PARKLAND MEMORIAL HOSPITAL

MEDICAL GRAND ROUNDS

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THE POLYMYALGIA RHEUMATICA-GIANT CELL ARTERITIS SYNDROME

POLYMYALGIA RHEUMATICA

Historical. In 1888, Bruce (1), a Scottish physician described five elderly patients who suffered from widespread muscle pain. He named the disease "senile rheumatic gout" even though he recognized the difference between this entity, gout and rheumatoid arthritis. Since then, and particularly after 1950, numerous investigators have reported elderly patients suffering from an illness characterized by severe proximal muscle pain and stiffness associated with constitutional manifestations such as fever, weight loss, anemia and depression.

The condition has been given a variety of names (Table I) but the term "polymyalgia rheumatica" (PMR) coined by Barber in 1957 (2) is the one used by most clinicians at the present time.

TABLE I

SYNONYMS FOR POLYMYALGIA RHEUMATICA

Senile rheumatic gout (1)
Secondary fibrositis (3)
Periarthrosis humeroscapularis (4)
Peri-extra-articular rheumatism (5)
Myalgic syndrome of the elderly with systemic reaction (6)
Pseudo-polyarthrite rhizomelique (7)
Anarthritic rheumatoid disease (8)
Rheumatism inflammatoire rhizomelique desgens agés (9)
Polymyalgia arteritica (10)

Prevalence. In Europe, PMR has been recognized as a relatively common condition. Kogstad (11) in Sweden identified 70 cases over a 2-year period in a hospital with an annual admission of 1000 patients with rheumatic diseases. Hamrin (12) also in Sweden recognized an average of 12 new patients per year in a community of 100,000. Dixon (13) in England found an incidence of 1.3% in over 2200 new patients seen in a rheumatic diseases clinic over a period of 26 months. This number was similar to the number of new cases of gout or ankylosing spondylitis seen in the same period of time.

In this country, Davison (14) reported seeing 14 cases at one hospital in New York City over a two-year period and Wilske

and Healey (15) reported 18 cases seen at the Mason Clinic in Seattle in 18 months.

Epidemiology. Of the many large series of patients with PMR, in only one series of 18 patients (15) it is stated the racial distribution; all patients were Caucasian. It is almost certain that the Scandinavian papers include only Caucasians and a similar situation appears to be true with the English and French patients. In a conference on PMR held at the Mason Clinic on April, 1970 (16), the comment was made that the disease has an apparent predilection for Caucasians and upper socioeconomic groups. This may explain the fact that in the last ten years no patient has been discharged from Parkland Hospital with the diagnosis of PMR.

Sex Distribution. As shown in Table II, there is a 2 to 1 prevalence of females in a survey of 647 cases obtained from 20 published series (1, 2, 4-8, 11-13, 15, 17-24).

TABLE II

SEX INCIDENCE IN 647 PATIENTS WITH POLYMYALGIA RHEUMATICA

	Male	Female	Total
Number	223	424	647
Per cent	34.5	65.5	100

Age. One of the hallmarks of polymyalgia rheumatica is its occurrence in the elderly population. The mean age at onset calculated from 289 patients turned out to be 65.2 years. As a matter of fact, with very few exceptions, the age at onset of the disease was over 50 years in the vast majority of patients.

Clinical Picture. The onset of the disease is often abrupt with pain and stiffness of the muscles of the proximal portions of the limbs and neck. It is not unusual for some patients to be able to date the onset of pain to within 1 or 2 days. In a series of 92 patients published by Hamrin (12), 40 per cent had acute onset of symptoms. In the remaining patients, the mode of onset was more insidious, evolving over a period of a month or more. The distribution of primarily affected muscle groups in Hamrin's patients is shown in Table III.

TABLE III

LOCALIZATION OF MUSCLE PAIN IN POLYMYALGIA RHEUMATICA

Affected Muscle Groups	Number of Patients	Percent
Brachio-cervical	59	64
Caudal	30	32
Brachio-cervical and caudal	2	2
Back	1	1
Diffuse	1	1

The great majority of patients develop myalgia in the neck and shoulders, hips and thighs, with the brachio-cervical region being affected more commonly. The symptoms are usually symmetrical. Pain also involves periarticular tissues such as biceps tendons or popliteal area or even the peripheral joints. Muscles and tendons are often tender to palpation. Pain is accentuated with active motion and can result in near incapacitation in some patients. In general, morning stiffness of the type seen in rheumatoid arthritis is present. Although the patients may appear weak because of apprehension or severe pain on motion no true muscle weakness is detectable.

