such a defect ataxia is present, but formal cerebellar testing is normal because the control of the γ-system is intact (Kremer, 1958).

Summary

A case of pneumococcal meningoc-encephalitis is described where initial disability was great, yet recovery surprisingly complete, probably due to glucocorticoid therapy.

Confusion of the body image and ataxia persisted to a limited extent, and the mechanisms of these are discussed.

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PUSEO-RHEUMATIC LEUKAEMIA

A Case Report with Special Attention to the Influence of Treatment


Senior Medical Registrar, King George Hospital, Ilford, Essex

Acute leukaemia, particularly in children, may present in many guises. Skeletal involvement as a cause of pains in the limbs is well recognized. Less commonly polyarthritis occurs as a prominent clinical feature, and the illness can then mimic acute rheumatic fever or juvenile rheumatoid arthritis very closely. In several well-documented reports, emphasis has been laid on the ineffectiveness of salicylates in relieving symptoms, an observation which might lead to consideration of acute leukaemia as an alternative diagnosis in such patients. The case described now illustrates such a mode of presentation; in addition, attention is paid to the influence of treatment, mainly with a steroid preparation, on the subsequent course of the illness.

Case Report

In June 1959, a schoolboy aged 15 was referred to the London Hospital with the provisional diagnosis of juvenile rheumatoid arthritis. During the preceding three weeks he had complained of intermittent aching pain in both knees and mild pain and stiffness in the neck. The left knee had appeared swollen for a few days, some two weeks before his first visit to hospital. During this fortnight he suffered from general malaise, lethargy and anorexia. He had received aspirin and paracetamol without relief of his joint pains.

At the time of his first visit to hospital there were few abnormal features. He was a thin boy of normal development with mild pallor of mucous membranes. No adenopathy was detected and there was little objective change in the joints, apart from tenderness over the femoral condyles of the left knee. ESR (Westergren) 32 mm./hr., haemoglobin 64%, and radiological appearances of the chest and knee joints normal. Over a period of three weeks, while awaiting admission to hospital, he was given calcium aspirin 1.3 g. q.i.d. and ferrous sulphate 0.2 g. t.i.d. Flitting joint pains continued with increasing intensity and frequency and involved wrists and shoulders as well as knees. Short bouts of intense pain in the knees were particularly distressing at night. Malaise increased, he experienced night sweats and his weight fell.

On examination on entering hospital on July 5, 1959, he was febrile (100°F) and sweating; he looked pale and ill; there was tachycardia and a soft apical systolic murmur was audible. No enlargement of superficial lymph nodes was detectable; liver and spleen were not palpable. There was now objective evidence of abnormality in several joints. Movement was restricted by pain at the left elbow and right shoulder; both knees were warm and contained effusions. Three days after his arrival in hospital the tip of the spleen could be felt and was tender.

A clinical diagnosis of juvenile rheumatoid arthritis was still favoured. ESR was now 80 mm. and haemoglobin 60%. The latex agglutination test was negative. A routine total and differential white cell count revealed acute leukaemia as the true diagnosis. There were 39,400 wbc/cu. mm., of which 'blast cells' constituted 89%, polymorphs 9%, and lymphocytes 2%. Examination...
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symptom for several weeks in the early part of the illness and during the first relapse, they were overshadowed by the general physical deterioration in the last two months of life.

Autopsy. (Dr. P. G. I. Stovin): Several ecchymoses were noted on the skin and herpetic crusting was present about the mouth. Slight generalized enlargement of lymph nodes existed and the spleen (650 g.) weighed about three times normal. Periportal infiltration was visible in the liver; vertebral and femoral marrow looked red and gelatinous; broncho-pneumonic consolidation had occurred in the left lung. No abnormality was visible to the naked eye in synovial membranes of the right knee or hip joint and the cartilages looked normal. Microscopically the existence of leukemic infiltration was confirmed, but was sparse, and the enlargement of lymph nodes and spleen was in part the result of congestion. In the synovium of the right knee synovial cells were swollen and a few lymphocytes were noted. Cytomegalic inclusion virus disease was present in the lungs.

