml of human rabies immunoglobulin [HRIG] (Kamrab) as eye drops in each eye. She was also administered Purified Chicken Embryo Cell (PCEC) vaccine (Rabipur, double dose) by the intramuscular route with subsequent doses given on days 3, 7, 14 and 28. On days 7 and 14 post-vaccination, 4 ml of blood was taken for estimation of rabies virus neutralizing antibody (RVNA) titre. RVNA titres estimated by the rapid fluorescent focus inhibition test (RFFIT)⁵ were more than 7.5 i.u. per ml, indicating that she had developed adequate protection against rabies. She was kept under observation in the hospital for 28 days. When last seen in the middle of 2011, she was alive and healthy.

On the same day that the first recipient died, 35 of the hospital staff along with the patient's attendants were administered rabies prophylaxis using the updated Thai Red Cross (TRC) schedule.

DISCUSSION

Human-to-human transmission has occurred among 8 recipients of transplanted corneas.⁶ Each of the donors had died of an illness compatible with or proven to be rabies. These 8 cases have been reported from five countries—two each from Thailand, India and Iran and one each from the USA and France. However, a history of exposure to rabies virus could not be elicited in the majority of donors from whom the corneas were transplanted. In the USA, four recipients have died of rabies following non-corneal organ

transplantation.² In Germany three of six organ recipients had died of rabies.⁷

Rabies developed in our first recipient following corneal transplantation. However, the source of infection remains unclear as the donor died without any clinical features of rabies and an autopsy was not done. The second recipient who received the other cornea from the same donor was given rabies post-exposure prophylaxis and is still alive. India being a rabies endemic country, such situations may be encountered, which warrant a thorough investigation and careful management.

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