Letter to the Editor / *Editöre Mektup*

DOI: 10.5578/ced.20240407 • *J Pediatr Inf 2024;18(4):e255-e256*

A Rare Cause of Fever of Unknown Origin and Pseudotumor Cerebri in Children: Mediterranean Spotted Fever

Çocuklarda Nedeni Bilinmeyen Ateş ve Psödotümör Serebrinin Nadir Nedenlerinden: Akdeniz Benekli Ateşi

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Cite this article as: Karakaya N, Kara Y, Kızıl MC, Yarar C, Us T, Kılıç Ö, et al. A rare cause of fever of unknown origin and pseudotumor cerebri in children: Mediterranean spotted fever. J Pediatr Inf 2024;18(4):e255-e256.

To the Editor

Rickettsiae are obligatory intracellular bacteria with cell membranes similar to gram-negative bacteria (1). *Rickettsia conorii* is the causative agent of Mediterranean spotted fever (MSF). Although the most common presenting symptoms are fever, headache, vomiting and rash, it may manifest diverse clinical symptoms. Although the prevalence of rickettsial diseases is increasing all over the world, identification of the disease and considering it in differential diagnosis may be challenging (2,3). Herein, a patient with MSF who presented with fever of unknown origin and clinical manifestations of pseudotumor cerebri was presented.

A 17-year-old male patient was admitted to our clinic with high fever, headache, vomiting, myalgia and rash persisting for nearly 10 days. On physical examination, a pink-purple 3 x 3 cm rash on the arms was revealed (Figure 1). Remarkable laboratory test results were as follows: WBC= 6500/mm^3, platelet= 122.000/mm^3, C-reactive protein= 148 mg/L, ALT= 163 U/L, and AST= 117 U/L.

During follow-up, headache accompanied with double vision necessitated application of lumbar puncture to obtain cerebrospinal fluid (CSF) material. CSF pressure was 34 cm/H_2O, and its glucose concentration was 71 mg/dL (blood glucose= 104 mg/dL) (7-15). Papilledema was detected in the fundus examination. In cranial and orbital MRI scans, optic nerves were tortoised bilaterally and intracranial pressure was high. Based on these findings, pseudotumor cerebri (idiopathic intracranial hypertension) was considered as the pre-diagnosis and acetazolamide treatment was initiated. In the follow-up, thrombocytopenia and lymphopenia were accompanied by high levels of acute phase reactants. Bone marrow aspiration biopsy performed by pediatric hematology department could not disclose the presence of blast cells. The results of ANA, Anti-ds DNA, C3, C4 tests requested from pediatric rheumatology department to exclude collagen tissue diseases were within their normal ranges. High fever and accompanying symptoms persisted, and we could not arrive at a diagnosis based on biochemical and invasive test results. Then, we learned that the patient had worked in the

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Received: 18.01.2024 **Accepted:** 28.03.2024 **Available Online Date:** 13.12.2024

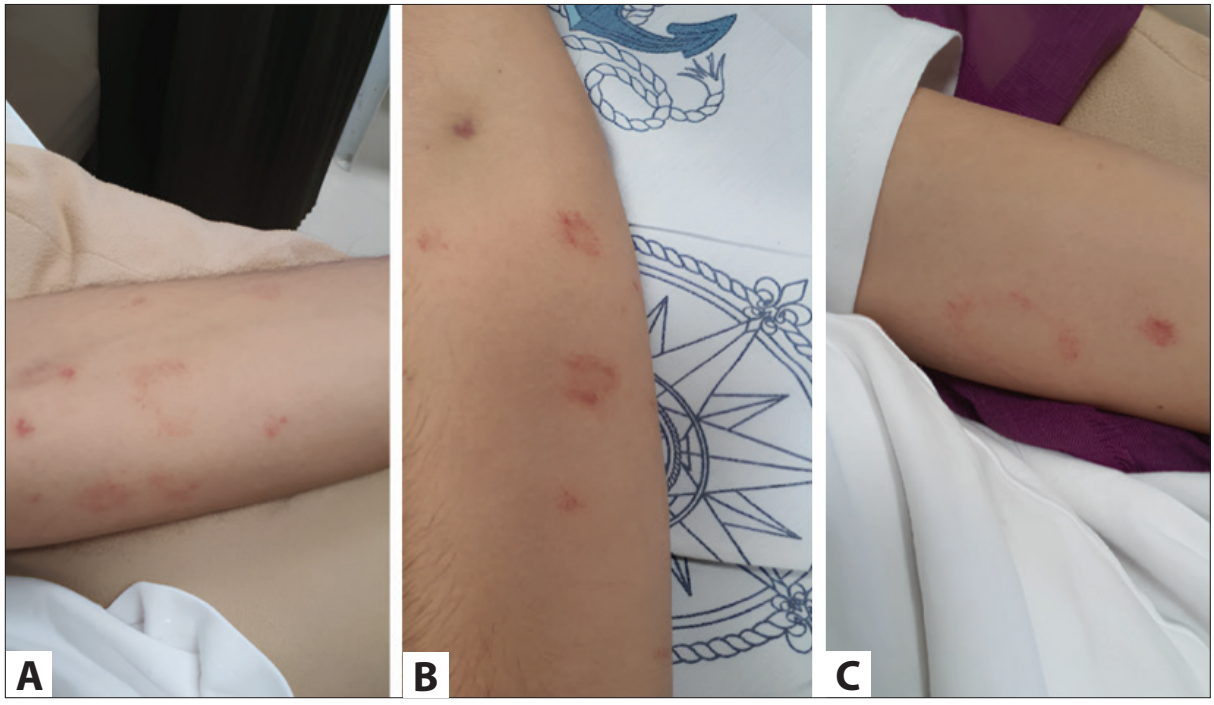

Figure 1. A. Rickettsial maculopapullar rash on the anterior forearm. **B.** Rickettsial maculopapullar rash on the leg. **C.** Rickettsial maculopapullar rash on the posterior forearm.

woods intermittently and was occasional exposed to insect and tick bites. A blood sample sent to a reference laboratory was examined in terms of Crimean-Congo Hemorrhagic Fever and rickettsial diseases. In the indirect immunofluorescence test (IFA), *R. conorii* IgM was positive 1/192 (<1/64). Based on the diagnostic criteria of the European Society of Clinical Microbiology and Infectious Diseases (ESCMID), ESCAR study group (ESCMID Study Group for *Coxiella, Anaplasma, Rickettsia* and *Bartonella*) diagnosis of MSF (diagnostic score: 31) was made, and doxycycline treatment was started (3). On the third day of the treatment, he was discharged with the recommendation of continuation of oral acetazolamide therapy and maintenance of oral doxycycline therapy up to 10 days.

Although our country is an endemic region for rickettsiosis, insufficient recognition and awareness of its symptoms is a challenging issue. Residency in rural areas, and exposure to ticks should be definitely questioned in patients with typical MSF symptoms especially in summer (2-5). In cases of suspected MSF, accessibility to gold standard diagnostic laboratory tests such as IFA should be fascilitated. It should be kept in mind that patients with MSF may also present with atypical clinical findings.

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