

Acquired immune deficiency syndrome without the recognized risk factors

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Summary: We report two cases of acquired immune deficiency syndrome (AIDS), apparently without the usual exposure factors, in whom a temporal association was detected after detailed epidemiological investigation.

The index case, a 45 year old housewife, had provided terminal home-nursing care for a 33 year old African man, who died from an undiagnosed encephalitis. At that time she had fissures of the skin of both her hands. Review of post-mortem pathology specimens of the African man allowed a retrospective diagnosis of AIDS with cerebral toxoplasmosis to be made.

The type of home-nursing care given by the index case was quite different from that normally provided by health care workers with the training and facilities to prevent the spread of infection.

Introduction

Whilst the modes of transmission of the human immunodeficiency virus (HIV) are now well established, it is important to retain a clinical awareness of the possibility of this infection in patients without apparent exposure to the recognized risk factors. We report such a case where clinical diagnosis was delayed because of the lack of obvious transmission source.

Case report

The index case, a 45 year old housewife, was admitted with a 3-week history of a disseminated herpes zoster infection together with both oral and vaginal candidiasis. Three years before she had been investigated for a glandular fever-like illness but despite bone marrow and lymph node biopsies, no diagnosis was made and she continued to complain of persistent symptoms of ill health, weight loss and cervical lymphadenopathy until presentation in 1985.

On admission the patient was unwell with a fever, axillary and cervical lymphadenopathy, modest hepatomegaly, and an extensive toxic erythema. Initial investigations were: haemoglobin 9.9 g/dl, total white cell count $6.9 \times 10^9/l$ with 86% granulocytes and 11% lymphocytes, platelets $250 \times 10^9/l$, and an ESR of 60 mm in the first hour. Liver function was slightly deranged but blood urea and electrolytes were normal.

A chest X-ray at this time was unremarkable and a complement fixing antibody titre of >2048 to varicella-zoster virus was consistent with her recent infection.

A working diagnosis of immunosuppression secondary to an underlying neoplasm, probably a lymphoma, was made. Further investigations including abdominal computed tomographic (CT) scan and nuclear isotope liver scan were normal. A number of biopsies including bone marrow, lymph node, jejunal mucosa, and liver showed either non-specific or reactive changes only.

The possibility of this being associated with a HIV infection was considered, but the patient had had no history of drug abuse or blood transfusions and her last sexual intercourse had been seven years previously.

The patient's clinical condition gradually deteriorated with increasing breathlessness and a non-productive cough. Chest X-ray showed diffuse interstitial shadowing which progressed rapidly over the next few days. *Pneumocystis carinii* pneumonia was felt to be the most likely diagnosis and this was confirmed by histological examination of transbronchial biopsies.

The patient responded dramatically to high dose oral co-trimoxazole, being afebrile within 36 hours, and symptom free within the week. Subsequently the patient was shown to be HIV antibody positive conferring a diagnosis of the acquired immune deficiency syndrome (AIDS).

In depth interviewing failed to elicit a history of drug abuse, contact with blood products, or any recent sexual intercourse; however she recalled looking after a sick neighbour in September 1982 who subsequently died from an undiagnosed encephalitic illness.

The patient remained well for 6 months but then gradually deteriorated and died from *Pneumocystis carinii* pneumonia 9 months after discharge from hospital.

Contact case

A 33 year old Ghanaian man was admitted to hospital in October 1982 having collapsed at home. There was a history of several weeks headache with general aches and pains and then a rapid onset of drowsiness. He was lymphopaenic with a total lymphocyte count of $0.9 \times 10^9/l$. A CT head scan demonstrated a mass of mixed attenuation in the left parietal region with further scattered small masses. His condition deteriorated rapidly and he died soon after admission from a presumptive encephalitic illness. In view of the history of the nursing contact obtained from the index case, a review of post-mortem histology allowed a retrospective diagnosis of cerebral toxoplasmosis to be made. Analysis of the stored serum and cerebrospinal fluid samples showed them both to contain HIV antibodies.

