POLYMYALGIA RHEUMATICA AND TEMPORAL ARTERITIS*

BY

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There is a fairly common form of rheumatism which afflicts the elderly with pains in the head, neck, back, and proximal parts of the limbs, with stiffness of movement and with constitutional illness. It is associated with a raised sedimentation rate (E.S.R.) and a mild hypergammaglobulinaemia, and women are affected more frequently than men. There is as yet no specific test for this condition, but the rapid response to corticosteroid therapy, combined with the negative results from investigations to detect other diseases, provide a reproducible diagnostic situation which has been called "polymyalgia rheumatica" (Barber, 1957).

Two schools of thought exist. One regards polymyalgia rheumatica as a variant or prodromal phase of temporal arteritis. The other regards it as an entity in its own right linked, perhaps, to rheumatoid arthritis (Kersley, 1951) and various suitable names have been proposed, *e.g.* "anarthritic rheumatoid disease" (Bagratuni, 1953, 1956) or "pseudo-spondylite rhizomélisque" (Forestier and Certonciny, 1953). Gordon, Rennie, and Branwood (1964) presented biopsy studies on six cases of polymyalgia rheumatica showing evidence of low-grade chronic inflammatory changes around the shoulder joint and the fibrous supporting tissues of the shoulder girdle. Other students of polymyalgia rheumatica have not found clinical signs of temporal arteritis (Barber, 1957; Gordon, 1960; Boyle and Beatty, 1961; Chalmers, Alexander, and Duthie, 1964; Tweed, 1965; Andrews, 1965), but temporal arterial biopsies were not done routinely in their series.

Those authors who have linked polymyalgia rheumatica to temporal arteritis, also called cranial or giant cell arteritis (Paulley and Hughes, 1960; Russell, 1959) have supported this concept with arterial biopsies (see also Alestig and Barr, 1963; Hamrin, Jonsson, and Landberg, 1964) and with

angiography (Hamrin, Jonsson, and Landberg, 1965). The names "senile arteritis" and "polymyalgia arteritica" have been proposed on the basis of their studies.

Present Investigations

The present study concerns all new out-patients seen by the authors in the study period January, 1963, to April, 1965, in whom the diagnoses of polymyalgia rheumatica or temporal arteritis were made. All out-patients have routine estimations of the erythrocyte sedimentation rate (Westergren method) at the first attendance. If the rate is raised, the estimation is repeated at the next visit. All patients whose E.S.R. was found to be greater than 40 mm. in one hour and who were suffering from central pain and stiffness were tentatively labelled as "polymyalgia rheumatica" and were admitted for investigation. If no other disease was discovered, the diagnosis was considered to be confirmed. All such patients were submitted to temporal artery biopsy.

Thirty biopsies were performed. One was on a patient who subsequently developed rheumatoid arthritis and has been omitted from this series. Of the remainder, eighteen biopsies were normal or showed sclerotic changes consistent with the patient's age, and ten were positive for granulomatous arteritis. In one patient the temporal artery could not be found at biopsy, but as no pulsation could be detected clinically in either temporal artery and as she had complained of temporal scalp pain, she has been included in the biopsy-positive group for purposes of comparison. We have also included for purposes of comparison two patients who attended the out-patient department with proved temporal arteritis which had been diagnosed before the study period began. As proof of temporal arteritis we accepted a biopsy showing granulomatous inflammation of the artery wall; not all biopsies showed giant cells, but this is not essential for this diagnosis.

During the study period, we saw 2,222 new

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patients, so that the overall clinic incidence is 1·3 per cent.* About half this total was seen in St. Mary Abbots Hospital, where the number of referrals for rheumatoid arthritis at all ages during the study period was 150 and for polymyalgia or temporal arteritis thirteen, a ratio of roughly twelve to one. If the comparison is limited to those aged 70 years or above, then polymyalgia and temporal arteritis were about half as common as rheumatoid arthritis. Irrespective of age, polymyalgia and temporal arteritis were more common than ankylosing spondylitis and about as common as gout. Our out-patients are drawn from an area of West London where the preponderance of women over 60 is much higher than the national average. This area is relatively generously supplied with active rheumatology departments which do not have excessively long waiting lists. These seem to be the major local selection factors which help determine the clinic incidence of a relatively benign condition affecting elderly women.

