ORIGINAL PAPER 6

Polymyalgia rheumatica: a general practice experience

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SUMMARY. In the eight years from April 1974 to April 1982, 10 diagnoses of polymyalgia rheumatica were made in an urban general practice. The natural history of the condition, diagnostic problems and occasional unpredictability of outcome are discussed. It is suggested that general practice research may provide answers to some unsolved questions.

Introduction

POLYMYALGIA rheumatica was the name suggested by Barber (1957) for a condition known for many years under other names, for example senile rheumatic gout (Bruce, 1888). Accepted diagnostic criteria include pain and morning stiffness, chiefly in shoulders and neck but often in the pelvic girdle, occurring in patients almost always aged over 50 years, with an erythrocyte sedimentation rate (ESR) usually above 50 mm in one hour and showing a rapid response to treatment with steroids. Involvement of peripheral joints may occur. Fever, anorexia, weight loss, anaemia and depression are common. Liver function tests often show abnormality, with rises in alkaline phosphatase in 50 per cent of cases (Healey and Wilske, 1978) and in alpha and gamma globulins. Liver biopsy is unhelpful, while muscles also show no diagnostic change.

The cause of polymyalgia is unknown. Viral infection, an autoimmune process, an association with the hepatitis B antibody, and the keeping of pet birds are all among theories which have not been substantiated. Instances of familial aggregation have been quoted: Barber (1957) mentioned polymyalgia in two sisters, Hamrin (1972) recorded four pairs of siblings among 93 patients, and Nielson (1980) reported its occurrence in a husband and wife within three months of one another. Women are more often affected than men, the ratio in most studies being about four to one. Polymyalgia is rare in blacks and orientals.

There is a frequent but inconsistent relationship with giant-cell arteritis. This condition, also affecting the elderly, most often involves the cranial arteries, temporal arteritis being its commonest presentation. Headache, jaw claudication, and thickening, tenderness and redness of the affected arteries occur, with the risk of sudden blindness should the retinal arteries become involved. Less often, patients with giant-cell arteritis present with symptoms reflecting changes in other arteries causing aortic aneurysm, stroke, myocardial infarction or limb claudication. Constitutional symptoms similar to those of polymyalgia rheumatica are met with, anaemia and loss of weight being common. The ESR is high and the response to steroids dramatic, but the initial dose needed to control symptoms and prevent blindness is 40–60 mg daily, compared with the 15 mg generally adequate for polymyalgia. Biopsy of affected arteries shows destruction of the internal elastic lamina and infiltration of giant cells.

Like polymyalgia, giant-cell arteritis has been reported as affecting several individuals in the same family, sometimes in association with polymyalgia. Kvernebo and Brath (1980) described five members of one family who developed one illness or both within the space of eight years, and Liang and colleagues (1974) recorded four pairs of relatives similarly affected among 250 patients studied. The two conditions may present simultaneously but, in a minority of those with polymyalgia alone giant-cell arteritis follows despite apparent successful treatment, occasionally after a gap of years. Temporal artery biopsy in polymyalgia may reveal active symptomless arteritis, and this cannot be excluded even if the biopsy is negative, since the specimen may be taken from an unaffected segment of an artery inflamed elsewhere in its course.

Belief that polymyalgia rheumatica and giant-cell arteritis are variants of the same condition has led some writers to use the term polymyalgia arteritica to describe both (Paulley and Hughes, 1960). Another suggestion is that polymyalgia rheumatica is a syndrome and not an entity, and that it represents a response to a variety of

Details of the 10 patients with polymyalgia rheumatica.

Case number	Year of diagnosis	Age	Sex	Previous history	Results of initial tests: ESR (mm)	Hb (g/dl)	Liver function	Referred	Treatment
1	1974	66	F	Depression	94	Not recorded	Alkaline phosphatase↑	Yes	Analgesics
2	1976	70	M	Nephrectomy, glaucoma, angina, obstructive airways disease	56	9.5	Not recorded	No	Steroids
3	1977	63	F	Duodenal ulcer	73	12.2	Normal	Yes	Analgesics
4	1977	69	F	-	50	11.0	Not recorded	Yes	Steroids
5	1979	83	F	Congestive heart failure	104	11.4	Normal	Yes	Steroids
6	1979	74	F	Depression, osteoarthritis, ischaemic heart disease	94	13.3	Not recorded	No	Steroids
7	1980	55	F	Hypothyroidism (8 years)	46	12.0	-	Yes	Steroids
8	1980	79	F	Osteoarthritis, obesity	114	9.6	-	No	Steroids
9	1981	70	F	-	61	12.0	Aspartate transaminase↑	No	Steroids
10	1982	69	M	Diabetes (11 years)	84	14.0	Alkaline phosphatase↑ γ glutamyl transpeptidase↑	No	Steroids

