Kikuchi-Fujimoto disease
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In February, 2013, a 30-year-old Ugandan-born woman, working in the UK, presented to the accident and emergency department with a 4-week history of arthralgia, fever, and lethargy, and a recent episode of possible tonsillitis. During the third week of symptoms, she developed epistaxis, periorbital swelling, and rash. She had returned from Uganda 7 days before the onset of symptoms, having spent time in Kampala and Lira. She did not complete her doxycycline malarial prophylaxis. 4 weeks earlier, there had been an Ebola outbreak near Kampala. She denied exposure to tick or flea bites, rural villages, funerals, hospitals, or unwell contacts. On examination she had a maculopapular, hyperpigmented, pruritic rash affecting her upper arms and face (figure A); subsequently, she developed a vasculitic rash affecting the digital pulps of her extremities. She had oedematous tonsillar fauces and a unilateral anterior cervical lymphadenopathy. She spiked fevers of 38.9°C up to three times daily with mild tachycardia. Examination was otherwise unremarkable. During admission she received multiple regimens of broad-spectrum antibiotics.

Initial investigations revealed a white blood cell count of 1·9 × 10⁹ per L, neutropenia (0·8 × 10⁹ per L), lymphopenia (0·6 × 10⁹ per L), C-reactive protein 18 mg/L, erythrocyte sedimentation rate 44 mm/h, alanine aminotransferase concentration 282 IU/L, lactate dehydrogenase 1345 IU/L, and activated partial thromboplastin time ratio 1·59 (partly correctable). Blood cultures were negative as were serological tests for malaria, HIV, hepatitis A virus, hepatitis B virus, hepatitis C virus, hepatitis E virus, cytomegalovirus, Epstein-Barr virus, adenovirus, influenza, respiratory syncytial virus, Mycoplasma, Chlamydia pneumoniae, dengue virus, tick-borne encephalitis, yellow fever virus, west Nile virus, Rift valley virus, chikungunya, sindbis virus, Sandfly fever group, spotted fever group (rickettsiae), Coxiella, Bartonella, Strongyloides, filariasis, and Toxoplasma. The result for typhus was equivocal and so doxycycline was restarted (repeat serology was negative). An autoimmune screen showed positive antinuclear antibody, antiphospholipid, and anti-dsDNA antibodies. C4 was marginally raised and lupus anticoagulant was equivocal. A CT scan of her chest, abdomen, and pelvis confirmed cervical and axillary lymphadenopathy only. Cervical lymph node core biopsy was sent for histology, and showed patchy necrosis on haematoxylin and eosin, CD20, and CD34 stains with widespread histiocytic infiltration on CD68 staining (figure B), suggestive of a histiocytic necrotising lymphadenitis, in keeping with Kikuchi-Fujimoto disease. At follow-up in July, 2013, our patient was well, with resolution of symptoms and regression of antinuclear antibody titre. She had not developed features of connective tissue disease, but remains at risk of doing so.

Kikuchi-Fujimoto disease was first reported in Japan in 1972. It typically affects people of Asian descent, and women are four-times more likely to be affected than men. It has rarely been reported in the black African population. The disease is suspected to be caused by a virus, but no specific agents have been identified. The disease is characterised by a unilateral posterior cervical lymphadenopathy. Lactate dehydrogenase and alanine aminotransferase concentrations can be raised; neutropenia and lymphopenia are common. Histologically, the dermatological manifestations of Kikuchi-Fujimoto disease have features in common with systemic lupus erythematosus. Rarer manifestations include serositis, aseptic meningitis, myocarditis, pneumonitis, hepatitis, and acute kidney injury. Diagnosis is based on lymph node histology with excision biopsy providing the best diagnostic accuracy. Treatment is supportive. Corticosteroids are considered if the clinical course is severe or persistent. Generally, the disease is self-limiting with recovery in weeks to months. Patients are at increased risk of developing autoimmune diseases; by 53% according to the results of one study with an average of 10 years of follow-up. This case serves as a reminder that differential diagnosis of pyrexia of unknown origin should not be unduly skewed by an exotic travel history.

Contributors
JD and RS cowrote the report. JDe helped finalise the report. All authors approved the final version. Written consent to publish was obtained.

References

Figure: Kikuchi-Fujimoto disease
(A) Rash affecting upper arms. (B) Haematoxylin and eosin stain of lymph node core biopsy, with CD68 stain inset.