Discussion

The existence of a remarkably uniform clinical syndrome emerges from a study of previous reports on the character of the polyarthritis that is sometimes seen in patients with acute leukemia. Joint manifestations of this type rarely occur in leukemia other than the acute lymphatic form; the occurrence of arthritis may precede other evidence of illness by weeks or months; flitting polyarthralgia with affection of the larger joints of the limbs is typical; treatment for a presumed rheumatic condition has nearly always been given before the correct diagnosis has become established.

In 1913, Struha referred to cases of acute lymphatic leukemia which 'set in with drawing pains in the extremities or with pains and swelling in the joints similar to articular rheumatism'. Baldridge and Awe (1930) list rheumatic fever and Still's disease among the conditions for which acute lymphatic leukemia may be mistaken. They mention the examination of the synovial membrane from an affected joint in one of their patients; no pathology could be recognized. In the same year Seward (1930) reported the occurrence of severe flitting joint pains in a man aged 31 with lymphatic leukemia. Examination of the joints at autopsy of this patient also failed to reveal special pathology. In particular, there was no evidence of hemorrhage, the synovial membranes and articular surfaces of the bones appearing normal. Poynton and Lightwood (1932) described a girl aged 3 who had a seven-week history of flitting joint pains and proved to have acute lymphatic leukemia. Sutton and Bosworth (1934) observed recurrent joint symptoms over a period of six months in a child. During this time, which included two admissions to hospital, polyarthritis diverted attention from the true underlying condition of acute lymphatic leukemia. They stressed the importance of protracted polyarthritis without evidence of carditis as an observation which might have led to more serious doubt about the true existence of rheumatic fever. Conybeare (1936) recorded the details of a child of 8 who had a long pyrexial illness accompanied by
bone and joint pains. The clinical diagnosis was at first rheumatic fever and later Still's disease before the presence of acute leukaemia was recognized.

Several descriptions relating to joint manifestations in acute lymphatic leukaemia appear in the French literature. Debray, Michaux and Sinton (1931) mention a boy who for several weeks had joint pains, characterized by their abrupt onset and intense severity, by their fleeting character, and by their tendency to affect larger joints. Effusions developed in the knee joints in this patient and fluid aspirated from one joint was yellow, sterile and contained few cells. A further patient, a girl aged 7 reported by Debré, Lamy, Soulie and Gabriel (1936), complained of pains in shoulders, hips and left knee; she was mildly anaemic, and a soft systolic murmur was audible. After a month an epistaxis occurred and the spleen and liver were then found to be enlarged. The possibility of leukaemia was entertained although no abnormality existed in the peripheral white cell count. Migratory joint pains continued to involve the knees, hips, shoulders and wrists; the epiphyses adjoining the larger joints became tender; and flexion became limited at the right wrist. Examination of the marrow 3 months after the onset of joint symptoms confirmed earlier suspicions of acute leukaemia.

The recognition of joint manifestations as the main clinical feature in some patients with acute lymphatic leukaemia is made in more recent publications. Bichel (1948) reviewed 'arthralgic leukaemia' in children. Dresner (1950) stressed that rheumatic fever might be closely simulated and that these patients failed to respond to salicylates. Dameshek and Gunz (1958) and Hayhoe (1960), in their monographs, recognize the 'pseudo-rheumatic' presentation of lymphatic leukaemia that sometimes occurs. By contrast, leukaemia is rarely mentioned as a possible cause of polyarthritis in discussions on the differential diagnosis of the rheumatic diseases.

As examination of affected joints at postmortem has seldom revealed either gross or microscopical abnormality, it has been proposed that small areas of leukaemia infiltration in adjacent subperiosteal or bony tissue might be responsible for pain and effusions. In the present case joint symptoms subsided as soon as haematological remissions were induced either by steroid or cytotoxic treatment, perhaps suggesting that the drugs were acting on such areas of leukaemic infiltration. Such a process seems more likely than the coexistence of a separate disease such as rheumatic fever, Reiter's disease or gout as a cause of polyarthritis. Levels of serum uric acid were raised as high as 9.5 mg/ml in the present patient, but the rise occurred immediately after remissions had been entered, at times when the joint symptoms had disappeared.

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D. D. Gibbs

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