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### **Case Report**

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# Aplastic crisis in a child with hereditary spherocytosis with a strong family history resolving with supportive care

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#### **ABSTRACT**

Hereditary spherocytosis (HS) is a familial hemolytic disorder with marked heterogeneity. Clinical features range from asymptomatic to fulminant hemolytic anaemia. The Human Parvovirus Virus B19 induced aplastic crisis can unmask several hereditary hematological disorders that have been normally compensated. Among these conditions, hereditary spherocytosis has been extensively reported. The clinical importance of this report is that in the case of an abrupt onset of unexplained severe anemia and jaundice, one should consider underlying hemolytic anemias mostly hereditary spherocytosis complicated by HPV B19 aplastic crisis. We herein report a 13-year-old boy who is a known case of hereditary spherocytosis presented with severe pallor, jaundice and pancytopenia after a febrile illness.

**Keywords:** Aplastic crisis, Hereditary spherocytosis, Severe pallor

#### **INTRODUCTION**

Human parvovirus B19 is a well-known cause of aplastic crisis in patients with Hereditary Spherocytosis.<sup>1,2</sup> Human parvovirus (HPV) B19, being first discovered and introduced in 1975, is a non-enveloped single-stranded DNA virus from the Parvoviridae family.3 The virus is transmitted by respiratory droplets and the prevalence is estimated to be high since most of the individuals are infected by the age of 15.4 The virus has a predilection for infecting the erythroid progenitor cells of the bone marrow resulting in their lysis and aplastic anemia.<sup>5</sup> The clinical syndrome associated with HPV B19 strongly depends on the host; for instance those suffering from hemolytic disorders, including sickle cell disease, hereditary spherocytosis (HS), autoimmune hemolysis, thalassemias, and paroxysmal nocturnal hemoglobinuria (PNH) are susceptible to aplastic crisis. 6 HPV-B19 infects erythroid progenitor cells and inhibits erythropoiesis, leading acute erythroblastopenia to reticulocytopenia.<sup>7</sup> Thus the patient presents with signs

and symptoms of abrupt onset severe anemia. Fever, nonspecific flu-like symptoms, and abdominal symptoms such as nausea or vomiting, abdominal pain, and diarrhoea may occur in patients with HPVB19-induced aplastic crisis.<sup>8</sup>

It is self-limiting and generally requires only supportive measures and blood transfusions to bridge the gap between marrow suppression and recovery. Rarely, patients will need immunoglobulin treatment, which is dramatically effective in reversing the aplasia. The diagnosis of transient aplastic crisis due to parvovirus B19 is often presumptive, based on a falling hemoglobin value and a low reticulocyte count in a patient with a hemolytic anemia. Specific DNA probes allow definitive diagnosis by PCR since the viremia is robust. A rising IgM antibody to the virus is another means of diagnosing parvovirus B19 infection.

We herein, report a known case of hereditary spherocytosis with a history of fever and vomiting since 4

days with severe anemia and pancytopenia on peripheral blood.

#### **CASE REPORT**

A 13-year-old boy, who is a known case of hereditary spherocytosis, with a sick look was presented with fever, vomiting and fatigue since 4 days. On physical examination, he had severe pallor, pulse rate: 96 bpm, respiratory rate: 24/min. The spleen was palpable about 6 cm below the left costal margin and the liver palpable 3 cm below the right costal margin. On auscultation, a systolic flow murmur is heard.

There is a past history of jaundice on day 3 of life and was treated with exchange transfusion followed by phototherapy. The child had uneventful course until 2 years of life when he was diagnosed to have hereditary spherocytosis at a higher centre when he presented with complaints of jaundice, splenomegaly and was on folic acid supplementation since then. There is a positive family history where his father and grandfather are diagnosed to have hereditary spherocytosis.



Figure 1: Known case of hereditary spherocytosis who presented with aplastic crisis.

Owing to the abrupt symptoms of fever, vomiting, severe pallor, splenomegaly with known history of hereditary spherocytosis we further investigated for aplastic crisis which is occurs in many patients due to infection with parvovirus. Our workup showed hemoglobin of 4.2 gm%, RBCs: 1.35 million/cumm, WBCs: 2200 cells/cumm, platelets: 1 lac/cumm and many spherocytes on peripheral smear examination, reticulocyte count: 0.1%. Total bilirubin: 4.1 mg/dl and direct bilirubin: 0.4 mg/dl. Because of pancytopenia accompanied with a marked decrease in reticulocytes as well as a history of HS, it was natural to believe that the patient in the present case was suffering from aplastic crisis due to viral infection; therefore, we did not perform bone marrow aspiration. The boy received packed red blood transfusion,

dobutamine infusion and other supportive care and was recovered in 7 days. Folic acid supplements were started.

#### **DISCUSSION**

This case was a typical example of the occurrence of aplastic crisis due to HPV B 19 complicating Hereditary spherocytosis in an otherwise apparently asymptomatic child. Previously, Green et al in 1984 reported an adult sibling pair with HS who developed aplastic crisis after a febrile illness, which was further diagnosed to be HPV B19 infection. They also found that the children of one of the patients also developed HPV B19 induced aplastic crisis, which was resolved with supportive care. These two adult patients were treated by blood transfusion and supportive care and were discharged after 6-8 days of hospital care. 10 Although the major targets of HPB19 are erythroid progenitor cells, HPB19 induced bone marrow damage does not exclusively involve a single cell lineage. In addition to reticulocytopenia, neutropenia and thrombocytopenia may be observed in patients with HPB19 infection.<sup>11,12</sup> In another case study, the authors describe an adolescent girl with aplastic crisis induced by HPB19 infection as an initial presentation of hereditary spherocytosis (HS).9

The bone marrow in patients with transient aplastic crisis is characterized by an absence of maturing erythroid precursors and presence of giant pronormoblasts.<sup>15</sup> Although giant pronormoblasts are suggestive of parvovirus B19 infection, they are not diagnostic of the disease.<sup>16,17</sup> Because of pancytopenia accompanied with a marked decrease in reticulocytes as well as a history of HS, it was natural to believe that the patient in the present case was suffering from aplastic crisis due to viral infection.

In the present study we described a case of hereditary spherocytosis with aplastic crisis owing to the occurrence of pancytopenia with fever and vomiting. One of the theories behind the etiology of transient pancytopenia in HPV B19 infections is that the virus could be responsible for the temporary arrest of hematopoiesis that leads to aplastic crisis in persons with chronic hemolytic anemia. The other hypothesis is the occurrence of HPV-associated hemophagocytosis leading to pancytopenia. <sup>13,14</sup>

#### **CONCLUSION**

Although the major targets of HPB19 are erythroid progenitor cells, HPB19 induced bone marrow damage does not exclusively involve a single cell lineage. So, here we report a typical presentation of aplastic crisis in known case of hereditary spherocytosis which was transient and the recovery with supportive care happened within 7 days.

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