stimuli (MacKenzie, 1969). Huskisson and colleagues (1977) described six cases of what they termed complicated polymyalgia: at the time polymyalgia was diagnosed, three of their patients were found to have malignant disease as well, another patient had infective endocarditis, one had rheumatoid arthritis and a sixth developed Hodgkin's disease two years later. Malignant disease has been reported in several other studies; thyroid disease, primary biliary cirrhosis and collagen vascular disorders are among other conditions that have occurred in association with polymyalgia.

The incidence of polymyalgia rheumatica is unknown. Dixon and colleagues (1966) described it as more common than ankylosing spondylitis, as common as gout and, in patients over 70 years of age, as common as rheumatoid arthritis. It seems likely that general practitioners caring for an ageing population will encounter it more often than in the past.

Case reports

In an urban general practice with a list size that increased from 8,000 to 11,000 between 1974 and 1982, 10 new cases of polymyalgia were diagnosed, eight of the patients being female. Only 8 per cent of practice patients were aged over 65 years, little more than half the national average.

Clinical features are shown in the table. Patients usually reported symptoms early, the average length of history at first attendance being only two to three weeks. The two histories which follow illustrate some of the problems of diagnosis and management.

Case 4

A woman aged 69 years presented with an effusion in her right knee and a weight loss of 4.5 kg. Shortly after, she developed persistent pain in her neck and shoulders. An orthopaedic surgeon diagnosed cervical spondylosis, but further loss of weight and a rise in the ESR from 50 to 102 mm led to referral to a physician. She had a normochromic normocytic anaemia, and a stool specimen was reported positive for occult blood. Results of barium meal, barium enema, sigmoidoscopy and intravenous pyelography were all normal. Laparotomy followed renewed surgical consultation, but nothing abnormal was found. Care of the patient reverted to the physician, but she remained unwell with pain successively in her shoulders, left foot and left ankle. Finally, two-and-a-half years after her illness began, she was referred to a rheumatologist and the diagnosis of polymyalgia was made.

Hindsight makes this problem much simpler than it appeared at the time.

Case 2

A man aged 70 years, blind owing to glaucoma, had a history of ischaemic heart disease and obstructive airways disease, and had also had a nephrectomy for renal calculus. His new symptoms of shoulder pain and stiffness led to prompt diagnosis, and steroids produced swift relief. Four months later he became diabetic and also developed herpes zoster. Just over a year after his first symptoms he had a deep venous thrombosis in his right calf, followed after admission to hospital by a pulmonary embolus and a haematemesis from which he died. Post-mortem examination revealed a carcinoma of stomach.

This is the only instance of complicated polymyalgia in this study.

Other features

Nine patients complained of pain in shoulders or hips or both areas; the tenth patient had upper chest pain and,

like all the others, severe morning stiffness. Four patients had pain in peripheral joints and two had headache, though none showed overt evidence of giant-cell arteritis. Two cases of temporal arteritis were identified in the practice during the eight years of the study.

Five patients were referred for consultant opinion because of diagnostic uncertainty.

Two patients (cases 1 and 3) were not treated with steroids, because in both there was initial doubt about the diagnosis and later spontaneous improvement. Of the eight patients treated with steroids, two developed diabetes, in case 6 four weeks and in case 4 four months after starting treatment. Herpes zoster occurred during treatment in three cases, in case 2 after four months of treatment and in both cases 1 and 6 after nearly two years of steroids. Two patients had had herpes zoster preceding polymyalgia, in case 6 three months and in case 7 18 months before the major illness.

The only patient known to have died was case 2, but two patients (cases 4 and 8) moved away while still under treatment and were lost to follow-up. Steroids were stopped after one year in case 7, but have been continued in the other cases. Three patients relapsed on treatment, in case 9 three months, in case 5 eight months and in case 6 one year after treatment had started, relapses being characterized by a return of symptoms, a rise in the ESR and a response to increased steroid dosage.

