Still’s disease and myocarditis associated with recent mumps infection

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Summary: We describe a 16 year old patient who developed Still’s disease with evidence of myocarditis. A rise in the mumps ‘V’ antigen indicated that the disease was associated with recent mumps infection.

Introduction

The characteristic features of Still’s disease are fever, an evanescent rash and polyarthritis lasting at least 3 months, occurring in patients below the age of 16. The disease is associated with systemic manifestations which may include lymphadenopathy, splenomegaly, neutrophil leucocytosis, hepatitis, pleuritis, myocarditis and pericarditis. An adult form of the disease was first recognized in 1971. The aetiology of Still’s disease is unknown and case control studies have failed to demonstrate an association with virus infection. We present a case of Still’s disease with evidence of recent mumps infection.

Case report

A 16 year old Chinese male patient was admitted with a one week history of pyrexia, mild diarrhoea, polyarthritis and a macular rash on the upper limbs and trunk. There had been no response to oral erythromycin. The patient was studying in England and had recently returned from a visit to Hong Kong. Past medical history was unremarkable and he denied any sexual contact or intravenous drug abuse.

Physical examination revealed pyrexia (39.5°C), widespread lymphadenopathy, mild pharyngitis and a maculopapular rash affecting the wrists, abdomen and thighs. The knees, ankles and right elbow were swollen, warm and tender with reduced range of movement. He had a sinus tachycardia, 120/min, but initial examination of the heart and chest was otherwise normal.

Investigations in the acute stage showed haemoglobin 13.4 g/dl, white cell count 20.4 x 10⁹/l (89% neutrophils); no malarial parasites were seen on several blood films, plasma viscosity 2.01 cP (normal 1.5–1.72), negative Paul Bunnell test on two occasions, electrolytes, glucose, corrected calcium and renal function were normal. Liver function tests were mildly deranged, bilirubin 19 μmol/l (normal <17), alanine aminotransferase (ALT) 131U/l (2–53), gamma glutamyl transferase (GGT) 94U/l (0–50), alkaline phosphatase 1031U/l (40–130). C-reactive protein was raised at 32 mg/dl (normal <1), immunoglobulins normal, auto-antibody screen and rheumatoid factor (IgM and IgG) were negative, and hepatitis B surface antigen test was negative. Multiple blood cultures were negative, microscopy and culture of throat swabs, urine and faeces were also negative; joint aspirate from the right elbow showed numerous red cells and white cells on microscopy but the Gram stain and culture were negative. Chest radiograph, abdominal ultra-sound and X-rays of affected joints were reported as normal. Admission electrocardiogram (ECG) showed right axis deviation and partial right bundle branch block.

His fever continued with daily spikes to 40°C and he complained of mild central chest pain, worse on inspiration. On the sixth day after admission he developed a grade 4 ejection systolic murmur in the aortic area. The following day this became less prominent but a transient pulmonary diastolic murmur was noted, an echocardiogram was performed which showed no valvular abnormalities. Eight days later a new ejection systolic murmur was heard in the pulmonary area, a repeat echocardiogram and sector scan were normal. A chest radiograph at this stage showed mild cardiomegaly and the ECG demonstrated first degree heart block (PR interval 0.22 seconds) with T wave inversion in the ventricular leads. Cardiac enzyme analysis showed an elevated hydroxybutyrate dehydrogenase of...
430 IU/l (normal 100–240) although there was no corresponding rise in creatine kinase. Liver function tests became increasingly deranged, at 2 weeks after admission bilirubin was 20 μmol/l, ALT 548 IU/l, GGT 202 IU/l, alkaline phosphatase 244 IU/l.

Initial diagnoses of rheumatic fever or subacute bacterial endocarditis were considered and he was commenced on aspirin 1.5 g/day, intravenous benzyl penicillin and gentamicin. He made little response, his temperature persisted and Still’s disease was diagnosed. Antibiotics were discontinued after 6 days and he was commenced on prednisolone 20 mg/day and indomethacin 50 mg three times daily, with immediate improvement. The arthritis, hepatic and cardiac abnormalities gradually resolved over several months and the dose of prednisolone was reduced and stopped after 6 months. At 7 months after admission he remains asymptomatic on indomethacin 25 mg three times daily, and the chest X-ray and ECG have returned to normal.

Viral cultures from throat swabs, urine and faeces were negative. Serology showed no evidence of infection with hepatitis A, coxsackie B, Mycoplasma pneumoniae, cytomegalovirus, yersinia or toxoplasma; however, serology to mumps revealed a rise of the ‘V’ antigen from 1/16 on admission to 1/128 after 5 days, declining to 1/32 after 3 months. There was no rise in the ‘S’ antigen. This pattern would be compatible with a recent mumps infection.

Discussion

This patient presented with fever, polyarthritis, rash and lymphadenopathy. There was also evidence of myocarditis and hepatitis. A clinical diagnosis of Still’s disease was made and he responded to treatment with indomethacin and prednisolone. There were several unusual features including the age of onset, prominent myocardial involvement and the association with mumps infection. Still’s disease normally present between the ages of 3 and 10 with a second group presenting over the age of 16 with a median age at onset of 25 defined as adult onset disease.4 Our patient presented at the age of 16 and therefore lies between the adult and juvenile forms of the disease although the epidemiology of the condition in Chinese people has not been well documented. The illness was complicated by myocarditis with tachycardia, ECG changes, cardiomegaly and a rise in cardiac enzymes. Transient changing murmurs were heard over several days suggesting a valvulitis or endocarditis but echocardiography on two occasions was normal. Cardiac involvement in juvenile Still’s disease is well recognized, occurring in 5–10% clinically5–7 and up to 36% investigated by echocardiography.8 The cardiac manifestations include pericarditis, myocarditis and valvulitis which may be complicated by arrhythmias and cardiac failure. In contrast, myocarditis in association with adult onset Still’s disease has only been reported once.9

Arthritis is a rare complication of mumps infection. The most frequently described joint manifestation is a migratory polyarthritis although monoarticular arthritis and arthralgia have also been reported.10 There is also a single case report of mumps infection presenting as adult onset Still’s disease.11 The cardiac complications of mumps include ECG changes in up to 15% but clinically myocarditis is very rare.12

The differential diagnosis between a prolonged viral infection and Still’s disease is often difficult to make. Follow-up in this case was for 7 months and the arthritis had settled by this time, further joint problems or radiological evidence of carpal fusion or apophyseal joint fusion in the neck would substantiate a diagnosis of Still’s disease. Although viral infections have not been clearly established as the cause of Still’s disease, intercurrent infections are associated with exacerbations of the condition.13 Our case demonstrates that Still’s disease and myocarditis may complicate mumps infection which should be considered in a patient presenting with a fever, rash, arthritis and cardiac abnormalities.

References