Comparison of Biopsy-negative and Biopsy-positive Cases

All patients were over 50 years of age, the eldest being 92. For purposes of analysis they have been divided into biopsy-negative and biopsy-positive groups. The two groups did not differ significantly with regard to the number of women, the age at onset, or the duration of the disease before treatment (Table I). Nor did they differ in the values for the E.S.R. at first attendance. Although an arbitrary lower limit of 40 mm. in one hour has been chosen, most values were over 80 mm. in one hour, there being no tendency for high values to concentrate in either group. Since these patients were referred by their doctors to rheumatism departments they were

all complaining of rheumatic pains of some sort. Central pain and stiffness were dominant symptoms in eleven out of the thirteen biopsy-positive cases and in fourteen out of the eighteen biopsy-negative cases. We were unable to distinguish anything about these pains which would have been of help in predicting the outcome of the temporal artery biopsy. "Severe morning stiffness", i.e. sufficient to stop the patient from carrying on her normal activities unaided, was a frequent symptom. One of our patients, biopsy-negative, was so stiff as to be unable to get out of bed in the morning to answer the telephone, or to open the door to the doctor. By 11 am. she could manage fairly normally. Another patient, biopsy-positive, could not get out of bed without her husband's assistance and had to go downstairs by sitting and sliding from step to step. Morning stiffness of this order was present in seven of the biopsy-negative and in three of the biopsy-positive group. Specific complaints of pains in the head and of scalp tenderness had some predictive value, since they were recorded in twelve of the thirteen patients with positive biopsies (Table II), but in only four of the eighteen patients with negative biopsies. Other symptoms and signs specifically related to the cranial arteries were, as might be expected, more prominent in the biopsy-positive group (Table III). Thus three

TABLE I
CLINICAL PARTICULARS

Temporal Artery Biopsy				Positive	Negative
Total Patients				13	18
Sex	Male			1	4
	Female			12	14
Age (yrs)	50-59			0	5
	60-69			5	6
	70-79			7	6
	80-89			0	1
	90-99			1	0
Duration before Treatment (mths)	Range			1-18	2-24
	Average			5	7

*Other physicians work in the rheumatism departments concerned. The numbers of new patients seen in the departments is, of course, larger.

TABLE II
PAIN AND STIFFNESS

Temporal Artery Biopsy		Positive	Negative
Total Patients		13	18
Symptoms	Spinal and proximal limb pain	11	14
	Severe morning stiffness	3	7
	Head pain and scalp tenderness	12	4

TABLE III
SYMPTOMS AND SIGNS RELATED TO CRANIAL ARTERIES

Temporal Artery Biopsy		Positive	Negative
Total Patients		13	18
Visual Changes	Retinal artery block	3	0
	Minor visual symptoms	4	2
Temporal Arteries	Pulsation present	3	1
	Artery thickened and tender		
	Pulsation Absent in both arteries	6	0
	Absent in branch	2	1
Excessive Carotid Sinus Sensitivity		4	1
Claudication of the Jaw		1	1

of the latter showed occlusion of the central artery of the retina (bilateral in two, unilateral in one) and this had not occurred in the biopsy-negative group. Minor visual disturbances (difficulty in focusing, mistiness of vision, flashes of light, etc.) occurred in both groups.

