Giant cell arteritis

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Summary

Giant cell (senile, temporal or cranial) arteritis occurs three times more frequently in women and the majority of patients are over 70 years of age. This series of forty-seven patients presented with visual symptoms and most also had a constitutional upset. The ocular signs were of ischaemic papillopathy (in 80%), which was unilateral in 61% and bilateral in 17%; or central retinal artery occlusion (13%). The ESR was always elevated above 40 mm, and in 28% it was above 100 mm. The main ophthalmic differential diagnosis was to determine whether it was arteritic or atherosclerotic ischaemic papillopathy in this age group, and principal differences are outlined (Table 6). Following corticosteroid therapy, vision improved in 23%, remained unchanged in 64% and deteriorated in 13% of patients. Visual deterioration predominated in elderly women who received steroids late in the course of the disorder. Early steroid therapy is recommended in elderly patients with visual symptoms and significantly raised ESRs.

During the first 10 years of a new Medical Ophthalmology Unit (1963-1972), there were 3200 admissions, of whom forty-seven patients had giant cell arteritis. This report analyses the clinical features, course and management of this personally investigated and treated series. We have used the more comprehensive term giant cell, rather than temporal or cranial arteritis, since it expresses the basic histology. The term temporal or cranial arteritis is too narrow a description for a disorder which causes widespread damage to many arteries of the upper half of the body in addition to the temporal or cranial arteries. The alternative term is senile arteritis for it affects those over 60 years of age.

Patients and methods

Fifty-eight patients were admitted to the Royal Eye Hospital Medical Ophthalmology Unit, located at Lambeth Hospital, with clinical features which suggested a diagnosis of giant cell arteritis but subsequent investigations resulted in a change of diagnosis in eleven patients, so this series comprises

forty-seven hospitalized patients. The eleven patients who were initially diagnosed as giant cell arteritis but subsequently recategorized, constitute the differential diagnosis and are analysed under this section.

On admission, all patients were examined by an ophthalmic surgeon and a physician interested in inflammatory eye disease. They were intensively investigated by routine blood count, ESR, blood biochemistry, serological antibody tests, and X-rays of chest, spine and skull, similar in many respects to those already outlined elsewhere for the investigation of uveitis patients (James *et al.*, 1969). In addition, temporal artery biopsy was undertaken in thirty-one of forty-seven (66%) patients.

Results

Sex and age

There were thirty-five females and twelve males, all of whom at the time of presentation were over 60 years of age and three-quarters were over 70 years (Table 1).

TABLE 1. Age-groups at onset of forty-seven patients with giant cell arteritis

Age (years)	No.	%
Under 60	0	
60-69	11	24
70-79	25	76
80+	11	
Total	47	100

Clinical presentations

The most frequent general symptoms were headaches in two-fifths, hyperalgesia of the scalp in one-quarter and pain on chewing or neck stiffness in one-fifth of patients (Table 2). Anorexia or weight loss were also significantly common, certainly more often than polymyalgia rheumatica. Associated with these general symptoms were visual symptoms in all but one patient. Two-thirds of patients complained of loss of vision, which was unilateral twice as often as it was bilateral (Table 3). Blurred vision

TABLE 2. Predominant presenting symptoms of forty-seven patients with giant cell arteritis

Symptom	No.	%
Headache	19	40
Hyperalgesia of scalp	13	28
Pain on chewing	10	21
Neck stiffness	10	21
Weight loss	8	17
Anorexia	7	15
Polymyalgia	4	8
Lumps on head	4	8
Angina	2	4
Facial pain	1	2

TABLE 3. Presenting visual symptoms in forty-seven patients with giant cell arteritis

Visual symptom	No.	%
Loss of vision		
unilateral	20	42
bilateral	11	23
Blurred vision	11	23
Diplopia	2	4
Amaurosis fugax	1	2
Field loss	1	2
None	1	2

was another frequent visual symptom in one-quarter of patients.

