ATYPICAL RHEUMATIC FEVER IN YOUNG ADULTS

BY

P. C. REYNELL

Experiences gained in the armed forces have contributed much to our knowledge of rheumatic fever in adults. It has become clear that the "textbook" descriptions of the disease give a misleading picture of its commoner features as observed in servicemen. Several large series of cases have now been reported in America (Griffith, 1947; Sokolow and Snell, 1947; Ferguson, 1943; Rosenberg, 1946); but, apart from Copeman’s description of atypical rheumatic fever in the Middle East (1944), the subject has not received the attention which it deserves in this country. This paper is an analysis of the salient clinical features of a consecutive personal series of twenty-five cases of rheumatic fever admitted to the medical wards of a military hospital in Italy during a twelve-month period.

Table 1

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<tbody>
<tr>
<td>Cases</td>
<td>4</td>
<td>3</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>6</td>
<td>0</td>
<td>2</td>
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</table>

Aetiology

The age incidence was 18 to 40 years, and the distribution was approximately representative of the population at risk, with a slight bias towards the younger age groups. The dates of admission to hospital are shown in Table 1, which illustrates the expected drop in case incidence in the late summer and early autumn.

Twenty-one cases gave a history of an infection within 5 weeks of the onset of symptoms. The infections were as follows: tonsillitis (9), febrile coryza (9), septic foot (1), cystitis (1), and gonorrhoea (1). Of the remaining four cases, one suffered from a severe subacute naso-pharyngitis. The interval between the infection and the onset of symptoms is shown in Table 2.

Clinical Features

Jones (1944) has emphasized the importance of accepting set standards for the diagnosis of rheumatic fever, and twenty-four of the present series fulfil his criteria. The remaining patient differed from the others only in that he was afebrile by the time he was admitted to hospital. The most useful diagnostic criteria were the association of arthritis or arthralgia with fever and raised erythrocyte sedimentation rate, together with a history of an acute infection within the preceding five weeks. In six cases there was a history of a previous attack of rheumatic fever.

Onset.—The illness commonly began with fleeting joint pains and malaise, and the patient was usually admitted to hospital within a week of the onset of symptoms (18 cases) but sometimes the onset was extremely insidious and prodromal symptoms might last for two weeks (1 case), three weeks (3 cases), four weeks (2 cases), or even five weeks (1 case). With this insidious type of onset, diagnostic difficulties were often encountered, and treatment had often been given for “sprained knee” or “flat feet” until the involvement of other joints or the clinical thermometer provided further evidence. In two cases erythema multiforme preceded the onset of pains in the joints. The following case illustrates this subacute type of onset.

Table 2

<table>
<thead>
<tr>
<th>Interval in weeks</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>2</td>
<td>6</td>
<td>8</td>
<td>4</td>
<td>2</td>
</tr>
</tbody>
</table>
ANNALS OF THE RHEUMATIC DISEASES

Case 1.—A trooper, aged 18 years, had tonsillitis seven weeks before admission. Three weeks after the tonsillitis he noticed aching and stiffness of the knees in the mornings, which wore off during the day. A few days later the right knee became sufficiently painful to prevent him from carrying out his duties. He was improved by a few days rest in bed, but after getting up he again began to complain of pain and swelling of the right knee and right ankle, although he is said to have been afebrile. Eventually the shoulders were also involved; he became feverish, and was admitted to hospital with a temperature of 101° F. and an erythrocyte sedimentation rate of 50 mm. in the first hour. When examined he had painful swelling of the right knee and ankle, and movements of the shoulders and lower spine were limited by pain. Temperature and pains in the joints subsided after four days on salicylates. Three weeks after admission the erythrocyte sedimentation rate was 10 mm. in the first hour, and he returned to his unit after five weeks in hospital.

Arthritis.—The story of pain and stiffness in the mornings was very characteristic of the milder arthralgias in this series. All grades of joint involvement were seen from subjective arthralgia to the classical hot, red, swollen and exquisitely painful joint. Seven patients had pains in the joints without demonstrable swelling. In two cases one knee only was involved, but the others all developed a migratory polyarthritis. The frequency with which different joints were involved is shown in Table 3, which gives an idea of the frequency with which the joints of the lower limb were affected.

Simple Type.—Fifteen cases could be classified under this heading. Fever was usually moderate and the temperature rarely exceeded 102° F. The pulse rate was not disproportionately raised, and rates of over 100 per minute were uncommon. Response to full doses of salicylates was prompt, and the patients were usually afebrile within three or four days. In most cases pains in the joints had subsided within ten days and the patients remained well thereafter.