The temporal arteries were carefully palpated in all patients, the most frequent abnormality found being absence of pulsation without enlargement of the artery. Nodular thickening of the artery was present in three patients with a positive biopsy and simple thickening in one with a negative biopsy. Two patients with positive biopsies had temporal arteries which had been completely normal on clinical examination. In one of these the only pointer towards arterial disease was marked sensitivity of the carotid sinus, defined as a systole of longer than two beats following pressure on the carotid sinus. The diagnostic significance of absence of pulsation in a temporal artery and of undue sensitivity of the carotid sinus depend on the frequency of occurrence of these signs in normal and diseased control populations. Two of us (Y.T.W. and A.S.D.) examined 48 elderly normal men, 163 elderly normal women, and 3 men and 54 women with advanced rheumatoid arthritis. In a preliminary training period observations were made on forty patients by two observers independently, the difference between observers being checked by further estimations together until a reproducible standard had been achieved. Observations were also made on the frequency with which systolic bruits could be heard over the common carotid arteries in these control subjects (Table IV). Pulsation was palpable in all control subjects up to and beyond the bifurcation of the temporal artery, except in one patient with rheumatoid arthritis who had been treated with corticosteroids and in whom gross "mooning" with thickening of the tissues made it

impossible to palpate the temporal artery. Carotid sinus sensitivity was occasionally found in the older control subjects and was more frequent with advancing age. It was, however, uncommon, affecting only fourteen of the control arteries examined (2.8 per cent.) and being usually unilateral, as opposed to four out of thirteen (31 per cent.) of the patients with positive temporal artery biopsies (usually bilateral). The presence of arterial bruits has been suggested by Hamrin and others (1965) as having diagnostic value. Arterial bruits were found in five of the 235 right carotid arteries and ten of the 235 left carotid arteries in the controls. There were no untoward effects from pressing on the carotid sinuses in these control subjects.

Of the eight patients with absence of pulsation in the temporal arteries, none gave a history of a previous phase of nodular enlargement of these arteries.

Case Reports

One patient illustrated the change from polymyalgia to temporal arteritis.

A married woman, a retired publisher aged 70, had had central pain and stiffness and general illness for one month before attending the clinic. She was found to have an elevated E.S.R. After a period of 3 months' treatment with analgesics and physiotherapy she had made no improvement and was admitted to hospital. Full investigation disclosed no underlying disease and the temporal arteries were normal to palpation. She was accordingly labelled as "polymyalgia rheumatica" and was treated with intramuscular corticotrophin, 15 units per day, with rapid improvement; 5 months later she accidentally reduced the dose and found that she developed headaches, difficulty in focusing, and tenderness of the jaws. This lasted for 2 weeks before she was seen again when examination disclosed absence of pulsation in both temporal arteries. A biopsy showed classical giant-cell arteritis.

TABLE IV
NUMBER OF CAROTID ARTERIES TESTED

Age (yrs)	Normal		Rheumatoid Arthritis		Total	Showing Asystole >2 beats on Compression	
	Male	Female	Male	Female		No.	Percentage
-60	8	0	1	13	22	0	0
-70	30	21	1	21	73	2	2.7
-80	40	107	1	15	163	2	1.2
-90	18	147	0	5	170	7	4.1
-100	0	41	0	1	42	3	7
All Ages	96	316	3	55	470	14	2.8

Other patients have not been so fortunate.

An unmarried woman, a retired shop assistant aged 69, had been previously seen by an experienced specialist in rheumatic diseases for her complaint of pain in the neck, arms, hips, and knees. No definite diagnosis was made and the patient was given symptomatic physiotherapy. However, she seemed generally unwell and her doctor referred her to an out-patient department of general medicine where she was admitted to hospital for further investigation. Again no diagnosis was achieved and she was transferred to a convalescent home. The E.S.R. was not estimated in either department. In the convalescent home she developed sudden complete blindness, and investigations at this stage revealed an E.S.R. of 120 mm./hour and temporal arteritis with central retinal occlusion. She improved with corticosteroid therapy but her sight did not return. It is difficult not to believe that a routine E.S.R. would have alerted medical attention to the problem sooner and in time to save her sight.

Arthritis.—We have not been impressed with the frequency of arthritis in this group.