Discussion

The diagnosis of polymyalgia rheumatica is not always easy. Coomes and colleagues (1976) found that in only 4 per cent of patients with polymyalgia seen at two London hospitals between 1964 and 1969 had the condition been diagnosed correctly by the referring doctor, but the success rate rose to 10 per cent of patients between 1970 and 1974. Jones and Hazleman (1981a) reported correct diagnosis before referral in 39 per cent of patients with polymyalgia or giant-cell arteritis seen in hospital at Cambridge between 1974 and 1979. These studies suggest that general practitioners are becoming more familiar with the clinical picture of polymyalgia, but neither study reveals how often the illness was managed without referral. Often elderly patients are already suffering from osteoarthritis, depression and other chronic illness which may cause a change in symptoms to be overlooked. In the present study five of the 10 cases of polymyalgia were diagnosed without referral, four of them rapidly and the fifth after three months of uncertainty. Of the five patients referred to hospital, in one patient (case 7) the condition was diagnosed on her first outpatient attendance; the diagnosis had been considered by her family doctor but rejected as her age (55 years) was thought to be against it and her ESR at 46 mm was the lowest of the group. A second patient (case 5) was admitted for investigation before the diagnosis became clear. Two other patients (cases 1 and 3) were thought for several months to be suffering from seronegative arthritis, while case 4, described already, was not diagnosed for two-and-a-half years.

There are still some unknown factors in the natural history of polymyalgia. Identification of co-existing giant-cell arteritis is not possible unless there are local signs or a positive biopsy is obtained. Beevers and colleagues (1973) reported an instance of blindness in a patient treated for six months with 15 mg prednisone daily. Rynes and colleagues (1977) described the onset of temporal arteritis in a patient after treatment for a year with prednisone. Many similar cases have been reported in other studies, while relapses during treatment are common.

There is little argument that steroids should be given as soon as the diagnosis is made, for although polymyalgia responds to salicylates and to non-steroidal anti-inflammatory drugs, the relief after taking steroids is rapid and complete. Steroids probably brought about the development of diabetes in two study patients; in both patients it was controlled by diet and hypoglycaemic drugs. Herpes zoster during treatment presumably reflected an altered immunological state; Coomes and colleagues (1976) reported five cases among 102 patients with polymyalgia. The duration of steroid treatment is in addition uncertain. Earlier writers (Russell, 1959; Meadows, 1966) suggested that six months was adequate. Fauchald and colleagues (1972) recommended a period of two years. Coomes and colleagues (1976) found that 84 per cent of their patients still required steroids after five years, while Beevers and colleagues (1973) believed that a small permanent maintenance dose was advisable. The unpredictability of the outcome was shown by Jones and Hazleman (1981b), one of whose patients developed a hemiplegia and blurred vision six months after stopping steroids taken continuously for eight-and-a-half years. While it seems that some patients will remain well indefinitely if steroids are withdrawn after one or two years, there is no means of identifying those patients in whom it will be unsafe to stop treatment, and the possibility of relapse or of dangerous complications cannot be ruled out. It is likely that these questions will be solved only by prolonged and detailed recording and follow-up. This research should be carried out in the community, since hospital studies depend upon referred cases, which may not be typical.

No general practitioner can expect to encounter many cases of polymyalgia in one practice, but careful search among ailing elderly patients may increase the yield. Sustained and detailed study by groups of general practitioners pursuing a common interest might result in a better understanding of a mysterious and fascinating condition which is enormously satisfying to treat.

THE INFLUENCE OF TRAINERS ON TRAINEES IN GENERAL PRACTICE

Occasional Paper 21

The latest Occasional Paper on vocational training reports on the educational progress of a group of trainees in the North of England. Two groups of trainees were identified, those who underwent the greatest change and those who underwent the least change precourse to postcourse, and their characteristics were compared with the characteristics of their trainers. This is the first time this has been done and several new findings have emerged.

These findings are fully consistent with those of Occasional Paper 18 and add still further support for the present system of selecting training practices. The report will therefore need to be considered by regional general practice subcommittees, course organizers, and regional advisers, and is recommended to all trainers and trainees.

The Influence of Trainers on Trainees in General Practice, Occasional Paper 21, can be obtained, price £3.25 including postage, from the Publications Sales Department, Royal College of General Practitioners, 14 Princes Gate, Hyde Park, London SW7 1PU. Payment should be made with order.

