

Fatal Haemorrhagic Chickenpox Complicating Nephrotic Syndrome

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Summary

We report a 14 year old Indian-Muslim girl who developed a fulminant, disseminated and fatal varicella infection while receiving steroids for nephrotic syndrome. The terminal phase of her illness was complicated by a bleeding dyscrasia and circulatory collapse.

Varicella infection in healthy children is a benign disease. However in neonates and immunosuppressed patients it may be severe and often fatal. There are many reports of fatalities occurring in cancer patients receiving chemotherapy, patients on immunosuppressives for asthma, haemolytic anaemia, rheumatic fever, and renal and bone marrow transplantation.

Patients with nephrotic syndrome receiving cyclophosphamide treatment are at particular risk of developing severe chickenpox infection. To our knowledge, there has been only one report of fatal chickenpox infection in a child who received steroids for nephrotic syndrome¹.

We report here a case of fatal haemorrhagic chickenpox complicating nephrotic syndrome.

Key words: Chickenpox, Nephrotic syndrome

Case Report

KMH was a 14 year old Indian-Muslim girl who presented at a district hospital with nephrotic syndrome in June 1992. She was started on prednisolone. In early September, she was readmitted to the same hospital with fever, low back pain and a generalised vesicular rash. Three weeks prior to admission, she was exposed to chickenpox when two of her siblings developed the disease. Soon after admission, her general condition deteriorated and she was referred to our unit for further management.

On arrival at our unit, her general condition was poor. She was febrile, drowsy and toxic looking. There was a generalized vesicular rash which was more dense over the face and trunk but sparing the palms and soles (Figure 1). Some of the vesicles contained haemorrhagic fluid. She was also bleeding from multiple ruptured vesicles in her mouth and extensive crusting of the mucosa was seen. Her face was swollen and she had pitting edema up to her sacrum. Her blood pressure was 130/80 mm Hg supine and her pulse rate was 106 per minute. Examination of the other systems were normal.

Preliminary investigations showed that her haemoglobin was 13.5g/l, total white cell count $31.0 \times 10^9/l$ and platelet count 43.0×10^9 . The blood film showed a leuco-erythroblastic picture with

a left shift of the neutrophils. Renal profile, liver profile and chest radiography were normal. Urinalysis showed 4+ proteinuria. Her blood culture did not grow any pathogen. The diagnosis of chickenpox was confirmed by immunofluorescent examination of a skin biopsy of a vesicle. She was commenced on intravenous gammaglobulin, acyclovir and hydrocortisone. She also received a combination of cloxacillin and ceftazidime. However, despite these measures, her condition deteriorated over the next 24 hours. She continued to bleed from her mouth ulcers as well as some of the ruptured vesicles on her trunk and face. She also developed spontaneous bruising over her left wrist and at venipuncture sites. Towards the terminal phase of her illness, she became progressively hypotensive and succumbed to her illness about 40 hours after admission despite resuscitative measures.

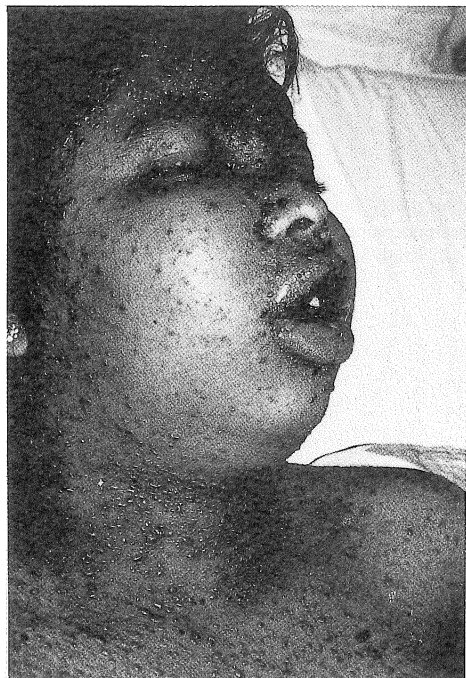


Fig. 1: Extensive vesicular rash, mouth ulceration and facial oedema.

Discussion

Chickenpox is a benign illness in immunocompetent patients and complications and fatalities are rare. However in immunocompromised patients it can be severe and often fatal.

The course of illness in this patient was very brief, terminating in circulatory collapse and disseminated intravascular coagulopathy as suggested by the bleeding diathesis and thrombocytopenia. Unfortunately we were not able to confirm this with laboratory tests because of her rapid demise over the weekend. A review of the literature revealed that disseminated intravascular coagulopathy can be the cause of death in immunocompromised patients with chickenpox infection². These patients have been classified to have malignant chickenpox with purpura. The disease has a high mortality rate and death usually occurs within 48 hours of the onset of bleeding. Death from acute adrenal insufficiency was unlikely in this patient because she was receiving hydrocortisone. There are two reasons for the fatal course of the infection in this patient. Firstly, her steroid therapy has reduced her resistance to infection. Secondly, her heavy proteinuria may have caused significant loss of immunoglobulins and other important serum proteins normally involved in combating infections thus rendering her defenceless against the varicella virus. It is recommended that such high risk patients be given varicella zoster immune globulin upon exposure to chickenpox as this measure has been shown to prevent or modify the course of

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the infection³. Recently, polyvalent immune serum globulin has been shown to confer antibody levels comparable to those achieved with specific varicella zoster immune globulin. These preparations may thus be suitable alternatives to varicella zoster immune globulin if the latter is not available. Acyclovir may be life saving in such circumstances but to be effective, it must be given as early as possible. Unfortunately, despite receiving both agents, the patient died soon after transfer to our unit.

Management of chickenpox infection in immunocompromised patients should be considered as a matter of great urgency. Such patients must avoid contact with patients with chickenpox and if that fails, early administration of immune globulin and acyclovir may be critical. Vaccination may also be considered in such circumstances.

References

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