Only one patient with polymyalgia developed definite arthritis. This was a woman aged 54 (biopsy-negative) who had been maintained on simple analgesics for severe stiffness of the limbs for 2 years. (Prednisolone had been given briefly at the onset of her illness but had produced severe dyspepsia and had not been tried again.) She developed a tense swelling of the left knee, and painful limitation of the left gleno-humeral joint. She continued up and about despite this, and then developed an acute rupture of the left knee into the calf, for which she was admitted to hospital. Fluid withdrawn from the knee was sterile, turbid, yellow, and of low viscosity, *i.e.* resembling that of rheumatoid arthritis. Similar fluid was withdrawn from the calf. Treatment with corticotrophin, 20 units per day, was started, with rapid disappearance of the arthritis, which has not recurred in 3 years of follow-up, although she still requires adrenal stimulation therapy to be maintained free from severe stiffness. Many sheep cell and latex tests for rheumatoid factor have been negative.

One other patient (biopsy-positive) complained of a painful shoulder and showed tenderness but no swelling of the left acromioclavicular joint. Injection of this joint with hydrocortisone relieved the pain, which has not since recurred.

Patients initially presenting to us with polyarthritis, high E.S.R., and negative tests for rheumatoid factor have been diagnosed as "atypical polyarthritis" and are not included in this study. No such patient has subsequently developed signs of temporal arteritis.

Seasonal Incidence.—Kinmont and McCallum (1965), following up an observed association between light sensitivity, dermatitis, and cranial arteritis, found that there was an association between the incidence of onset of the acute phase of temporal arteritis and the season, and more specifically with the hours of sunshine. We found no correlation between month or season, either with the date of onset of polymyalgic symptoms, or with the date of onset of cranial arteritis, and none of our cases has shown a light-sensitivity dermatitis.

Treatment

An attempt was made to treat as many as possible of these patients with corticotrophin rather than with oral corticosteroids, following the reports of the West London (Savage, 1962) and Sheffield (West, 1962) groups that this was less addictive and less liable to lead to complications than oral corticosteroids. However, this has not been very successful, and of twelve patients started on corticotrophin only five continue with this treatment, six have had to change to prednisolone because of poor control or difficulty with injections, and only one has gone into remission. Of the five patients who experienced serious complications on this treatment, four showed fluid retention with some degree of heart failure and one developed diabetes. In contrast, of fifteen patients started on prednisolone treatment, eleven are continuing with this and four have gone into remission. Four other patients (with polymyalgia rheumatica) have been treated with simple analgesics. Patients were not, however, randomly allotted to one or other treatment.

Prognosis and Duration of Disease

Eight of the biopsy-positive and fifteen of the biopsy-negative groups were still "active", requiring treatment as judged by return of symptoms and raised E.S.R. if the dose of corticosteroid was reduced (Table V). The duration of disease to date

TABLE V
DURATION AND PROGNOSIS

Temporal Artery Biopsy		Positive	Negative
Number of Cases still "Active"		8	15
Duration (mths)	Mean Range	15.0 7-31	13.8 2-32
Number Recovered		2	3
Number Dead		3	0
Duration (to death or recovery)	Mean Range	14.0 6-19	10.7 7-15

is much the same for both groups. Three patients, biopsy-positive, have died (of heart failure, pneumonia, and suicide respectively) and in two of these the disease was "active" at the time of death. Only five have so far gone into remission. A "polymyalgia" clinic has been set up and patients attend every 2 or 4 weeks for adjustment of the steroid dosage, which is kept as low as is consistent with freedom from symptoms and maintenance of an E.S.R. below 20 mm./hour.

Discussion

This study has confirmed the high incidence of histological evidence of temporal arteritis in patients presenting with polymyalgia rheumatica (Paulley and Hughes, 1960; Alestig and Barr, 1963; Hamrin and others, 1964, 1965). It also demonstrates that temporal arteritis cannot be excluded in such cases on clinical grounds alone. In contrast to standard texts (*e.g.* Beeson and McDermott, 1961), which suggest that irregular cord-like thickening or painful nodular enlargement of the temporal arteries are characteristic signs of temporal arteritis, absence of pulsation and difficulty in palpating the temporal arteries were the most frequent arterial signs in our patients with positive biopsies. Undue sensitivity of the carotid sinus to pressure was also found to be a useful sign of temporal arteritis. That these were valid signs was shown by reference to a control series of subjects with advanced rheumatoid arthritis and elderly normal subjects.

