Interactive case report

Fever of unknown origin: case outcome

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Four weeks ago we presented the case of a 19 year old student who presented with a spiking fever (2 September, p 484). Her condition deteriorated and she required intensive care. As she had no confirmed diagnosis her parents requested a second opinion (9 September, p 541). The parents were informed that her underlying disease had triggered a systemic inflammatory response affecting various organs with an uncertain prognosis. Although it is generally acknowledged that patients do not have an automatic right to a further opinion, doctors should always respect a patient's and relatives' wishes for a second opinion unless there are justifiable reasons for refusal—for example, if it may be harmful to the patient. Such requests should be handled sensitively and patients should be made to feel comfortable.

Our working diagnosis was of adult onset Still's disease. She was started on intravenous hydrocortisone, angiotensin converting enzyme inhibitors, and β blockers. The second medical opinion agreed with our assessment and diagnosis.

Several diagnostic criteria exist for adult onset Still's disease. The box lists Yamaguchi's diagnostic criteria. Although our patient satisfied the diagnostic criteria for Still's disease, she went through a difficult course with several complications rarely seen in this condition. High serum ferritin concentrations (>1500 $\mu\text{g/l}$) in patients with fever of unknown origin can

indicate adult onset Still's disease. Ferritin synthesis is increased by hepatocytes in response to inflammatory cytokines, and ferritin concentrations are correlated with disease activity. In the absence of an alternative diagnosis and with the clinical characteristics detailed, the patient's illness was thought to be consistent with Still's disease.

Her fever and C reactive protein concentrations improved considerably with the steroids. She was also given intravenous immunoglobulin. Her recovery was complicated by the onset of pancytopenia. A bone marrow aspirate showed a hypocellular picture with no evidence of haemophagocytosis, and the conclusion was that she had drug related marrow suppression secondary to broad spectrum antibiotics. This recovered spontaneously once the antibiotics were stopped. She made a good recovery with clear improvement in her clinical condition, and her left ventricular function was normal on subsequent echocardiography. The concentrations of inflammatory markers normalised, her serum ferritin concentration came down to 733 (normal 20–200) $\mu\text{g/l}$, and she was switched to oral prednisolone at a tapering dose.

Clinical management was later frustrated by a renewed opinion on the lymph node biopsy, which raised the possibility of peripheral T cell lymphoma. No lymphadenopathy or organomegaly was found on repeat computed tomography of the chest, abdomen, and pelvis and no features of lymphoma have been apparent over six months follow-up after discharge. She is doing well on a small dose of prednisolone and weekly methotrexate. We asked her parents to comment since some of the most difficult communication challenges occurred when the patient was unconscious.

Competing interests: None declared.

doi 10.1136/bmj.38950.395868.68

This is the final part of a three part case report, which describes the outcome and summarises the comments made by readers during the case presentation. Further responses are welcome through bmj.com

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BMJ 2006;333:691–4

Yamaguchi's diagnostic criteria for adult onset Still's disease

Major criteria

Fever of 39°C for more than 1 week
Arthralgia or arthritis for more than 2 weeks
Typical evanescent rash
Neutrophilia

Minor criteria

Sore throat
Lymphadenopathy or splenomegaly
Liver dysfunction
Negative test results for rheumatoid factor and antinuclear antibody
Patients must meet five or more criteria, including at least two major criteria.

We welcome contributions of interactive case reports. Cases should raise interesting clinical, investigative, diagnostic, and management issues but not be so rare that they appeal to only a minority of readers. Full details of the criteria are available at: bmj.com/cgi/content/full/326/7389/564/DC1