International Journal of Contemporary Pediatrics
Pandian N et al. Int J Contemp Pediatr. 2024 Dec;11(12):1816-1818
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20243419

Case Report

Dengue fever with atypical manifestations in a child with beta thalassemia major post splenectomy

Nidarshana Pandian[1], Dhaarani Jayaraman[2]*, Naveen Chandher K.[3], Sarala Premkumar[3], Sudeep Gaddam[2], Julius X. Scott[2]

[1]Sri Ramachandra Institute of Higher Education and Research (SRIHER), Chennai, Tamil Nadu, India
[2]Department of Pediatric Hemato-Oncology, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Chennai, Tamil Nadu, India
[3]Department of Pediatrics, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Chennai, Tamil Nadu, India

Received: 13 October 2024
Revised: 03 November 2024
Accepted: 04 November 2024

***Correspondence:**
Dr. Dhaarani Jayaraman,
E-mail: dhaaranij@yahoo.com

ABSTRACT

Dengue is one of the significant public health concerns in endemic countries. With plasma leakage as the primary pathophysiology in the process of severe dengue infection with hemoconcentration and thrombocytopenia being the significant laboratory diagnostic features in children, the presentation in children with underlying hemato-oncological conditions is atypical and often, anemia is found in these children, instead of hemoconcentration. We describe a young boy with beta thalassemia major who underwent splenectomy for hypersplenism 5-months ago with dengue illness, characterized by features mimicking a severe post splenectomy opportunistic infection with fever, loose stools and abdomen tenderness. Leucopenia, thrombocytopenia, minimal pleural effusion, ascites and gall bladder wall edema prompted us to think of dengue and was found to be positive. Child improved with appropriate supportive care measures. Pediatricians should be aware of the differences in the presentation of tropical infections in children with hemato-oncological disorders. Tropical infections should be kept in mind even in children with underlying hemato-oncological conditions, to ensure diagnosis and appropriate management.

Keywords: Dengue fever, Dengue with warning signs, Post-splenectomy, Beta-thalassemia major, Anemia

INTRODUCTION

Dengue infection is a significant public health concern in developing countries like India and is spread through the *Aedes aegypti* and *Aedes albopictus* mosquito bite. Clinical manifestations grossly guide in the classification severity of dengue fever (DF). It varies with uncomplicated viremia with mild illness to DF with warning signs to dengue hemorrhagic fever (DHF) or dengue shock syndrome (DSS).[1,2]

Laboratory features like hemoconcentration and thrombocytopenia and leucopenia are grossly supportive during an outbreak before the antigen detection or serological tests are available.[1] In children with an underlying hemolytic anemia, hemoconcentration may be masked and hence, the atypical features have to be anticipated, not to miss the diagnosis.[3] We report a case of a 9-year-old boy with thalassemia major with dengue with warning signs, 5 months from the splenectomy.

CASE REPORT

Our index patient is known case of beta thalassemia major from a lower socio-economic status and rural background born to third degree consanguinity; he is on regular

transfusion and chelation therapy for 9 months of age. He developed hypersplenism and hence underwent splenectomy 6-months ago. He received appropriate vaccinations prior to splenectomy and was continued on oral penicillin prophylaxis after surgery. He was on regular transfusion once in 4 weeks and the last transfusion dated 20 days ago.

He presented to us with unrelenting high grade fever spikes with chills and rigors. He was reviewed at his native place and was started on intravenous antibiotics in view of post splenectomy status from D2. Despite antibiotics, fever persisted, his oral intake worsened, and he had severe abdomen pain with 4-5 episodes diarrhoea per day. He presented to our hospital on D5 of illness with fever (temperature 100.3-degree F), cold peripheries, minimal tachycardia (pulse rate 149/min) and borderline blood pressure records (90/50 mmHg). He required a fluid bolus in emergency room. His vitals stabilised with the management and child was further continued on maintenance IV fluids.

Baseline investigations done showed haemoglobin 8.7 g%, total leucocyte count if 2560 cells/cubic.mm with polymorphs 45% and lymphocytes 47% and platelet count of 2.11 lac/cubic.mm. Child was started on intravenous piperacillin tazobactam and metronidazole in view of significant fever and abdomen tenderness, with possibility of severe post splenectomy opportunistic infections (OPSI). Renal functions were normal; liver function tests showed mild transaminitis (SGOT 280 U/l and SGPT 145 U/l), serum bilirubin and albumin were normal.