Apart from signs and symptoms related to cranial artery involvement, there was nothing in the age, sex, duration of disease, or response to treatment which would have differentiated patients with "polymyalgia rheumatica" without a positive temporal artery biopsy from those with a positive biopsy.

Polymyalgia rheumatica is probably a common rheumatic disease, about as common as gout and ankylosing spondylitis and nearly as common as rheumatoid arthritis in the older age groups, judging from the number of cases which various observers have been able to assemble in relatively short periods. Just how common it is cannot at present be estimated, since this disease, although reasonably well defined, does not appear in surveys of rheumatic complaints in the population (*e.g.* Kellgren, Lawrence, and Aitken-Swan, 1953). Nor is it listed in the classification of rheumatic diseases published by the American Rheumatism Association (Blumberg, Bunim, Calkins, Pirani, and Zvaifler, 1964). The topic has been the subject of numerous reports in the British and Continental European literature, but

few observers or major rheumatism centres in the New World have yet commented upon it, suggesting regional differences in incidence.

Summary

In 29 patients with "polymyalgia rheumatica" (defined as central pains and stiffness and a raised E.S.R.), a temporal artery biopsy was positive for temporal arteritis in ten and normal in eighteen; in one subject the artery could not be found. Absence of temporal artery pulsation and undue sensitivity of the carotid sinus to pressure were useful signs predicting a positive biopsy. These signs were validated by reference to control populations of 57 patients with advanced rheumatoid arthritis and 211 normal elderly men and women. A comparison of patients in whom the biopsy was positive with those in whom it was negative showed no difference with regard to sex, age at onset, duration of disease, or response to treatment, although there were differences in symptoms corresponding to involvement of the cranial arteries. It seems likely that polymyalgia rheumatica and temporal arteritis constitute a single entity which is commoner than is generally recognized.

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La périarthrite rhizomélisque et l'artérite temporale

RÉSUMÉ

Parmi 29 malades atteints de périarthrite rhizomélisque (*polymyalgia rheumatica*, caractérisée par des douleurs centrales, des enraidissements et par la vitesse de sédimentation globulaire élevée) une biopsie de l'artère temporale révéla l'existence d'une artérite temporale chez 10 d'entre eux et une structure normale chez 18 d'entre eux; chez un malade on ne réussit pas à trouver l'artère. L'absence du pouls de l'artère temporale et une sensibilité exagérée du sinus carotidien furent des signes utiles pour prédire une biopsie positive. La valeur de ces signes fut confirmée chez des témoins: 57 malades atteints d'arthrite rhumatismale avancée et 211 hommes et femmes normaux

d'âge avancé. Une comparaison des sujets à biopsie positive avec ceux à biopsie négative ne révéla aucune différence en ce qui concerne le sexe, l'âge du début de la maladie, la durée de la maladie ou l'effet du traitement, bien qu'il y eût des différences dans la symptomatologie correspondant à l'implication des artères crâniennes. Il est bien possible que la périarthrite rhizomélisque et l'artérite temporale constituent une seule entité, plus fréquente qu'on ne le croit.

La polimialgia reumática y la arteritis temporal

SUMARIO

Entre 29 enfermos con *polymyalgia rheumatica* (caracterizada por dolores centrales, rigidez y velocidad de sedimentación eritrocitaria elevada) la biopsia de la arteria temporal reveló la presencia de la arteritis temporal en 10 de ellos y una estructura normal en 18; en un enfermo no se pudo hallar la arteria. La ausencia del pulso temporal y la sensibilidad exagerada del seno carotideo permitian anticipar una biopsia positiva. La validez de estos signos fué confirmada en grupos de testigos: 57 enfermos con artritis reumatoide avanzada y 211 hombres y mujeres normales de edad avanzada. La comparación de los sujetos con biopsia positiva con los demás no reveló diferencia alguna respecto al sexo, la edad del comienzo de la enfermedad, la duración de la enfermedad o el efecto del tratamiento, pero se vieron diferencias en la sintomatología derivada de la implicación de los nervios craneales. Es pues probable que la polimialgia reumática y la arteritis temporal formen una sola entidad, más frecuente de lo que se realiza generalmente.