Multisystem disease in post-streptococcal arthritis

Avi Livneh, Kumar Sharma, Kathryn L Sewell, Harold D Keiser

Abstract

The case presented is of a patient with migratory polyarthritis and serological evidence of a recent streptococcal infection, consistent with the diagnosis of acute rheumatic fever, who in addition had multisystem disease manifestations. This case supports the concept that the sequelae of streptococcal infection can encompass a broader clinical spectrum than is suggested by the Jones criteria for the diagnosis of acute rheumatic fever.

In recent years acute rheumatic fever as defined by the revised Jones criteria¹ has been a comparatively rare disease in most of the developed world. Arthritis and less often carditis have been the only 'major' manifestations noted, particularly in adults.²⁻³ At the same time it has become apparent that patients with acute rheumatic fever may develop disease manifestations not included in the revised Jones criteria, such as pneumonitis, nephritis, dermal vasculitis, or neuritis.⁴⁻⁷ The following case report illustrates the occurrence of these multisystem manifestations in a patient with a post-streptococcal arthritis.

Case report

A 58 year old black man with no appreciable medical history developed a migratory polyarthritis affecting the ankles, knees, and elbows two weeks before admission. Indomethacin, 25 mg three times a day, relieved the joint pain, but after three days the patient noted a rash on his hands, feet, and ankles and a painless penile ulcer. He stopped the indomethacin, but the rash persisted and migratory arthritis affecting the ankles and the knees appeared again. On the night of admission the patient had severe left pleuritic chest pain and shortness of breath. He denied recent sore throat, any oral, ocular, urethral, or gastrointestinal abnormalities, exposure to venereal disease, drug or alcohol abuse.

On physical examination his temperature was 38.5°C and pulse 120. Palpable purpuric lesions were present on both aspects of the hands and dorsal aspect of the feet. There was a non-tender indurated deep ulcer 1 cm in diameter, starting to heal, on the shaft of the penis. The right ankle and the left knee were red, swollen, and tender. Rales were heard in the lung bases. The remainder of the physical examination was normal. Laboratory findings on admission included a Westergren erythrocyte sedimenta-

tion rate of 140 mm/h, slightly raised values of alkaline phosphatase, transaminases, and lactic dehydrogenase, serum creatinine 115 µmol/l, haemoglobin 110 g/l with normochromic normocytic indices, white blood cell count $16 \times 10^9/l$ with 80% neutrophils, and arterial partial oxygen pressure 68 mmHg. The urine sediment showed 10 red blood cells and two white blood cells per high power field and 1 gram of protein in a 24 hour specimen. Chest radiography showed a small left pleural effusion and left basilar atelectasis. Fluid aspirated from the knee had a white blood cell count of $10 \times 10^9/l$ and a normal glucose concentration; no crystals or micro-organisms were found. Other blood chemistry tests and enzyme determinations, the electrocardiogram, and a ventilation/perfusion lung scan were normal. The patient's chest pain subsided spontaneously shortly after admission. Throat, genitourinary tract, and blood cultures were obtained initially, and then intravenous penicillin was started for presumed mixed infection with syphilis and gonorrhoea.

During the first week in hospital the patient's fever continued, rising to 39°C, and his right knee, left ankle, and both elbows became swollen and painful with marked periarticular swelling and erythema. Premature ventricular contractions were noted on the cardiogram. Liver enzyme activity remained mildly abnormal, the haemoglobin dropped to 90 g/l without evidence of bleeding or haemolysis, the platelet count rose to $700 \times 10^9/l$ and the white blood cell count to $40 \times 10^9/l$, with 80% neutrophils. Repeated Venereal Disease Research Laboratory and fluorescent treponeme antibody absorption tests, antinuclear antibody test; rheumatoid factor test, total haemolytic complement, antinative DNA antibody, hepatitis B surface antigen and antibody, cryoglobulins, gonococcal cultures of body orifices, multiple blood cultures, cultures of the throat, urine, and synovial fluid, bone marrow biopsy and culture, sonogram and computed tomographic scan of the abdomen, renal and coeliac angiography, echocardiography, and slit lamp examination of the eyes were all normal or negative. Biopsy of a purpuric lesion showed mild perivascular inflammation. Dark field examination and biopsy of the penile ulcer were not performed because the dermatologists considered that the penile ulcer was healing and unlikely to yield diagnostic information.

Early in the second hospital week the anti-streptolysin O titre measured on admission was reported to be 500 units. A diagnosis of acute rheumatic fever was suggested; penicillin was

Department of Medicine,
Sheba Medical Center,
Tel-Hashomer, Israel,
and the Department
of Medicine,
Albert Einstein College
of Medicine, Bronx,
NY, USA
A Livneh
K Sharma
K L Sewell
H D Keiser

Correspondence to:
Dr Avi Livneh,
Department of Medicine,
Sheba Medical Center,
Tel-Hashomer 52621, Israel.

Accepted for publication
19 March 1990

continued to complete a 10 day course and aspirin, 900 mg every four hours, was begun. The arthritis, rash, and penile ulcer gradually resolved, the patient's temperature dropped to 38°C, the white blood cell count decreased gradually, and the urine analysis and chest radiograph returned to normal.

At the beginning of the third week in hospital the patient noted a small mass in the lower lateral aspect of his left thigh without inguinal adenopathy. Aspiration of this subdermal abscess yielded 5 ml of bloody pus, which was negative for micro-organisms, including mycobacteria, on smear and culture. The patient's condition continued to improve and he was discharged free from symptoms with tapering doses of aspirin at the end of the third hospital week. The antistreptolysin O titre at the time of discharge was still 500 units. On follow up examinations up to one year later no physical or laboratory abnormalities were found and the antistreptolysin O titre was less than 50 units.

