

Unusual Presentation of Rickettsial Fever as Henoch-Schonlein Purpura : A Case Report

Nigwekar P*, Kavar Y**, Shrikhande DY***

Abstract

Rickettsial fever (RF) is common ailment in many parts of India with high mortality and morbidity. It is prevalent in and around Loni. Henoch-Schonlein purpura (HSP) is generalized vasculitis with classical triad of palpable purpura, arthriagia, abdominal pain. We present an unusual case of RF presenting as HSP.

Key words : *Rickettsial fever, Weil-Felix test, Henoch-Schonlein purpura.*

Introduction

Rickettsial fever is a vector borne disease caused by the gram negative intracellular organism that presents as epidemic typhus, scrub typhus, spotted fever etc. It is prevalent all over India especially in Himachal Pradesh, Tamilnadu, Karnataka and some part of Maharashtra. Rickettsial fever is re-emerging, like dengue and leptospirosis in various parts of India[1,2,3]. Rickettsial fever has high mortality and morbidity, if not diagnosed early, and successful treatment require high index of suspicion. In 1915, a discovery by Weil and Felix led to the development of the widely used serological test for diagnosis of Rickettsial infections[6]. RF affect all age groups mainly children below 10 years with female preponderance[7,8,11].

Case report

Eight year old female presented with fever since 8 days, rash over body, joint pains and abdominal pain of 5 days duration. The rash was seen on lower limbs on extensor aspect and this later became purpuric in nature. Simultaneously both ankle joints were swollen and were painful. (Fig.1) Same day child had nonbilious vomiting and colicky abdominal pain. No history of oliguria and hematuria was present.

On examination patient was febrile (101°F). On general examination purpuric rash was palpable and was present

*Associate professor, ** PG student, *** Professor.

Department of Pediatrics, Rural Medical College, PIMS,

Address for Correspondence :

Dr. Prashant Nigwekar, Associate Professor, Dept. of Pediatrics, Rural Medical College, PIMS, Loni-413736 (M.S.) Maharashtra, India.413736, Email: nprashant59@gmail.com

over the lower limbs. Classical distribution was on extensor aspect over shin of tibia, back of legs and feet. Systemic examination was within normal limits, except mild splenomegaly.

Investigations revealed: Hb-10.3%, TLC-16000/cumm, with 75% polymorphonuclear leucocytes. Platelet count-4.8 lacs/cumm. Urine routine was normal. Liver function tests and renal function tests were within normal limits. Blood culture was sterile. X-ray chest was normal. Cerebrospinal fluid examination was within normal limits.



Figure 1 a) : Rash over feet and ankles



Figure 1 b) : Swelling ankle and wrist joints

Antistreptolysin (ASO) titre, C reactive protein (CRP) was negative. Prothrombin time and activated partial thromboplastin time was within normal limits. USG abdomen showed mild splenomegaly. Weil-Felix (WF) test revealed a strongly positive titre : OX2 1:160, OX19 1:160.

Patient was treated symptomatically. No apparent improvement was seen. WF test result became positive after two days. Hence oral doxycycline, 5mg/ kg body weight was started on the day of report. Dramatic improvement was noticed. There was remission of fever from the very next day of starting treatment. The rash gradually started fading, abdominal pain subsided. Oral doxycycline was continued for 7 days. Child was discharged on 8th day. The recovery was remarkable and uneventful.

Dissussion

Rickettsial fever is reemerging as an infection like dengue and leptospirosis. In India, spotty fever group and typhus are reported in different parts of the country, especially in Himachal Pradesh, Tamilnadu, Karnataka and some part of Maharashtra. Rickettsial disorders affects skin, central nervous system, heart, lungs and skeletal muscles. Proliferation of rickettsiae within endothelial cell cytoplasm leads to lymphohistiocytic or leukocytoclastic vasculitis of small venules and capillaries resulting in petechial skin lesion, microvascular leakage, tissue hypoperfusion and possibly end-organ ischaemic injury[11]. Fever and nonconfluent maculopapular rash involving palm and soles is a characteristic. Weil-Felix test demonstrates heterophile antibodies to strains of proteus mirabilis (ox-19, ox-2, ox-k). This test is positive in 50% cases. Though the test is non-specific it is the only easily performed test available in India. Even though WF agglutination test is not very sensitive, when positive, it is a very specific test[5]. Good correlation between the results of WF test and detection of IgM antibodies by an Immuno-fluorescence assay has been demonstrated [4]. In our case according to the WF titres, the most probable Rickettsial disease would be tick born spotted fever or epidemic typhus. Since no louse infestations (scalp and body infestation, lymphadenopathy) were seen and the patient belonged to low socioeconomic status from rural area, increased chances of tick infestation is likely. Cattle house is in close vicinity of patient's home which is additional supportive evidence. Further investigations like PCR could have been carried out to detect the specific Rickettsial organism. ELISA for *R. Coroni* is available in India. More definitive tests like Immunofluorescence, PCR

are expensive and not easily available. Rickettsia are dangerous pathogens. Typical rash and high index of suspicion, especially in endemic areas is required for early diagnosis and treatment. Excellent response is seen to doxycycline and chloramphenicol. Death may occur due to delay in treatment, due to encephalitis, Acute Respiratory Distress Syndrome (ARDS), myocarditis, hepatitis and renal failure. Early treatment on a high index of suspicion and necessary empirical specific treatment is justified as delay in initiating the treatment could be fatal[10].

HSP is one of the commonest vasculitis of childhood. Classical description is palpable purpura, arthralgia, and abdominal symptoms. Glomerulonephritis is seen in 10-20 % of patients[11]. The diagnostic criteria for HSP are palpable purpura (mandatory) in the presence of at least one of the following four features: Diffuse abdominal pain, arthritis (acute) or arthralgia, renal involvement (any haematuria and/or proteinuria), any biopsy showing predominant IgA deposition[9]. Abdominal pain is usually colicky. Intermittent vomiting is seen in 60% cases but hematemesis and malena rare. Most of the clinical features are self limiting and resolve in few days.

Conclusion

We suspected Rickettsial fever in this patient of classical HSP, due to typical rash and RF is also because endemic in and around Loni. A large number of rickettsial fever occur during September to February every year from nearby areas with a variety of presentations. Though palpable rash is described in RF, no such case report of association of RF and HSP is found in literature[10]. Association of RF and HSP can be explained on basis of generalized vasculitis which is seen in both conditions. Dramatic response to doxycycline supports a diagnosis of RF[11].

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