



ORIGINAL RESEARCH PAPER

General Medicine

A CASE OF SCRUB TYPHUS WITH SECONDARY HEMOPHAGOCYTTIC LYMPHOHISTOCYTOSIS

KEY WORDS: Scrub typhus with secondary hemophagocytic lymphohistiocytosis, fever, acute febrile illness,

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ABSTRACT

Scrub typhus is an acute fever sickness caused by Orientia tsutsugamushi and spread via the bite of the trombiculid mite chigger. At the bite site, there may be a single painless eschar with an erythematous ring. Fever, skin rash, myalgia, organomegaly, and lymphadenopathy are a few examples of the non-specific clinical symptoms. Meningoencephalitis, pneumonitis, acute renal failure, etc. are common side effects of scrub typhus. A rare complication is hemophagocytic lymphohistiocytosis (HLH). We report a case of 40 year old female presented with history of high grade continuous fever associated with macular rash on the legs later the Diagnosis of hemophagocytic lymphohistocytosis secondary to scrub typhus was made.

INTRODUCTION

Scrub typhus is an acute fever sickness caused by Orientia tsutsugamushi and spread via the bite of the trombiculid mite chigger. At the bite site, there may be a single painless eschar with an erythematous ring. Fever, skin rash, myalgia, organomegaly, and lymphadenopathy are a few examples of the non-specific clinical symptoms. Meningoencephalitis, pneumonitis, acute renal failure, etc. are common side effects of scrub typhus. A rare complication is hemophagocytic lymphohistiocytosis (HLH).

CASE DESCRIPTION

A 40 year old female presented with history of high grade continuous fever since 4 days associated with macular rash on the legs. She also had history of shortness of breath, myalgia, abdominal pain. There was no past history of diabetes, systemic hypertension, coronary artery disease, asthma, tuberculosis, chronic kidney disease. On examination pallor, icterus present.

Eschar was present. On systemic examination splenomegaly was present. IgM of scrub typhus was positive. Diagnosis of hemophagocytic lymphohistiocytosis secondary to Rickettsial infection was made with the following findings: Triglyceride: 300 mg/dL, Ferritin: 1800 mcg/dL, Fibrinogen: 90 mg/dL, Bone marrow aspiration and biopsy: Hyperactive macrophages with erythrophagocytosis, LDH: 2000 U/L, Bi-cytopenia: Hb:9 g/dL platelet count 90,000. Doxycycline (4 mg/kg daily) was added and patient received intravenous dexamethasone (10 mg/day), then tapered to 5 mg/day.

DISCUSSION

Hemophagocytic lymphohistiocytosis (HLH) is characterised by increased immune activity. The symptoms of HLH include fever, hepatosplenomegaly, lymphadenopathy, blood cytopenias, altered mental state, increased serum ferritin, and abnormal liver functions. Many HLH patients have a predisposing genetic abnormality and/or an immunologic trigger, which might be an infection, a cancer, a rheumatologic condition, or conditions linked to immunological dysregulation. Infections like scrub typhus may act as catalysts in the secondary type of HLH.

Diagnosis of HLH is by Presence of five of eight diagnostic criteria: (1) persistent fever, (2) splenomegaly, (3) unexplained cytopenia, at least two cell lines (haemoglobin <

90 g/L, platelets < 100,000 cells/mm³, absolute neutrophil count < 1000 cells/mm³), (4) hypofibrinogenaemia (≤150 mg/dL) and/or hypertriglyceridaemia (≥265 mg/dL, (5) hyperferritinaemia (≥500 ng/mL), (6) haemophagocytosis (in bone marrow, liver, spleen, or lymph nodes without evidence of malignancy), (7) low activity of NK cells, (8) high concentration of soluble CD25.

Rickettsia is an obligatory intracellular, non-motile, gram-negative proteobacteria. They are in coccus, bacillus, and threads forms. There is no human-to-human transfer. They usually require a vector for transmission, such as fleas, lice, mites, or ticks. An significant cause of acute febrile sickness in South and East Asia and the Pacific is scrub typhus, commonly referred to as bush typhus. It is caused by the gram-negative alpha proteobacterium of the Rickettsiaceae family, a intracellular parasite Orientia tsutsugamushi, which was initially discovered and described in Japan in 1930. Since it lacks both peptidoglycan and lipopolysaccharide in its cell wall, it differs from other Rickettsiae.

CONCLUSIONS

Hemophagocytic lymphohistiocytosis (HLH) is a rare life-threatening condition. Scrub typhus with hemophagocytic lymphohistiocytosis can result in DIC and multiorgan failure. Despite its rarity, scrub typhus may be lethal; as a result, practitioners must be aware of the necessity of detecting and treating suspected cases as soon as possible.

REFERENCES:

- [1] Pazhaniyandi S, Lenin R, Sivathanu S. Hemophagocytic lymphohistiocytosis with a leukemoid reaction in an infant with scrub typhus. J Infect Public Health 2015;8:626-9.
- [2] Agrwal S, Dabas A, Mantan M, Yadav S. Hemophagocytic lymphohistiocytosis with neurological manifestations in an infant with scrub typhus: A rare fatal occurrence. Trop Doct 2019;49:52-3.
- [3] Henter JI, Horne A, Aricó M, Egeler RM, Filipovich AH, Imashuku S, et al. HLH-2004: Diagnostic and therapeutic guidelines for hemophagocytic lymphohistiocytosis. Pediatr Blood Cancer 2007;48:124-31.
- [4] Bal M, Mohanta MP, Sahu S, Dwivedi B, Pati S, Ranjit M. Profile of pediatric scrub typhus in Odisha, India. Indian Pediatr 2019;56:304-6.
- [5] Ogawa M, Hagiwara T, Kishimoto T, Shiga S, Yoshida Y, Furuya Y, et al. Scrub typhus in Japan: Epidemiology and clinical features of cases reported in 1998. Am J Trop Med Hyg 2002;67:162-5.
- [6] Rathi N, Rathi A. Rickettsial infections: Indian perspective. Indian Pediatr 2010;47:157-64.
- [7] Naoi T, Morita M, Kawakami T, Fujimoto S. Hemophagocytic Lymphohistiocytosis Associated with Scrub Typhus: Systematic Review and Comparison between Pediatric and Adult Cases. Trop Med Infect Dis 2018;3:19.
- [8] Kwon HJ, Yoo IH, Lee JW, Chung NG, Cho B, Kim HK, et al. Life-threatening scrub typhus with hemophagocytosis and acute respiratory distress syndrome in an infant. J Trop Pediatr 2013;59:67-9.