The commonest ocular signs were those of ischaemic papillopathy which occurred in thirty-seven out of forty-seven patients (80%); it was unilateral three times oftener than bilateral papillopathy (Table 4). In two instances, unilateral ischaemic papillopathy became evident after the patients had been admitted to hospital and commenced on steroid therapy. Six patients presented with central retinal artery occlusion, five of whom also had evidence of contralateral ischaemic papillopathy.

Ophthalmoplegia due to lateral rectus palsy was noted in two patients.

As might be expected in this age group, there were additional factors contributing to visual loss, comprising cataract in five patients, glaucoma in two patients or old post-traumatic optic atrophy in one patient.

TABLE 4. Presenting ocular signs in forty-seven patients with giant cell arteritis

Ocular sign	No.	%
Ischaemic papillopathy	37	79
unilateral	29	62
bilateral	8	17
Central retinal artery occlusion	6	13
Cataract	5	10
Ophthalmoplegia	2	4
Glaucoma	2	4
Optic atrophy	1	2

Nearly one-third of patients reached hospital within 1 month of symptoms, and almost all (forty-four out of forty-seven) within 6 months.

Helpful investigations

Temporal artery biopsy

This was undertaken in thirty-one of the forty-seven patients, providing evidence of a giant cell arteritis in twenty-three. The biopsy proved to be technically unsatisfactory in six, and the sample submitted was normal in two instances.

Blood tests

The haemoglobin level was less than 12.0 g/100 ml in eighteen (38%) and less than 13 g in twenty-nine (62%). The leucocyte count was greater than 9000/mm³ in twenty (42%). The erythrocyte sedimentation rate (ESR) was always elevated. It was more than 100 in thirteen (28%) and more than 50 in thirty (64%). It was never below 40 mm (Table 5). There were raised α_2 globulins in seventeen (36%).

The alkaline phosphatase was only slightly elevated to 14.4 King-Armstrong units in one of eight patients in whom it was performed.

TABLE 5. Blood test results in patients with giant cell arteritis

Test	No.	%
Haemoglobin (G)		
<13	29	62
<12	18	38
Leucocytes over 9000/mm ³	20	42
ESR mm/1 hr		
>100	13	28
50-100	30	64
40-49	4	8
<40	0	
Raised α_2 globulin	17	36

Differential diagnosis

Eleven patients were admitted to hospital with the diagnosis of giant cell arteritis but subsequent investigation and observation led to a change of diagnosis to atherosclerotic ischaemic papillopathy in eight, and to a diagnosis of syphilis, monoclonal gammopathy or orbital pseudotumour in the other three patients. In ophthalmic practice it is sometimes difficult to distinguish ischaemic papillopathy due to arteritis from that due to atherosclerosis, particularly since both may co-exist in patients in this age group. The history, physical findings and ESR may provide differential points (Table 6). but if there is any doubt it is better to embark on a therapeutic trial of systemic steroids rather than withhold them.

TABLE 6. Differences between arteritic and atherosclerotic papillopathy

Differential features	Ischaemic papillopathy due to	
	Arteritis	Atherosclerosis
History of		
polymyalgia rheumatica	+	-
intermittent claudication	-	+
angina of effort	±	+
Signs of		
generalized atherosclerosis	±	++
absent pulses	-	+
attenuated retinal vessels	±	+
wide pulse pressure	±	++
ECG	Usually normal	Frequently ischaemic
ESR	Elevated	Normal
Immunoglobulins	Abnormal	Normal
Histology	Vasculitis	Atherosclerosis
Response to steroids	Dramatic	Nil

Response to oral steroids

All patients were treated with oral corticosteroids in a dose of 40 mg or more daily for 12–24 months. Thirty-two patients were also given 100 mg intravenous hydrocortisone on admission, followed by intramuscular hydrocortisone 100 mg every 8 hr during the first 36 hr of admission. The results are compared (Table 7). Irrespective of the route of administration, the vision of eleven patients (23%) improved, thirty (64%) remained unchanged, and six (13%) deteriorated despite treatment. There was no significant difference whether oral steroids were used alone or supplemented by intravenous and intramuscular administration. There was deterioration of vision despite steroids in six elderly women with longer histories before commencing treatment. Their length of history before steroid therapy averaged 12 weeks compared with 6 weeks in those who showed improvement with treatment. Irrespective of the visual response, the ESR always returned to normal, almost always within a month (Table 8).