His fever persisted and his repeat hemogram showed haemoglobin 6.4 g%, total leucocyte count if 5450 cells/cubic.mm with polymorphs 39% and lymphocytes 55% and platelet count of 1.01 lac/cubic.mm by D7 of illness. Ultrasonography of abdomen showed mild ascites, minimal bilateral pleural effusion and gall bladder wall edema (Figure 1). In view of fever, features of third spacing, leucopenia and thrombocytopenia, dengue NS1 antigen assay was sent and was found to be positive.

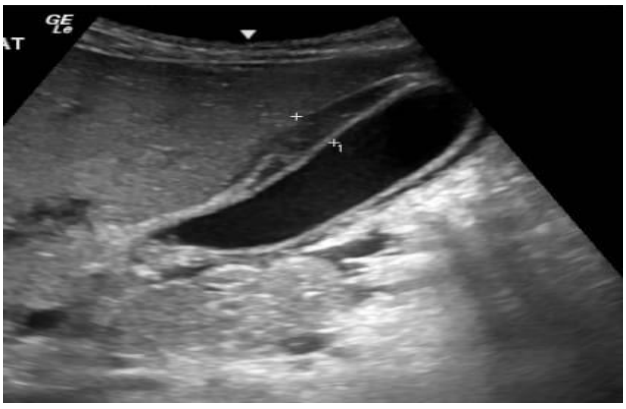

Figure 1: Ultrasonography picture showing gall bladder wall edema seen in dengue infection.

In view of persistent high grade fever spikes by D8 of illness and platelets dropping to 34,000/cubic.mm, serum ferritin was done and was found to be elevated (42,000 ng/ml). Possible need for IVIG in case of worsening has been considered; however, since D9 of illness, his fever and pain reduced, and platelets improved to 58,000/cubic.mm. Serum ferritin repeated was 16000 ng/ml. There were no bleeds. He was managed with 1 unit of leuko-filtered packed red cells transfusion during the course of illness in view of severe anemia. Serial hemogram showed improvement in WBC and platelet count by D11 of illness and could be discharged home by day-12 of illness.

DISCUSSION

DF is characterized by fever of 2-7 days duration with two or more features of headache, retro-orbital pain, myalgia, arthralgia, rash and bleeding manifestations. DF is associated with higher morbidity and mortality in very young children. Due to early diagnosis and appropriate management protocol guidelines, the mortality rates have declined over the last 2 decades. DF with warning signs is characterized by more clinical features from among abdominal pain/tenderness, persistent vomiting, third-spacing, mucosal bleeds, higher mental changes, hepatomegaly, increase in hematocrit and thrombo-cytopenia. Severe plasma leakage with hypotension, severe bleeding or severe organ involvement characterizes the more severe form, called as DHF.[1,2]

Anemia instead of hemoconcentration in the course of dengue fever is usually seen in severe DHF due to significant hemorrhage dropping the hematocrit. Rarely in very young children severe pancytopenia might ensue due to significant bone marrow suppression or possibly secondary to secondary hemophagocytic lymphohis-tiocytsosis (HLH).[3]

Beta thalassemia is an autosomal recessive condition with ineffective erythropoiesis leading to hemolytic anemia with a transfusion dependent state and iron overload eventually with repeated transfusions. Children with beta thalassemia often are reported to present with anemia during dengue infection, rather than hemoconcentration.[4]

In a study by Pongtanakul et al from Thailand, 20 patients with hemoglobinopathies and dengue fever were reported with varying severity of illness. It was noted that 90% had anemia, rather than hemoconcentration and 75% required packed red cell transfusion. Two of them developed secondary HLH requiring intravenous immunoglobulin (IVIG).[4] Hence, getting a baseline hematocrit in these children is very vital for the management of dengue illness.

