A patient with spotted fever group rickettsiosis mimicking connective tissue disease

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Introduction

Common clinical features associated with spotted fever group (SFG) rickettsioses include fever, headache, arthralgia, arthritis, gastro-intestinal disturbances, lymphadenopathy, hepato-splenomegaly and a discrete maculo-papular rash [1]. However, diverse clinical manifestations associated with SFG rickettsioses have been reported, some presenting with a continued febrile illness with no definite features suggestive of rickettsioses [2]. We describe a patient with SFG rickettsiosis who had small joint arthropathy, cervical lymphadenopathy, hair loss, oral ulceration, and intermittent high fever over a 4 month period. She was later confirmed of having SFG rickettsiosis and all her clinical features resolved with anti-rickettsial antibiotics.

Case report

A 35-year old married sub-fertile woman presented in April 2009 with high intermittent fever associated with chills and rigors, a dry cough, loss of appetite, lethargy, symmetrical small joint arthropathy, (which had appeared over about two weeks of initial febrile illness and persisted with varying degrees of severity) and painful cervical lymphadenopathy for over 4 months, with worsening of symptoms over the past 10 days. She had undergone many haematological, histological (lymph node biopsy) and biochemical investigations, but no definitive diagnosis had been made. On examination she looked ill, had oral ulcers, but was not pale or icteric and there was no clinical evidence of a vasculitis. Her hair was sparse and thin but there was no scarring alopecia. She had 1-2 cm, tender, posterior deep cervical lymph nodes on the left side and anterior lower deep cervical nodes on the right. There were inspiratory crackles in both lung fields, but abdominal examination was normal (there was no hepatosplenomegaly). Her clinical presentation was suggestive of a connective tissue disorder. The investigation results were as follows; WBC: 2.9×10⁹/μl, N-40%, L-55%, Hb-12.8 g/dl, Platelet count-223×10⁹/μl, blood picture - normal red cell morphology, numerous reactive lymphocytes but no abnormal cells. ESR 65 mm 1st hour, CRP 2.6 (normal 0-5mg/l), SGPT 28 IU/l, SGOT 32 IU/l, serum LDH 3236 IU/l, normal chest X ray, normal ultrasound of the abdomen. ANA, anti-DsDNA, HIV antibodies, EBV antibodies, toxoplasma antibodies, and mantoux test were negative. Two lymph node biopsies which had been done within a space of two months showed reactive changes, and was negative for tuberculosis by PCR. Because she was ill and had high fever she was given intravenous ceftriaxone and levofloxacin empirically, but had no clinical improvement over the next 5 days.

As she was having continued high intermittent fever associated with lymphadenopathy, we subsequently tested her for rickettsioses by IFA test using rickettsial antigens prepared from cell culture grown rickettsiae: Rickettsia conorii (Malish) (RC), Rickettsia typhi (Wilkinson), and Orientia tsutsugamushi (Karp) (OT). IgG antibodies were detected using fluorescein-conjugated goat anti-human IgG(γ) and IgM (KPL, Inc., Gaithersburg, MD) and the serum was strongly positive for SFG rickettsioses with a dilution titer of >1:8192. She was started on oral doxycycline, but reacted to the drug after 2 days with epigastric pain and vomiting. She was therefore given oral azithromycin and intravenous chloramphenicol to which she showed a good clinical response over three days. When reviewed 6 months and 12 months after leaving hospital she remained asymptomatic with complete regression of lymphadenopathy and oral ulcerations. She had re-grown her hair. Her ANA which was repeated remained negative and the IFA dilution titer using R. conorii antigen was 1: 128 after one year.

Discussion

This patient with SFG rickettsiosis presented with a clinical picture suggestive of a connective tissue disorder such as systemic lupus erythematosus or a lymphoma, but there was no evidence for these conditions. Infections such as infectious mononucleoses, toxoplasmosis, HIV and tuberculosis were also excluded. She had a good clinical response to a combination of azithromycin and chloramphenicol which have good anti-rickettsial activity.
Introduction

Cerebrotendinous xanthomatosis (CTX) is a rare disorder of lipid metabolism. We report two siblings with this genetic disorder.

Case report

A 26-year old woman was referred for evaluation of progressive difficulty in walking which started at the age of 12 years. She had developed bilateral cataracts at the age of 15. Examination showed bilateral pyramidal signs with cerebellar ataxia. She had xanthomas involving elbows, wrists, knees and ankles bilaterally. Further enquiry revealed a history of similar illness in a brother aged 31. He had developed seizures at the age of 4 years, difficulty in walking at 5 years, and bilateral cataracts at 10 years. Examination showed cerebellar ataxia with pyramidal signs, facial dysmorphism due to facial xanthomas and peripheral xanthomas (Figure 1).

Investigations of both siblings showed normal serum cholesterol, fasting blood sugar, renal function, urinalysis and thyroid profiles. X-rays of chest, and bilateral knee and ankle joints were normal. MRI scanning showed symmetrical and heterogenous signal changes in cerebellar dentate nuclei bilaterally (hypointense in T1, and hyperintense in T2 and FLAIR sequences), which are characteristic of CTX (Figure 2).

Two siblings with cerebrotendinous xanthomatosis

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