Epidemiological investigation

The index case was interviewed on three separate occasions by a medical epidemiologist from the PHLS Communicable Disease Surveillance Centre. A standard protocol was followed, consisting of an initial unstructured interview followed by the completion of a structured questionnaire administered by the investigator. An independent witness was later interviewed using similar techniques. Basic demographic details and personal characteristics of the index case were sought. Previous medical, social, sexual and travel histories were recorded, including potential exposures to recognized risk factors for HIV infection.

The index case denied previous exposure to parenteral drug abuse and contact with bisexuals. She had never had a blood transfusion and had received no parenteral medication in the five years prior to this interview. She had never travelled overseas.

In October 1982 she had nursed for several days a family friend, the contact case, who had been resident in the United Kingdom for ten years. At this time he was suffering from an undiagnosed illness, and was incapable of looking after himself. He became progressively more drowsy with vomiting, and was incontinent of urine and faeces. The nursing care given

by the index case involved exposing her hands to his body fluids including saliva and urine, although these were not obviously blood stained. At the time of this exposure she had some recent small cuts on the skin of her hands, as well as a primary irritant hand dermatitis which was exacerbated by regular hand washing of the contact case's bed linen which was soiled with urine and faeces. Approximately eight weeks later she developed an illness which was diagnosed as 'glandular fever'.

The index case strongly denied any sexual contact with the Ghanaian man, and this denial was corroborated by a female friend of the contact case, who had lived with him until eight months before he died. To the knowledge of this woman the contact case was not bisexual and did not abuse parenteral drugs. He had travelled to Hamburg in 1982 (the year of his death) for a two week holiday, but had not returned to Africa in the previous ten years. However, several years prior to his death, the contact case had worked in the entertainment industry, and it is possible that he may have been in contact with drug abusers at that time.

Discussion

These two cases of AIDS have been previously briefly reported.¹ The index case demonstrates several important clinical lessons.

Although the possibility of AIDS was considered at an early stage there was a certain degree of reluctance to perform the HIV antibody test as the patient did not fit into an 'at risk' group. As the reservoir of HIV infection in the community rises, so atypical presentations of HIV infection will increase. It is therefore important that the clinical awareness of potential HIV infection should be heightened and that the at risk group for HIV infection should perhaps include patients who have unexplained immunodeficiency together with a pyrexia of unknown origin even if there is no obvious virus transmission source.

Glandular fever type illnesses have been described in association with seroconversion to HIV infection² and it is of interest that she suffered from this type of illness documented at around 8 weeks after nursing the contact case. This was similar to the experience of two nurses who sero-converted after needle-stick injury sustained whilst caring for patients suffering with AIDS,^{3,4} and is highly suggestive that the HIV infection in the index case was contracted at the time her unprotected hands were repeatedly exposed to urine and faeces whilst nursing the contact case.

For the purposes of epidemiological surveillance there is only one documented case of accidental injury in a health care worker which strictly fulfills the criteria for occupationally acquired HIV infection,

although two further possible cases have occurred in the United States.⁵ In the United Kingdom no seroconversion has occurred in a group of 101 health care workers with accidental parenteral or mucosal exposure to blood or body fluids contaminated with HIV.⁶

References

1. Grint, P. & McEvoy, M. Two associated cases of the acquired immune deficiency syndrome (AIDS). *Commun Dis Rep* 1985, **42**: 4.
2. Cooper, D.A., Gold, J., Maclean, P. *et al.* Acute AIDS retrovirus infection: Definition of a clinical illness associated with seroconversion. *Lancet* 1985, **i**: 537–540.
3. Anonymous. Needlestick transmission of HTLV III from a patient infected in Africa. *Lancet* 1984, **ii**: 1376–1377.
4. Stricof, R.L. & Morse, D.L. HTLV III/LAV seroconversion following a deep intramuscular needlestick injury. *N Engl J Med* 1986, **314**: 1115.
5. Center for Disease Control. Update: Evaluation of HTLV III/LAV infection in health care personnel – United States. *MMWR* 1985, **34**: 575–583.
6. P.H.L.S. Surveillance of health care workers with accidental parenteral or mucosal exposure to blood or body fluids of patients infected with HTLV III. (unpublished data).

While it is extremely important that all possible exposures are followed up, the current available evidence suggests that if simple sensible precautions are followed, then the risk to health care personnel of contracting this infection is very small.