Arthralgias or transient synovitis usually lasting few days with objective joint swelling and increased amounts of joint fluid may occur. The joints more commonly affected are the knees, metacarpophalangeal, proximal interphalangeal and wrists. In the different series reviewed, the incidence of synovitis varied from 10 to 50% of the patients. Synovianalysis has been reported in a handful of patients only and the fluid appeared to be similar to that in rheumatoid arthritis.

About 50% of the patients with PMR have associated history or positive findings of temporal arteritis. In some cases the symptoms and signs appear weeks or months after the onset of polymyalgia.

As a rule most patients develop systemic manifestations early in the disease. Fever is a common finding with over half the patients reported being affected. The temperature is usually low-grade but occasionally is severe enough to suggest sepsis. Night sweats are also a common finding. PMR should always be considered in the diagnosis of Fever of Unknown Origin in the elderly population as discussed by

Petersdorf (25). Fatigue and general malaise is the rule in most patients. Over 60% of the patients in Hamrin's series lost 12 pounds or more. In most cases anorexia was present Depression and change in behavioral patterns have been reported in several patients. A summary of the clinical findings is shown in Table IV.

TABLE IV

CLINICAL FINDINGS IN POLYMYALGIA RHEUMATICA

	Per cent
Pain and stiffness localized to proximal limbs and neck	100
Arthralgias or arthritis	10-50
Temporal arteritis	50
Fever	50-90
Anorexia and weight loss	60-90
Fatigue and general malaise	80-90

As shown in Table V the majority of the patients are symptomatic for over 3 months before the diagnosis is made.

TABLE V

INTERVAL BETWEEN CLINICAL ONSET OF POLYMYALGIA RHEUMATICA AND DIAGNOSIS

Months	No. of Patients
0-3	31
4-6	20
7-9	19
10-24	14
>24	9

Associated Conditions. The most commonly reported condition associated with PMR is cranial arteritis. We will discuss this association in detail later.

A small number of patients presenting with the typical picture of PMR eventually develop rheumatoid arthritis or Sjögren's syndrome (7, 8, 13, 19). A few patients with systemic lupus erythematosus (26) or scleroderma (27) may

also present with a clinical picture compatible with PMR. These patients usually made no more than 5% of an aggregate of 247 patients from seven series.

A few patients with concomitant carcinoma also develop PMR (12, 27-29). The possibility of malignancy should be kept in mind when working these patients up.

Laboratory Findings. The hallmark of PMR is the finding of a strinkingly elevated erythrocyte sedimentation rate. In the series reviewed the average ESR values were commonly about 100 mm/hr using the Westergren method. With very rare exceptions, the ESR was over 50 mm/hr in all untreated patients. In part, this is due to the fact that an ESR of over 50 is one of the criteria used for inclusion in several series. Analysis of an aggregate of 176 cases from publications where an elevated ESR was not a required criterium revealed that 95% of the patients had elevated values.

A normochromic or hypochromic anemia is found very frequently in patients with PMR. The average hemoglobin concentration is usually around 11 gms%. These patients do not respond to treatment with iron and are considered to have a reutilization defect such as that seen in other chronic inflammatory diseases. In general, serum iron is low in these patients and iron binding capacity normal or low.

The incidence of positive rheumatoid factor tests taken from over 250 patients turned out to be 7.6% which is no higher than that for a control population of the same age bracket.

Whenever serum enzyme determinations have been done to rule out muscle damage, the results have been completely negative. Similarly, electromyographic studies performed in several series (14, 15, 19, 28-30) have shown no significant abnormalities.

Pathology. Muscle biopsies have shown few abnormalities and usually have been reported as normal. No evidence for myositis or vasculitis has been found. In about 15% of the muscle biopsies reported, slight chronic inflammation in muscle septa or perivascular areas were noticed.

Course. When PMR is not associated with any other condition, the prognosis is good. In general the disease will follow a protracted course with an average duration of 2 to 3 years before remission occurs.

In a small group of patients reported by Gordon (18) not treated with steroids, the average duration of symptoms was

nearly 3 years. All patients recovered when seen up to 3 years after remission. Bagratuni (31) reported on a group of 46 patients followed up for a maximum of 16 years. The average duration of the disease was 7.1 years in this group. All surviving patients except two had either improved or were symptom-free at the end of the study.