Discussion

Our series confirms previous experience (Table 9) that giant cell arteritis is more commonly seen in women over 60 years of age. Since it is a generalized systemic disorder, this diagnosis should always be considered when the elderly have multisystem symptoms and signs. Early diagnosis is essential if visual failure is to be averted by prompt corticosteroid therapy. If the patient has already reached the late stage of visual failure then the lesion is irreversible and the only benefit of treatment is an effort to prevent bilateral visual failure.

TABLE 7. Response of vision in giant cell arteritis to corticosteroids

Vision	Steroid by					
	Mouth		Oral and parenteral		Total	
	No.	%	No.	%	No.	%
Improved	4	27	7	22	11	23
Unchanged	10	66	20	62	30	64
Worse	1	7	5	16	6	13
Total	15	100	32	100	47	100

TABLE 8. Influence of corticosteroid therapy on elevated ESR

No. of weeks before ESR returns to normal	Patients	
	No.	%
1	5	10
2	17	36
3	13	28
4	6	13
5	2	4
6	3	7
>6	1	2
	47	100

How then can we recognize the disorder sufficiently early? Awareness of the diagnosis by the family doctor to whom the elderly first report symptoms seems crucial for he can arrange an erythrocyte sedimentation rate (ESR) without delay. The ESR is easy to perform and provides a valuable pointer to the underlying arteritis. If an elderly person with any of the symptoms listed (Table 2) has an elevated ESR of 40 mm/hr or more then this should be strong grounds for considering artery biopsy and/or steroid therapy. A therapeutic response with amelioration of symptoms within a day or so and a fall in ESR within 2 weeks provides further diagnostic evidence. The diagnosis is a clinical one and should not remain dependent on temporal artery biopsy if this means a delay in commencing steroids. The principal value of parenteral, in addition to oral, steroids is that the early treatment is ensured.

Visual failure in this group of patients is most likely to be due to lack of awareness of the diagnosis resulting in a considerable delay in initiating steroid therapy, or because of co-existent glaucoma and atherosclerosis. Early treatment is recommended in an effort to overcome one treatable cause of blindness.

TABLE 9. Features of giant cell arteritis in reported series

Authors	Year	No. of patients	% Women	Age at onset (years)	Grounds of diagnosis	% with artery biopsy	% visual disturbance	% improved by steroids	Remarks
Cooke <i>et al.</i>	1946	7	57	66-73	Histology	85	85	Not used	43% mortality without steroids
Anderson	1947	54	55	67	Histology and Clinical	53	42	Not used	12% mortality without steroids
Heptinstall <i>et al.</i>	1954	14	28	48-75	Histology	100	73	40	Corticotrophin ± Cortisone
Ross Russell	1959	35	51	73	Clinical	31	45	Visual failure did not occur in steroid-treated, but developed in one-third of patients	
Hollenhorst <i>et al.</i>	1960	175	54		Histology and clinical	69	58	60% were treated	
Alestig and Barr	1963	10	50	70	Histology and clinical	90	0	100	
Cullen	1967	17	77	69-81	Histology	100	89	One patient	
Harrison <i>et al.</i>	1967	50	70	71	Clinical	14	6	Not stated	
Hamilton <i>et al.</i>	1971	25	64	69	Histology	100	60	Dramatic symptomatic relief in most	
Present series	1973	47	74	60-88	Clinical	50	100	25	

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