# Hemorrhagic Varicella in Chronic Liver Disease

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## ABSTRACT

Hemorrhagic varicella is a serious complication of a relatively benign disorder and usually occurs in immunocompromised persons and those on immunosuppressive therapy. To the best of our knowledge, this is the first case report of hemorrhagic varicella associated with chronic liver disease in Indian literature. Our encounter with this case highlights that rare hemorrhagic varicella can also present in cases of chronic liver disease. Prompt diagnosis and treatment with acyclovir leads to complete recovery.

**Key words:** Acyclovir, Chronic liver disease, Hemorrhagic varicella, Immunocompromised

## INTRODUCTION

Primary infection by Varicella-zoster virus (VZV) results in varicella (chickenpox) which, a common and extremely contagious acute infection that occurs in epidemics among preschool and school-aged children, is characterized by generalized vesicular rash.[1] The typical clinical presentations of varicella are distinctive and readily recognized by most experienced clinicians. However, atypical clinical presentations and uncommon complications of this disease can pose diagnostic and therapeutic challenges.[1] There may be development of several unusual complications besides the usual complications of chickenpox, and these may be Guillain-Barré syndrome, acute transverse myelitis, acute disseminated encephalomyelitis, optic neuritis, acute pancreatitis, acute acalculous cholecystitis, immune thrombocytopenia, disseminated intravascular coagulation (DIC), hemorrhagic rashes, hemorrhagic stroke (due to vasculopathy), Steven-Johnson syndrome, myocarditis/pericarditis, acute nephritic/nephrotic syndrome and rarely orchitis, synovitis, tympanic membrane rupture or cerebral thrombophlebitis.[2] Hemorrhagic varicella is a serious complication of a relatively benign disorder and usually occurs in immunocompromised persons and those on immunosuppressive therapy.

After extensive literature search we did not find any case of hemorrhagic varicella associated with chronic liver disease. Probably our case is the first reported association of hemorrhagic varicella with chronic liver disease in Indian literature.

## CASE REPORT

A 12-year-old boy, follow-up case of chronic liver disease, was admitted in our hospital with 1-week history of fever, malaise and 5 days history of generalized discrete vesiculopustular rash over the entire body and oral ulcers. His palms and soles were also involved. Since the last 3 days, the rash became hemorrhagic with frank bleeding from several areas especially from the lesions on the face and also had malena. Since then, the patient became anorexic and weak. There was family history of similar illness in two younger siblings but of mild in nature and rashes were subsided in them without any treatment. He had history of jaundice off and on and abdominal distension since last 1 year. He was hospitalized two times for the complaint of jaundice and abdominal distension in the past 1 year. The patient belonged to a poor social background. On examination, the patient was looking toxic and he was conscious and oriented. He was febrile

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(temperature: 38.5°C) and had severe pallor. There were several hemorrhagic ulcers in his mouth and over the lips, multiple large hemorrhagic rashes over the face [Figure 1], with frank bleeding from a few lesions and multiple discrete hemorrhagic pustules over the entire body [Figures 2 and 3]. On per abdominal examination spleen was palpable 8 cm below costal margin and liver was palpable 3cm below costal margin. On investigations, hemoglobin level was 4.7 g/dl, total leukocyte count was $4.3 \times 10^9$/l and platelets were $29 \times 10^9$/l and ESR was 65 mm fall. Serum bilirubin was 3.0 mg/dl, serum albumin was 2.3 g/dl with SGOT 154 and SGPT 88. Prothrombin and thromboplastin time were also prolonged. Level of serum urea was 31 mg/dl and creatinine was 0.98 mg/dl. The patient was HBsAg negative and antibodies to hepatitis-C were absent. ELISA test for HIV was negative. Abdominal ultrasonography revealed hepatosplenomegaly with coarse echotexture of liver with portal vein dilatation. Tzanck smear revealed multinucleated giant cells. Anti-varicella IgM antibodies were positive. We did not perform liver biopsy because patient's general condition was low and he had thrombocytopenia too. On the basis of clinical condition and laboratory parameters the diagnosis of chronic liver disease with hemorrhagic chickenpox was made. During the hospital stay the patient has also developed pneumonia on day 2 of admission. We have given aggressive supportive treatment as well as intravenous acyclovir 10 mg/kg 8 hourly, intravenous antibiotics, including cefotaxime 75 mg/kg 12 hourly and vancomycin 15 mg/kg 8 hourly to him for 10 days and were transfused packed cell volume along with platelet concentrate and fresh frozen plasma. Fortunately the patient got improved on day 7 and we discharged him and referred to higher center on day 10 for further management of his chronic liver disease. Written consent was obtained from the patient's father for the publication of case report along with photographs.