GIANT CELL ARTERITIS (GCA)

Historical. At the time that Bruce published his paper on "senile rheumatic gout" another English clinician, Jonathan Hutchinson, described a patient with temporal arteritis (32) in a manner that deserves quoting: "The subject of this case was an old man named Rumbold...He was upwards of 80, and almost in his dotage. I was asked to see him because...he had red "streaks on his head" which were painful and prevented his wearing his hat...The "red streaks" proved, on examination, to be his temporal arteries, which on both sides were found to be inflamed and swollen. The streaks extended from the temporal region almost to the middle of the scalp, and several branches of each artery could be distinctly traced. The conditions were nearly symmetrical. During the first week that he was under my observation, pulsation could be freely detected in the affected vessels, but it finally ceased; the redness then subsided, and the vessels were left impervious cords. At no time any gangrene of the skin of the scalp threatened. The old gentleman lived, I believe, several years after this without any other manifestations of arterial disease. It was thought that the pressure of his hat on his temples had been the exiting cause of the arteritis.

In 1932 the clinical syndrome of "temporal arteritis" was rediscovered by Horton et al (33) who reported two cases presenting with fever, weakness, anorexia, anemia and painful tender areas over the scalp and along the temporal vessels.

Prevalence. The reported clinical incidence of GCA has ranged from 2 to 10 patients per 100,000 population per year (12, 34). A study conducted in an Italian institution for the aged, Nosenzo et al (34) found 9 patients of temporal arteritis among 100 elderly inmates. The high clinical incidence of GCA among the elderly population is also shown by pathology studies. Ainsworth et al (35) found 2 cases with temporal arteritis in 39 random post mortem examinations in patients over 60 years old.

Ostberg (36) examined 1097 temporal arteries obtained from 65 per cent of all persons dying during one year at Mälmo, a town with a population of 250,000. He found evidence of

arteritis in 12 or an incidence close to 1 per cent.

Age and Sex Distribution. As in PMR, GCA occurs most commonly in patients over 50 years old with an average age similar to that found in PMR. There are, however, occasional patients younger than 40 with well documented GCA (37-39). There is a 2 to 1 preponderance of females over males in some published series (21, 40) and in others the sex incidence is equal.

Initial Manifestations. GCA is usually associated initially with systemic manifestations such as fever, sweats, anorexia, weight loss, malaise, fatigue, depression, headache, myalgia or arthralgia. Because of the vagueness of these features, manifestations directly related to obliterative vascular changes are more likely to lead to clinician to the correct diagnosis early in the disease. These sometimes catastrophic occurrences are almost invariably preceded by vague systemic changes which are commonly either overlooked or misinterpreted as indications of neoplastic, infectious, degenerative or functional disorders. Unfortunately, the systemic manifestations may precede the onset of typical symptoms of GCA by many months or even years. In the series reported by Hamilton et al (41), symptoms were present for an average of five months before the diagnosis was finally established.

From the above considerations it follows that a good number of patients will not have local symptoms of arteritis when examined. Out of 109 patients with confirmed arterial biopsies taken from Hamrin (12) and Fauchald et al (24), 40 per cent had no symptoms or signs of temporal arteritis.

One of the most common symptoms recorded in patients with GCA is headache. Although it is usually localized to the temporal area it can be generalized and in patients with polymyalgia it may be occipital in character. These headaches may be of a nonspecific nature or may conform to the classical symptoms of "temporal arteritis" with abrupt onset with pains which may be localized to the temporal area or may radiate widely over the scalp, face, jaws and occiput. These pains are usually of a continuous boring quality with episodic lancinating exacerbations. Scalp tenderness may prevent resting the head on a pillow or brushing the hair. Fever is seen in 80 per cent of patients with GCA and since the absence of symptoms and signs of arteritis is not uncommon, these patients become diagnostic problems quite frequently (25, 42).

TABLE VI

INITIAL MANIFESTATIONS OF GIANT CELL ARTERITIS

Headache
Myalgia
Fever
Anorexia
Weight Loss
Malaise
Depression

Arterial Manifestations. As mentioned above, between 30 to 50 per cent of patients with confirmed biopsy findings do not have abnormal findings related to temporal arteritis. The most common positive finding is that of absent or diminished arterial pulsation. This physical sign is more common than the finding of tender nodular, painful arteries, seen in no more than 30 per cent of the patients. A small proportion of patients present with masticatory claudication, almost exclusively seen in patients with GCA and usually related to obliteration of the facial artery.

Dixon et al (13) have noticed carotid sinus hypersensitivity in 31 per cent of their patients with GCA.

Some authors (12, 23) have emphasized the presence of arterial bruits detectable in a fair proportion of patients. Hamrin (12) found an incidence of 59% in patients with GCA or PMR against 16% for a population of age matched controls. Bruits were heard most commonly over carotid, subclavian, axillary and brachial arteries.

