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Brief Report

Two unusual presentations of acute rheumatic fever

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Abstract Patients with acute rheumatic fever sometimes present with atypical signs and symptoms. In these circumstances, the Jones criterions may not be sufficient to make a clinical diagnosis. We describe here two patients with unusual presentations, highlighting that, both in regions where the disease is endemic, or where it is seen only sporadically, physicians should be more alert and careful in making the diagnosis.

Keywords: Acute rheumatism; abdominal pain; heart block

ACUTE RHEUMATIC FEVER IS A DIFFUSE inflammatory process involving the connective tissues that appears in approximately 0.3 percent of untreated patients suffering infections of the upper respiratory tract by the group. A beta-haemolytic streptococcus. Its importance stems from its morbidity and sequels rather than its mortality.[1–5] Minor manifestations, such as rheumatic pneumonia, epistaxis, and abdominal pain[1,5] are relatively non-specific, but are recognised as enhancing diagnostic suspicion. Sometimes, however, patients are admitted to hospital with particularly minor symptoms, which may be unusual.[6] It is axiomatic that the disease can only be diagnosed when it is suspected. In this light, we report here two interesting cases with very unusual presentations.

Case 1

The patient, a 12-year-old girl, had developed fever, a sore throat, and silent abdominal pain one week previously. Her infection of the upper respiratory tract had been treated for four days with amoxicillin-clavulanate. She was admitted to our hospital with severe abdominal pain, albeit without diarrhoea or vomiting.

On admission, she had a persistent sinus tachycardia, at 136 beats per minute, but was normothermic, at 37.8 degrees centigrade. Her blood pressure was 140 over 80 millimetres of mercury. On examination, we noted periumbilical abdominal tenderness. There were no audible cardiac murmurs, and no swollen joints. The rest of the physical examination was within normal limits. An abdominal ultrasonic examination revealed an oedematous and enlarged vermiform appendix. Appendectomy was done. Pathological sampling confirmed the presence of appendicitis and local peritonitis (Fig. 1). A few days later, her ankles became red, warm, swollen and painful, permitting

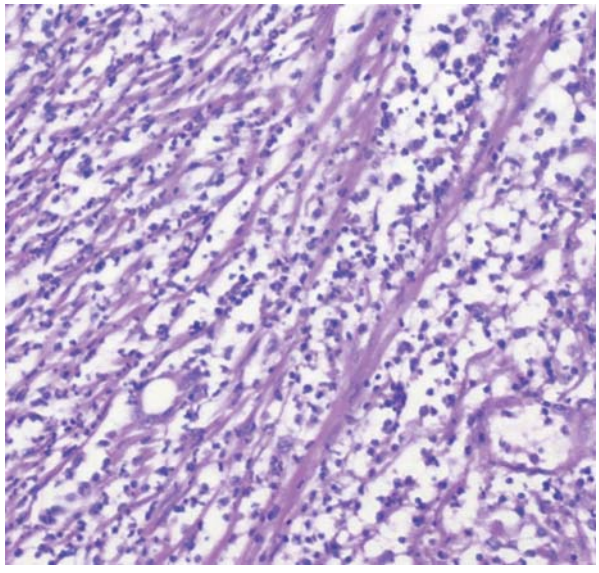

Figure 1.
The pathology specimen from our first patient shows massive polymorphonuclear lymphocytic infiltration through the muscular layers of the vermiform appendix. Haematoxylin eosin, with magnification by 100.

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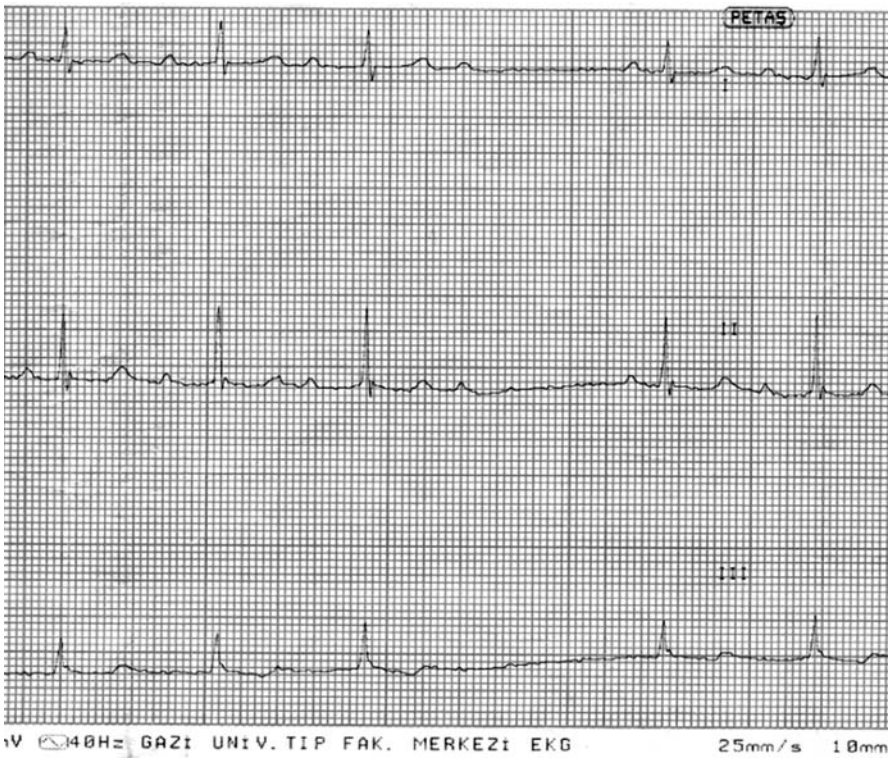

