A Case of Rabies

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Rabies encephalomyelitis is considered rare in Great Britain, but this is the second human case to occur in this country within six months. The patient, like the one reported by Ghosh (1964), was an immigrant from the East. As air travel increases such cases are likely to be met with more often than in the past and an awareness of this possible diagnosis is important, since the initial picture may be confusing and overshadowed by behavioural disturbances of a histrionic type.

CASE REPORT

The patient was a 41-year-old Indonesian woman who had arrived in England three months previously. Four days before admission her right hand began to ache and later that day when carrying a bag the right arm felt stiff and ached. The pain went up to the axilla, became shooting, and the arm felt tender. There were pins and needles in the hand and forearm, the skin felt prickly, and she scratched it. These sensory symptoms became worse and by evening she was drowsy and nauseated. She was better the next day, but by the following day there was an irritation of both sides of the face and of the skin at the back of the neck and right shoulder which she kept rubbing. On the morning before admission the right hand felt weak and the shooting pains recurred. Pains also began to shoot down the inner side of the left arm and there were pins and needles in the left hand. That evening when she tried to drink she became frightened and choked.

On admission she was fully alert and orientated, but very upset and tearful, and kept slapping her hand on the bedclothes in a peculiar, histrionic way. In conversation, however, she readily relaxed and talked naturally and quietly but when touched she became fearful. This was particularly noticeable when the corneal reflexes were elicited. When the cornea was touched she jerked back her head in an exaggerated manner, cried out, and trembled all over. Even more dramatic was the state of terror induced by offering a drink. This caused her to shake, perspire, hold up her hands in defence, and call out "Don't, don't" or "No, no." As the cup reached her lips she would throw back her head, cry out "I can't, I can't," and flop back in bed breathing heavily. With encouragement, however, she could be persuaded to take a few gulps of water and swallow it.

Apart from this odd psychiatric state there were no signs other than in the arms, where there was mild diffuse weakness on the right with minimal weakness of the left finger extensors and absent tendon jerks in both arms. There was severe impairment of all forms of sensation over the whole of the right arm except for the inner aspect, and minimal sensory loss in the left hand. Loss on the right extended into the neck and trunk from C3 to T4 in a patchy fashion. General examination was entirely negative; temperature, pulse, and blood-pressure were all normal.

The pains in the arms, strange demeanour, and exaggerated voluntary movements, together with the apprehension at the sight of water, all suggested hydrophobia and both patient and husband were questioned about dog bites. This was denied, though it emerged that the patient was a dog-lover and had brought three spaniels from Indonesia. The dogs were in quarantine, and six weeks previously one of the dogs had died. This was immediately followed up, but inquiry revealed that necropsy had shown that the dog had died of liver disease. Since rabies is so uncommon, and no clear history of infection was forthcoming, a diagnosis of viral radiculopathy with severe tension state was made.

Except for the fear of swallowing, which was for solid food as well as liquids, the patient remained fairly well until the following day, when her temperature rose to 100° F. (37.8° C.). She became restless and said that both sides of her face felt swollen, but no sensory loss could be demonstrated over the face, and the other physical signs were unchanged. The next day the temperature was 104° F.

(40° C.) and she had pain in the face similar to that in the arms. She also began to complain that she could not breathe properly, and when examined would sit up or roll away from the examiner like a startled animal. So bizarre was the patient's behaviour at this time that a visiting physician who examined her and could find nothing wrong in the chest considered her symptoms hysterical. She was sedated with Largactil and had a good night, though the temperature remained at 103° F. (39.4° C.). Next morning her breathing was worse and she became cyanosed owing to repeated respiratory muscle spasm. The difficulty in breathing was now obviously not hysterical and this coupled with the hyperpyrexia made the diagnosis of rabies certain. Soon afterwards she suddenly vomited twice, collapsed, and died.

At necropsy Professor W. Blackwood found diffuse encephalomyelitis particularly marked in the brain stem and cervical cord, where there was congestion, loss of cells, and perivascular cuffing, but no Negri bodies. Examination of the Ammon's horn also failed to reveal inclusions. Intracerebral inoculation of 10% suspension of brain tissue from the hippocampus and cervical cord into litters of newborn mice produced signs of encephalitis after five days. No specific cytoplasmic inclusions of rabies, however, were found in stained sections. The same material was inoculated into groups of six adult mice by the intracerebral route. In this case signs of encephalitis with tremors, ataxia, convulsions, and spastic paralyses appeared on the nineteenth day after inoculation with material from the hippocampus and on the twentieth day with material from the cervical cord. By the twenty-third day only three out of the six of the inoculated animals had sickened. Sections from the brain showed a diffuse encephalomyelitis with neuronal necrosis and perivascular cuffing; numerous intracytoplasmic eosinophilic inclusions, typical of Negri bodies, were present. The same material was kindly examined by Dr. A. D. Macrae at Colindale, who reported that numerous Negri bodies were present in smears stained by Seller's stain.

The unusual feature in this case was the long incubation period of rabies virus. Normally, inoculated animals sicken between six and eight days after inoculation, but according to Johnson (1956) some strains of street-rabies virus are characterized by an incubation period of 15 to 20 days and some as long as 65 days. A week before the virus was identified the husband disclosed that shortly before leaving Indonesia his wife had been bitten by a moribund cat which had been savaged by rabid dogs. Subsequently a further examination of the human material revealed a few intracytoplasmic inclusions in Purkinje cells in the cerebellum.

COMMENT

This case is reported, not just because it is a case of rabies, but because it illustrates some of the difficulties in diagnosis. The clinical picture can be misleading, and pathological confirmation may be delayed. Clinical diagnosis is therefore important, particularly if there is a risk to other humans (for the modern prophylactic treatment of rabies see Second Report of World Health Organization's Expert Committee on Rabies, 1954), and the disease should be borne in mind in persons entering the United Kingdom from countries where rabies is endemic.

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ALAN RIDLEY, M.D., PH.D., M.R.C.P.,
Resident Medical Officer,
The National Hospital, Queen Square, London.

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