Recurrents.—There was a group of cases showing delayed resolution. These patients were not the only cases in whom convalescence was for one reason or another prolonged, but they formed a sufficiently homogeneous group to justify description. The initial course of the disease was in no way remarkable, and all these patients appeared to be symptom-free within ten days of admission. A few days later, however, they would start to complain of a "second wave" of pains in the joints. These pains were less acute than those of the initial attack and were not accompanied by relapse of pyrexia, but they were far more persistent and symptomatic response to salicylates was less certain. Convalescence was often exceedingly prolonged and the erythrocyte sedimentation rate might remain elevated for two or three months. These cases showed some resemblance to Copeman's group of "rheumatic fever with chronic fibrositis" (1944) and would have been an interesting group to follow up, but most of them had to be evacuated to the United Kingdom for disposal. They were quite distinct from the "relapses", described below, and the following is an illustrative case.

Case 2.—A driver, aged 22, had had rheumatic fever at the ages of 17 and 21, and coryza one week before the illness here described. On June 22 he twisted his left knee, and next day had malaise, with both knees swollen and painful. On June 24 he was admitted to hospital with a temperature of 102° F. The left ankle and both knees were swollen, hot, and tender. There was rapid response to salicylates. By July 2 he was free from all symptoms, the erythrocyte sedimentation rate being 27 mm. in the first hour. On July 7 pain returned in the right shoulder and elbow. Low-grade fitting arthralgia persisted in spite of salicylates. On July 15 the erythrocyte sedimentation rate was 15 mm. in the first hour. On Aug. 5 there was slight residual morning stiffness of shoulders, knees, and fingers, the erythrocyte sedimentation rate being 3 mm. in the first hour. Thereafter he gradually recovered, and on Aug. 29 returned to his unit again and well.

Polycyclic Type.—There were two patients who had relapses when ambulant after all signs of activity had apparently subsided. These episodes were acute pyrexial episodes resembling the initial attacks, rather than the "second wave" type of arthralgia described above. In the following case the two relapses may have been precipitated by premature return to duty.

Case 3.—A private, aged 23, six weeks before admission had sore throat and febrile coryza lasting two weeks. On Dec. 16 he was admitted to hospital because of feverishness, painful swelling of left knee and ankle, and low-back pain for twenty-four hours. The temperature was 100° F., and the erythrocyte sedimentation rate 45 mm. in the first hour. The attack settled rapidly.

Table 3

<table>
<thead>
<tr>
<th>Joint</th>
<th>Pain only</th>
<th>Pain and swelling</th>
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<tbody>
<tr>
<td>Knees</td>
<td>24</td>
<td>18</td>
</tr>
<tr>
<td>Ankles</td>
<td>16</td>
<td></td>
</tr>
<tr>
<td>Shoulders</td>
<td>12</td>
<td>7</td>
</tr>
<tr>
<td>Metatarsals</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Wrists</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Fingers</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Back</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Hips</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Temperomandibular</td>
<td>1</td>
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with salicylates. On Dec. 29 he was up and well. The erythrocyte sedimentation rate was 3 mm. in the first hour. On Jan. 4 he returned to his unit, where he was sleeping under canvas. After a day or two he developed some stiffness of the lumbo-sacral region, and on Jan. 8 he was readmitted with the clinical picture of an acute lumbago with pain radiating down the backs of the thighs. No peripheral joints were affected. The temperature was 101° F., and the erythrocyte sedimentation rate 40 mm. in the first hour. He again made a rapid symptomatic recovery, the erythrocyte sedimentation rate fell to normal, and he was discharged to duty on Jan. 31. He remained well until Feb. 11, when he developed pain in the left hip and one superior tibiofibular joint was hot, red, and painful. Symptomatic recovery was again rapid, but this time he was sent away for prolonged convalescence.

Rheumatic Fever in Older Patients.—Among the older patients the onset of the disease tended to be more insidious and the course more prolonged as judged by duration of pains in the joints and the length of time during which the erythrocyte sedimentation rate remained elevated. Numbers are too small for statistical analysis, but the figures are suggestive. In four out of five patients over 30 the erythrocyte sedimentation rate remained elevated for more than thirty days, whereas in all but three of thirteen patients under 30 it had fallen to normal levels within this period. In three out of five patients over 30 prodromal symptoms had been present for two weeks before the patient was admitted to hospital, whereas in all but four of twenty patients under 30 the prodromal phase was less than two weeks. There was no correlation between these two factors (duration of prodromal symptoms and length of time during which the erythrocyte sedimentation rate was raised) when they were related to the series as a whole irrespective of age.