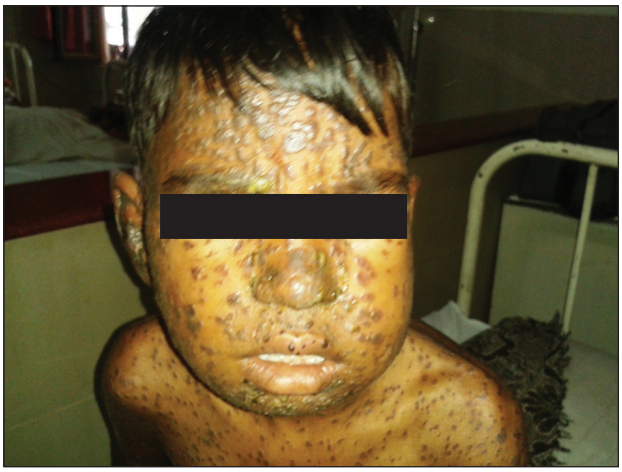

**Figure 1:** Hemorrhagic rashes over the face

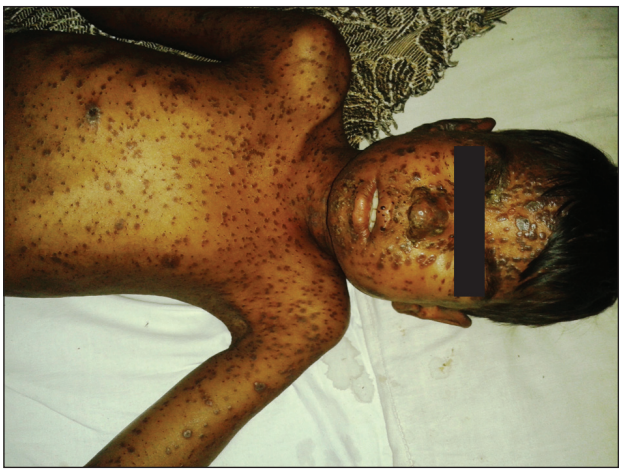

**Figure 2:** Multiple hemorrhagic rashes over the chest

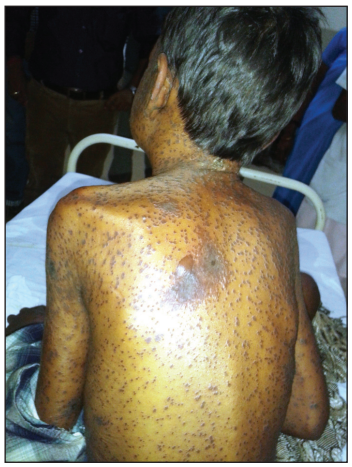

**Figure 3:** Multiple hemorrhagic rashes and large blister over the back

## DISCUSSION

An estimated 60 million cases of Varicella occurs worldwide each year and in India an incidence of 4.7 lakhs has been noted making it an almost inevitable disease of childhood. Complications of varicella are generally mild, but it can present in severe forms especially in immunocompromised children and adults.[2] Immunocompromised persons who get varicella are at risk of developing visceral dissemination (VZV infection of internal organs) leading to pneumonia, hepatitis, encephalitis, and disseminated intravascular coagulopathy.[3] They can have an atypical varicella rash with more lesions, and they can be sick longer than immunocompetent persons who get varicella. The lesions may continue to erupt for as long as 10 days, may appear on the palms and soles, and may be hemorrhagic.[3]

Hemorrhagic complications are seems to be common in the immunocompromised or immunosuppressed populations. There have been case reports in which fatal hemorrhagic chickenpox occurred in steroid dependent asthmatic patients and in nephrotic syndrome.[4,5] In cases of chronic liver disease there is impairment in cellular as well as in humoral immunity, probably this is the reason of hemorrhagic varicella in our case.[6,7]

To the best of our knowledge, this is the first case report of hemorrhagic varicella associated with chronic liver disease in Indian literature.

The predominance of uncomplicated cases in children tends to overshadow the morbidity and mortality associated with severe cases.[8] Complications of varicella requiring hospitalization in children are becoming more frequent than previously thought.[9] Even though a lethal outcome remains a rare occurrence, it may be of relevant concern when considering the overall incidence of chickenpox in the general population. Acyclovir may be life saving in such circumstances but to be effective, it must be given as early as possible. Intravenous foscarnet may be needed for cases that do not respond to acyclovir, however, our patient responded well to intravenous acyclovir. 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. 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 the infection,[10] however, our patient responded well to intravenous acyclovir alone.

The present case serves to emphasize the importance of including varicella vaccine in the childhood immunization program to help decrease varicella-related life-threatening complications.

## CONCLUSION

Our encounter with this case highlights that rare hemorrhagic varicella can also present in cases of chronic liver disease. Prompt diagnosis and treatment with acyclovir leads to complete recovery.

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