Discussion

This patient's illness with its multiple manifestations can best be understood as a complex case of post-streptococcal arthritis. He presented with migratory polyarthritis, fever, a raised erythrocyte sedimentation rate, and serological evidence of a recent streptococcal infection, thus satisfying the revised Jones criteria for diagnosis of acute rheumatic fever.¹ His anaemia, leucocytosis, and mildly inflammatory synovial fluid are regular though non-diagnostic findings in acute rheumatic fever.⁸ Furthermore, most of the other manifestations of this patient's illness, including the increased liver enzymes,³ pleuritis and pleural effusion,⁹ pulmonary infiltrate,^{4 9 10} haematuria, proteinuria, and glomerulonephritis,⁵ palpable purpura with perivascularitis,^{6 11} and cardiac arrhythmias in the absence of overt carditis,² have been described in patients with acute rheumatic fever or in association with previous streptococcal infection. Other systemic diseases, such as systemic lupus erythematosus or bacterial endocarditis, that might be thought to explain the manifestations of this patient's illness can be excluded by the history, the clinical and laboratory findings, and the clinical course.

An unusual feature of this patient's illness is the penile ulcer, which we have not found reported in association with previous streptococcal infection. Venereal disease seems highly unlikely in the absence of supporting history and laboratory evidence and the lack of response to antibiotic treatment. Behçet's syndrome or Reiter's disease are not considerations because the major distinguishing criteria for

these conditions were absent and these diagnoses would not account for the purpuric skin lesions and high antistreptolysin O titre.^{12 13} Instead, this patient's penile ulcer may be viewed as a manifestation of vasculitis after streptococcal infection, an established entity after such infection previously known to affect blood vessels of the skin, lung, tongue, kidney, heart, and nervous system.^{7 9 14 15} The sterile subdermal abscess found in the patient's thigh might also have been secondary to a focal vasculitis or, alternatively, the site of streptococcal infection which became sterile after the course of penicillin.

Acute rheumatic fever was perceived in the past as a systemic disease affecting many organs.⁹ More recently, a general decline in the incidence and severity of acute rheumatic fever, and widespread use of the Jones criteria, which emphasise the most specific and characteristic features of the disease in establishing the diagnosis, have tended to restrict our appreciation of its generalised nature and the variability of its presentation. Our patient's illness, with palpable purpura, pleuritis, liver and kidney abnormalities, and penile ulcer, together with the more common manifestations of acute rheumatic fever as it occurs now in adults,^{2 3} fits within the larger spectrum of post-streptococcal arthritis.

- 1 Stollerman G H, Markowitz M, Taranta A, Wannamaker L W, Whittemore R. Jones criteria (revised) for guidance in the diagnosis of rheumatic fever. *Circulation* 1963; 32: 664-8.
- 2 Ben-Dov I, Berry E. Acute rheumatic fever in adults over the age of 45 years: an analysis of 23 patients together with a review of the literature. *Semin Arthritis Rheum* 1980; 10: 100-10.
- 3 Barnert A L, Terry E E, Persellin R H. Acute rheumatic fever in adults. *JAMA* 1975; 232: 925-8.
- 4 Lustok M J, Kuzma J F. Rheumatic fever pneumonitis: a clinical and pathological study of 35 cases. *Ann Intern Med* 1956; 44: 337-56.
- 5 Bisno A L. The coexistence of acute rheumatic fever and acute glomerulonephritis. *Arthritis Rheum* 1989; 32: 230-2.
- 6 Samys W M, Thorne E G, Small P, Mass M F, McIntosh R M, Stanford R E. Leukocytoclastic vasculitis. *Arch Dermatol* 1976; 112: 219-26.
- 7 Lucas S B, Moxham J. Recurrent vasculitis associated with beta-hemolytic streptococcal infection. *BMJ* 1978; i: 1323.
- 8 Stollerman G H. Rheumatic fever. In: Kelley W M, Harris E D Jr, Ruddy S, Sledge C B, eds. *Textbook of rheumatology*. Philadelphia: Saunders, 1985: 1277-93.
- 9 Paul J R. Pleural and pulmonary lesions in rheumatic fever. *Medicine (Baltimore)* 1928; 7: 383-410.
- 10 Grunow W A, Esterly J R. Rheumatic pneumonitis. *Chest* 1972; 61: 298-301.
- 11 Goodyear P R, de Chadarevian J-P, Kaplan B S. Acute post-streptococcal glomerulonephritis mimicking Henoch-Schönlein purpura. *J Pediatr* 1969; 93: 412-5.
- 12 Weinberger H W, Ropes M W, Kulka J P, Bauer W. Reiter's syndrome. Clinical and pathologic observations. *Medicine (Baltimore)* 1962; 41: 35-91.
- 13 Chajek T, Fainaru M. Behçet's disease. Report of 41 cases and a review of the literature. *Medicine (Baltimore)* 1975; 54: 179-96.
- 14 Hodson A K, Doughty R A, Norman M E. Acute encephalopathy, streptococcal infection and cryoglobulinemia. *Arch Neurol* 1978; 35: 43-4.
- 15 Escudero J, Stanislawsky E, Escudero X. Fulminant acute rheumatic fever with multisystem involvement. *Am Heart J* 1983; 105: 161-2.