Figure 2.
The electrocardiogram of our second patient, on admission, showed second degree Mobitz I atrioventricular block.

only a limited range of movement. The changes in the ankles spontaneously disappeared on the second day. Her temperature was then 38.3 degrees centigrade, and she had a pansystolic murmur, of grade 2 or 3 out of 6, audible at the apex and radiating to the left axilla, along with an early diastolic murmur heard at the left side of the sternum. Laboratory evaluations revealed haemoglobin of 9.7 grams per decilitre; the white blood cell count at 12,600 per cubic millimetre, with 60 percent polymorphonuclear cells; a platelet count of 615,000 per cubic millimetre; titres of antistreptolysin O at 1660 international units; levels of C reactive protein at 183 milligrams per decilitre, and an erythrocytic sedimentation rate of 117 millimetres per hour. Other laboratory studies, including the electrocardiogram, were normal. Cross-sectional and Doppler echocardiography demonstrated second degree mitral insufficiency, first degree aortic insufficiency, but normal left ventricular systolic function. The cultures of the throat, urine and blood were negative. On the basis of these findings, we diagnosed acute rheumatic fever with carditis, and we commenced therapy with prednisolone and prophylactic benzathine penicillin. She was discharged from hospital in good condition, albeit still with aortic and mitral insufficiency.

Case 2

A 12-year-old boy presented with a low grade fever and pain in his left knee. A few days later, he noted that both knees had become warm, swollen and painful. He also complained of pain in the lower back and chest. On examination, his vital signs were normal. His left knee was warm and tender, albeit with a full range of motion. A pansystolic murmur, graded at 2 out of 6, was heard at the apex. Laboratory studies showed haemoglobin at 11.6 grams per decilitre; a white blood cell count of 9,940 per cubic millimetre, with 70 percent polymorphonuclear cells; a platelet count of 483,000 per cubic millimetre; a titre of antistreptolysin O at 499 international units, C reactive protein at 118 milligrams per decilitre, and an erythrocytic sedimentation rate of 76 millimetres per hour. Culture of the throat proved negative. An electrocardiogram revealed second degree Mobitz I atrioventricular block (Fig. 2). The chest X-ray was normal, but echocardiography revealed second degree mitral insufficiency.

Since the clinical and laboratory findings satisfied the Jones criterions for the diagnosis of acute rheumatic fever and carditis, we commenced treatment with prednisolone and prophylactic benzathine penicillin. The patient improved dramatically. Within two days, he was able to walk without pain and his electrocardiogram had reverted to normal. A week later, acute phase reactants were normal. He was discharged on prednisolone for six weeks, along with three weekly injections of benzathine penicillin.

Discussion

Abdominal pain is a nonspecific and relatively uncommon symptom of acute rheumatic fever, said

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to occur in less than one-twentieth of patients with acute rheumatic fever. When seen, it usually precedes other rheumatic signs by a few hours or a few days. Generally, it is mild and disappears quickly, but it can be severe and can mimic appendicitis.[1,7] Rheumatic abdominal pain can be difficult to distinguish from early acute appendicitis, particularly when the abdominal symptoms precede other rheumatic manifestations. As in our first patient, an abdominal investigation with ultrasonography can be helpful in establishing the correct diagnosis. The first diagnosis in our patient was acute appendicitis. Only a few days after the appendectomy did the patient develop arthritis and carditis. Pathological examination confirmed true inflammation of the appendix. This infiltration by inflammatory cells, however, may be a consequence of the acute rheumatic fever. Should serious doubt remain of the true cause of the appendicitis, it is better to remove a normal appendix than to risk rupture and generalized peritonitis.[8]

Our second unusual presentation was the finding of second degree Mobitz I atrioventricular block in the electrocardiogram on admission of the patient. In this boy, the diagnosis of acute rheumatic fever was suggested by the history of low-grade fever and arthritis in his left knee, clinical and echocardiographic evidence of carditis, and laboratory evidence of streptococcal infection. The patient regained normal sinus rhythm within two days with treatment with steroids. First degree heart block is a common electrocardiographic manifestation of acute rheumatic fever, and is included in the revised Jones criterions. Other electrocardiographic changes, such as sinus tachycardia, bundle branch blocks, nonspecific ST-T wave changes, and atrial and ventricular premature complexes, have been reported with variable frequency. Atrioventricular block of advanced degree, however, is an exceptionally rare manifestation. When seen, such advanced block with acute rheumatic fever appears to be self-limited in most instances. Specific treatment, such as insertion of a temporary pacemaker, should be considered only when clinical symptoms persist.[9] Unlike rheumatic valvitis, it is not associated with significant permanent morbidity. Second and third degree block should be considered a manifestation of acute rheumatic fever only when associated with other rheumatic signs, even in the absence of valvitis. In the absence of other major criterions, isolated atrioventricular block of second or third degree is most often caused by conditions other than acute rheumatic fever. The most common cause of electrocardiographic abnormalities in acute rheumatic fever is vagal hypertonia due to toxaemia. Direct involvement of the atrioventricular node, nonetheless, has been documented in some pathological studies of rheumatic heart disease.[10] In addition, although advanced atrioventricular block is rare during acute rheumatic fever, it may be more frequent than is thought, since as in our case, it subsequently resolves without specific treatment.

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