Study by Ampaiwan et al revealed a case-fatality rate of 3.64% versus 0.63% in children with pediatric hemato-oncological conditions. Despite severe plasma leakage, these children were noted to have lower hemoglobin levels

and causes of anemia included acute hemolysis and acute bleeds.[5]

Padyana described a rare complication of a 28-year-old man with dengue with splenic rupture leading to hemoperitoneum and shock.[6] Similar report of a 22-year-old man with HB-H Constant Spring disease who presented with shock and abdominal tenderness revealing splenic rupture as early as day-2 of illness.[7] Splenic rupture in dengue fever is probably caused by congestion of the spleen and thrombocytopenia.[8] Subcapsular hematomas are reported to be found in found in 15% of dengue hemorrhagic fever cases. Our patient had only featured of third spacing and no hematomas/occult or overt bleeds.[9]

Similar report of less common manifestations and prolonged course was described in the report by Dinand et al in a 16-year-old boy with G-6PD deficiency and thalassemia intermedia. The child was having non-transfusion dependent thalassemia, however, undergone splenectomy 2-months ago in view of features of hypersplenism. During the dengue illness, required transfusion due to hemolytic episode.[3] The severity might be misinterpreted in view of anemia and diagnosis could be missed early due to lack of classical hemoconcentration.[4]

Whilst we anticipate a significant course of bacterial sepsis post splenectomy, dengue fever has not been noted to have increased severity in splenectomised patients. Deficit of phagocytes and T-helper cells, lack of splenic macrophages which are the dengue-permissive cells in spleen in DHF and lack of antibody dependent enhancement of severity of DF to DHF/DSS might lead to a rather protective role in such children as per the literature reviews.[10,11]

CONCLUSION

Tropical infections including dengue might be considered as a differential for OPSI in children with hematological conditions post splenectomy, as early diagnosis is very imperative for an appropriate timely management. Children with underlying hemato-oncological conditions with DF often have anemia and a misleading presentation in the early days of illness and hence, a high index of suspicion is the key to early diagnosis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Tayal A, Kabra SK, Lodha R. Management of Dengue: An Updated Review. Indian J Pediatr. 2023;90(2):168-77.
2. World Health Organization, Special Programme for Research, Training in Tropical Diseases, World Health Organization. Department of Control of Neglected Tropical Diseases, World Health Organization. Epidemic, Pandemic Alert. Dengue: guidelines for diagnosis, treatment, prevention and control. 2009. Available at: https://www.who.int/publications/i/item/9789241547871. Accessed on 12 August 2024.
3. Dinand V, Metzler V, Yadav SP, Sachdeva A. Atypical dengue Fever in a splenectomized boy with glucose-6- phosphodehydrogenase deficiency and thalassemia intermedia. Indian J Pediatr. 2011;78(4):482-4.
4. Pongtanakul B, Narkbunnam N, Veerakul G, Sanpakit K, Viprakasit V, Tanphaichitr VT, et al. Dengue hemorrhagic fever in patients with thalassemia. J Med Assoc Thai. 2005;88(8):S80-5.
5. Chuansumrit A, Natesirinilkul R, Pongtanakul B, Tangnararatchakit K, Sirachainan N, Tantiworavit A, et al. Dengue Infection in Pediatric Patients with Thalassemia: Aggravation of Anemia. Southeast Asian J Trop Med Public Health. 2012;43(2):311-22.
6. Padyana M, Gopaldas JA, Karanth S. A stitch in time – dengue with spontaneous splenic rupture. Radiol Infect Dis. 2020;7(3):145-8.
7. Ungthammakhun C, Chueansuwan W, Changpradub D. Dengue Hemorrhagic Fever Complicated with Spontaneous Rupture of the Spleen among Patients with Thalassemia and G6PD Deficiency: A Case Report. Arch Clin Med Case Rep. 2021;5:873-7.
8. Bhamarapravati N, Tuchinda P, Boonyapaknavik V. Pathology of Thailand haemorrhagic fever: a study of 100 autopsy cases. Ann Trop Med Parasitol. 1967;61:500-10.
9. Pungjitprapai A, Tantawichien T. A fatal case of spontaneous rupture of the spleen due to dengue virus infection: case report and review. Southeast Asian J Trop Med Public Health. 2008;39:383.
10. Jirillo E, Mastronardi ML, Altamura M, Munno I, Miniello S, Urgesi G, et al. The immunocompromised host: immune alterations in splenectomized patients and clinical implications. Curr Pharm Des. 2003;9(24):1918-23.
11. Blackley S, Kou Z, Chen H, Quinn M, Rose RC, Schlesinger JJ, et al. Primary human splenic macrophages, but not T or B cells, are the principal target cells for dengue virus infection in vitro. J Virol. 2007;81(24):13325-34.

Cite this article as: Pandian N, Jayaraman D, Chandher NK, Premkumar S, Gaddam S, Scott JX. Dengue fever with atypical manifestations in a child with beta thalassemia major post splenectomy. Int J Contemp Pediatr 2024;11:1816-8.