MEMBERS' REFERENCE BOOK

The College has completely changed the format of the old *Annual Report* and has now produced a new reference book for members, which will appear annually. This contains not only the full report of Council, reports from faculties and financial accounts as before, but also for the first time the names and addresses of half the membership of the College. The second half will follow next year, and so a complete register will be available every two years.

The Reference Book also includes lists of regional advisers, course organizers, College tutors and faculty secretaries, and a number of special features such as a list of College policy statements. Current activities of the College are described and illustrated by photographs of places and personalities. In addition, the book contains a great many articles about different aspects of general practice written by well-known authorities in the field.

This large volume, comprising 450 pages, provides an invaluable source of information for all general practitioners. Copies can be obtained from the Publications Sales Department, Royal College of General Practitioners, 14 Princes Gate, Hyde Park, London SW7 1PU, price £17.50 including postage. Payment should be made with order.

References

Barber, H. S. (1957). Myalgic syndrome with constitutional effects: polymyalgia rheumatica. *Annals of Rheumatic Disease,* **16,** 230-237.

Beevers, D. G., Harpur, J. & Turk K. A. D. (1973). Giant-cell arteritis—the need for prolonged treatment. *Journal of Chronic Diseases,* **26,** 571-584.

Bruce, W. (1888). Senile rheumatic gout. *British Medical Journal,* **2,** 811.

Coomes, E. N., Ellis, R. M. & Kay, A. G. (1976). A prospective study of 102 patients with the polymyalgia rheumatica syndrome. *Rheumatology and Rehabilitation,* **15,** 270-279.

Dixon, A. S., Beardwell, C., Kay, A. *et al* (1966). Polymyalgia rheumatica and temporal arteritis. *Annals of Rheumatic Disease,* **25,** 203-208.

Fauchald, P., Rygvold, O. & Oystese, B. (1972). Temporal arteritis and polymyalgia rheumatica. Clinical and biopsy findings. *Annals of Internal Medicine,* **77,** 845-852.

Hamrin, B. (1972). Polymyalgia arteritica. *Acta Medica Scandinavica,* Supplement S533.

Healey, L. A. & Wilske, K. R. (1978). *The Systemic Manifestations of Temporal Arteritis.* London: Grune & Stratton.

Huskisson, E. C., Dieppe, P. A. & Balme, H. W. (1977). Complicated polymyalgia. *British Medical Journal,* **2,** 1459.

Jones, J. G. & Hazleman, B. L. (1981a). Polymyalgia rheumatica and giant-cell arteritis— a difficult diagnosis. *Journal of the Royal College of General Practitioners,* **31,** 283-289.

Jones, J. G. & Hazleman, B. L. (1981b). Prognosis and management of polymyalgia rheumatica. *Annals of Rheumatic Disease,* **40,** 1-5.

Kvernebo, K. & Brath, H. K. (1980). Polymyalgia arteritica: a report on five cases within a family. *Scandinavian Journal of Rheumatology,* **9,** 187-189.

Liang, G. C., Simkin, P. A., Hunder, G. G. *et al.* (1974). Familial aggregation of polymyalgia rheumatica and giant-cell arteritis. *Arthritis and Rheumatism,* **17,** 19-34.

MacKenzie, A. H. (1969). The polymyalgia rheumatica syndrome. *Geriatrics,* **24,** 158-164.

Meadows, S. P. (1966). Temporal or giant-cell arteritis. *Proceedings of the Royal Society of Medicine,* **59,** 329-333.

Nielson, J. L. (1980). Polymyalgia rheumatica in a husband and wife. *Scandinavian Journal of Rheumatology,* **9,** 177-178.

Paulley, J. W. & Hughes, J. P. (1960). Giant-cell arteritis of the aged. *British Medical Journal,* **2,** 1562-1567.

Russell, R. W. R. (1959). Giant-cell arteritis. *Quarterly Journal of Medicine,* **28,** 471.

Rynes, M. I., Mika, P. A. & Bartholomew, L. E. (1977). Development of giant-cell (temporal) arteritis in a patient 'adequately' treated for polymyalgia rheumatica. *Annals of Rheumatic Disease,* **36,** 88-90.

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Addendum

Since this paper was written a second patient (case 1) has died, eight years after the diagnosis of polymyalgia was made. The cause of death was a ruptured dissecting aneurysm of the thoracic aorta. She was not treated with steroids and had been free of symptoms for nearly seven years.