TABLE VII

ARTERIAL MANIFESTATIONS IN GIANT CELL ARTERITIS

Diminished or absent pulsation
Arterial pain or tenderness
Arterial nodularity
Arterial bruits
Masticatory claudication
Carotid sinus hypersensitivity
Visual symptoms
Aortic arch syndrome
Cerebrovascular accidents
Myocardial infarction

Eye Involvement. Loss of vision is the most serious sequela of GCA. The incidence of eye symptoms varies widely from one series to another: From a minimum of 6% recently reported by Fauchald (24) to about 60% reported by Meadows (43). A compilation of many cases with proven arterial biopsy showed an average evidence of eye symptomatology close to 40 per cent. Unilateral or bilateral blindness due to ischemic optic neuritis is the most frequently observed lesion occurring in two-thirds of the patients with visual symptoms. Onset of blindness is usually abrupt and is frequently preceded by episodes of transient blurring of vision. It should be emphasized that only in rare cases visual impairment was the initial manifestation of GCA. Hamilton et al (41) observed that the average duration of systemic manifestations of GCA was 3.5 months before the onset of blindness.

Other visual symptoms seen in the remaining one-third of patients include: transient blurring of vision and diplopia (44). Ophthalmoscopic examination shortly after the onset of blindness may reveal no abnormality. Within 1 to 2 days, edema of the optic disc is usually evident. The edema has been noted to resolve within ten days and cotton-wool patches may persist for two weeks. Optic atrophy is the end result in most of these patients.

Aortic Arch Syndrome. Histologic involvement of the aorta and its major brances is present in the vast majority of patients with GCA (12, 38, 45-47). As a matter of fact, the intensity of the inflammatory lesions diminishes centripetally. In most cases, aortic arch involvement remains clinically silent. Clinical symptomatology related to stenosis of the aortic arch and major vessels is seen in 15 to 20 per cent of patients with GCA. The most common manifestations include the appearance of "pulseless disease," aneurisms of the aorta and major brances, dissection and intermittent claudication. The affected patients present clinical and pathological features indistinguishable from Takayasu's arteritis (48, 50), an inflammatory arteritis involving major vessels observed in young females. The two diseases seem to differ only in the age group involved since the clinical and histologic features are remarkably similar. Nakao et al (50) reported a series of 84 cases, mostly women from age 10 to 49. In two-thirds of these patients, there was a positive history of systemic-manifestations such as fever, myalgia and elevated ESR. There is also good evidence of improvement in such cases following treatment with corticosteroids (52). Our ignorance regarding etiological factors in both entities makes it impossible to decide at the present time whether both diseases are related or not.

Cardiac Involvement. Giant cell arteries of the coronary arteries has been demonstrated in several cases at autopsy. Most patients with this finding died of myocardial ischemia (53-57). When giant cell arteritis presents as an acute myocardial infarction, the nature of the underlying disorder may be overlooked. An unduly prolonged fever, excessive or prolonged elevation of the ESR, a normocytic anemia or elevation of the alpha globulins in patients with myocardial infarction may be clues suggesting arteritis.

Neurologic Manifestations. Autopsy studies have demonstrated giant cell arteritis involving carotid, vertebral, meningeal and intracerebral arteries (38, 43, 47, 63). Hemiparesis and subarachnoid hemorrhages are widely recognized complications usually occurring at the time of exacerbation of the disease (40, 41, 47, 53, 63, 64). Diffuse cerebral dysfunction related to GCA may present as dementia, confusion, hallucinations or coma (42, 63, 64).

Laboratory Findings. These are identical to the findings in PMN. As a rule, most patients present with very elevated ESR values averaging close to 100 mm/hr (12, 21, 40, 41). They also show normochromic or hypochromic anemia of the same magnitude (28, 40-42, 58) and other indices of inflammatory activity such as elevated alpha globulins show by serum electrophoresis.

Pathology. Histological lesions are typically seen in a patchy distribution within the arterial wall. There is a prominent inflammatory infiltrate composed of lymphocytes, plasma cells and macrophages usually involving the entire thickness of the vessel. In some instances a few eosinophiles can be identified. Giant cells within the vessel wall are almost a constant feature of the arteries. They are absent, however, in the cases where inflammation is confined to the intima or when fibrosis is the predominant feature. Therefore, the presence of giant cells is not an absolute criterium for the diagnosis. In addition to the patches of cellular infiltrate, special stains reveal widespread fragmentation of the internal elastic lamina in the areas of inflammation. Thrombosis is common at the sites of active inflammation and evidence of recanalization suggesting previous thrombosis is frequently demonstrable. The intima displays loose fibrous tissue proliferation resulting in marked thickening.