The Erythrocyte Sedimentation Rate

The erythrocyte sedimentation rate was estimated at approximately ten-day intervals in every case and was accepted as a rough guide to the activity of the disease. Initial levels varied from 10 to 50 mm. in the first hour (Westergren). In some cases the level rose after admission to hospital, although clinical evidence of activity had subsided. One such case is described below, and is of particular interest as he might never have been admitted to hospital had he not attended for a routine examination.

Case 4.—A signalman, aged 19 years, on March 7 attended for a routine check-up twenty-five days after an attack of tonsillitis. On being questioned he admitted he had vague morning pains in the knees for two or three days and some stiffness of the adductor muscles. He had no definite physical signs. The temperature was 99-4° F., and the erythrocyte sedimentation rate 6 mm. in the first hour. He was admitted to hospital for observation. The next day the temperature rose to 101° F. in the evening; he complained of pain in the right foot, and the medial aspect of the right tarsus was hot, red, swollen, and tender. On March 11 he was afebrile and free from all pain, but the erythrocyte sedimentation rate had risen to 21 mm. in the first hour. On March 20, when he was up and well and asking to be discharged from hospital, the erythrocyte sedimentation rate was 33 mm. in the first hour.

Complications

No electrocardiographs were available, but there was no clinical evidence of carditis in any case. One or two transient systolic murmurs were heard, but they were neither widely propagated nor associated with cardiac enlargement. In no case was there a diastolic murmur, cardiac enlargement, persistent tachycardia, or pericarditis. The only patient who came under suspicion was kept under observation for three months, at the end of which time his heart was clinically and radiologically normal apart from a soft systolic murmur.

Nodules, purpura, and pneumonitis were not seen in any case.

Discussion

There can be little doubt that a diagnosis of rheumatic fever was justified in these cases. The clinical picture merges imperceptibly into that of the classical type, and the history of a preceding infection could usually be obtained when sought for. This benign adult form of the disease is probably much commoner than is usually believed, and it has not received adequate recognition only because such cases are not usually admitted to hospital and the clinical picture may be more familiar to the general practitioner than to the consultant. While there would be little disagreement with the view that these patients should be given adequate doses of salicylates by mouth for at least two weeks, the extent to which activity should be limited is controversial. Once a diagnosis of rheumatic fever has been made, it is customary to order rest in bed until all signs of activity have subsided (Sokolow and Snell, 1947; Ferguson, 1943); and Rosenberg (1946) insists on a minimum of six weeks rest in bed. These authors find that electrocardiographic changes during the active phase are common even in mild cases, and Rantz and others (1945) sometimes found such changes in a subclinical "post-streptococcal state" after tonsillitis. However, it is a highly questionable procedure to stigmatize patients with the "cardiac" label on the basis of electrocardiographic changes alone, and it is more important to know the risk of clinically significant cardiac sequelae in these cases. It can hardly be
denied that valvular disease of the heart does occasionally occur after mild or even subclinical attacks of rheumatic fever, but the risk of significant cardiac involvement in the type of adult rheumatic fever described here is probably greatly overrated. Copeman (1944) states emphatically that cardiac sequelae did not occur in his "benign" group; and in the series described here, although early activity was permitted, there was no clinical evidence of cardiac involvement although a longer follow-up would have been desirable and five cases were lost sight of before the erythrocyte sedimentation rate had fallen to normal levels. De Liee and others (1943) found that persistent cardiac sequelae were observed in only 7 per cent. of patients over 25 with first attacks of rheumatic fever, and they were presumably cases of the classical type usually seen in civilian hospital practice. All authorities are agreed that the incidence of carditis is very much greater in the classical than in the commoner benign type of adult rheumatic fever. A common-sense attitude in the matter is essential. The risk of cardiac involvement in the type of case described here is slight, and there is little evidence that activity insufficient to provoke tachycardia or aching of the joints. The patient should, of course, remain under medical observation until all signs of activity have subsided.

Summary

A consecutive series of twenty-five cases of rheumatic fever admitted to the medical wards of a military hospital is described and analysed. The course of the disease was characteristically benign, although arthralgia was persistent in some cases. Cardiac complications were not observed.

The management of mild adult rheumatic fever is discussed.

REFERENCES


Rhumatisme Articulaire Aigu Atypique
Chez de Jeunes Adultes

RÉSUMÉ

L'auteur a décrit et analysé une série consécutive de vingt-cinq cas de rhumatisme articulaire aigu admis dans les salles de médecine d'un hôpital militaire. L'évolution de la maladie était typiquement bénigne, bien que l'arthralgie ait persisté dans quelques cas. On n'a pas observé de complications cardiaques.

Il discute la conduite à tenir dans la forme bénigne du rhumatisme articulaire aigu chez l'adulte.
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