TABLE VIII

PATHOLOGIC FINDINGS IN GIANT CELL ARTERITIS

Mononuclear inflammatory infiltrate
Giant cells
Fragmentation of internal elastic lamina
Intimal fibrosis and thickening
Thrombosis

Diagnosis. The essential elements that enable the clinician to make the diagnosis of GCA are the realization that this is not an unusual disorder and to suspect its presence in patients with a broad spectrum of clinical presentations. As we have indicated previously, the presence of clinical findings of arterial involvement is the exception and not the rule so that the presumptive diagnosis of GCA should be entertained in elderly patients with vague systemic complaints, muscular aches and pains, FUO, unexplained anemia and particularly unexplained elevation of the ESR over 50 mm/hr.

Definitive diagnosis can only be made by microscopic examination of an affected artery. Excision of a segment of a temporal artery has been the diagnostic technique most widely used. Detectable abnormalities of these vessels on physical examination should not be considered a prerequisite to obtain a biopsy in suspected cases. The lesions of GCA may be extremely localized and this accounts for negative biopsies in obvious cases with the disease. It is important, therefore, to obtain as generous a portion of artery as seems feasible and to examine the whole specimen with serial sections.

Angiography may be a helpful procedure to localize involved areas. Recently, the technique of selective temporal arteriography (59) has been shown to be a useful tool to localize the areas of the temporal artery which are affected. The procedure is usually performed at the time of biopsy.

The presence of segmental lesions correlates well with positive biopsy findings. Hunder et al (60) found 7 positive arteriograms of 31 suspected cases of GCA. Of these 7 cases only 5 showed positive biopsy findings.

After such procedures have yielded negative results and the index of suspicion in any given patient is still high, the

performance of a clinical trial with corticosteroids may be indicated.

Course and Prognosis. The disease responds uniformly to corticosteroid treatment so that the incidence and morbidity of serious ocular involvement has been markedly diminished in the last 20 years.

The fatality rate in GCA is low even without treatment (61, 62). In a review of 57 cases reported prior to 1947, Anderson (53) recorded 7 deaths. Patients dying of GCA in recent years have either been untreated (63) or received inadequate therapy. Fatalities due to GCA are most often related to cerebral (47, 53, 63) or cardiac involvement (12, 55, 57).

THE RELATIONSHIP BETWEEN POLYMYALGIA RHEUMATICA AND GIANT CELL ARTERITIS

Porsman (17) in 1951 was the first to call the attention to the many similarities between what he called a special type of arthritis in old age and temporal arteritis.

Paulley and Hughes (64) in 1960 showed the high incidence of "anarthritic rheumatism" in 76 cases of giant cell arteritis. Finally, Alestig and Barr (21) demonstrated the presence of arteritis by biopsy in 7 of 9 patients with PMR. None of these patients had symptoms referable to the temporal arteries or abnormalities of these vessels on palpation.

The relationship between the two diseases must be examined from different points of view since the criteria for inclusion of patients in the published series has varied widely.

What is the incidence of PMR in patients with typical GCA? Selecting the series where the diagnosis of GCA was made on the basis of symptoms and signs of arteritis, the overall incidence of 52 per cent was found.

TABLE IX

INCIDENCE OF POLYMYALGIA RHEUMATICA IN PATIENTS WITH TYPICAL GIANT CELL ARTERITIS

Author	Number of Cases With GCA	Per Cent With PMR
Paulley and Hughes (64)	76	43
Hamilton et al (41)	25	25
Wadman and Werner (65)	52	62
Bevan et al (66)	37	38
Hamrin (12)	34	91
Total	224	52

It can be concluded that PMR is a common event in patients with overt giant cell arteritis.

The second question to be asked is the following: Is PMR always a manifestation of either overt or inapparent GCA? This problem was investigated in patients with typical PMR at the time of inclusion in prospective series where most patients had undergone a temporal artery biopsy. An aggregate of 164 patients from 5 series yielded an average of 49 per cent with positive biopsy findings.

TABLE X

INCIDENCE OF GIANT CELL ARTERITIS IN PATIENTS WITH POLYMYALGIA RHEUMATICA

Author	Number of Patients With PMR	Per cent With GCA
Alestig and Barr (21)	9	78
Dixon et al. (13)	29	38
Hamrin (12)	44	41
Bruk (23)	33	45
Fauchald et al. (24)	49	41
Total	164	49

This estimate is almost certainly low because of the segmental distribution of arterial lesions, biopsies may yield negative results in a proportion of patients with overt GCA. Excluding the small number of patients with PMR secondary to other causes such as collagen diseases and malignancies, there remains still a group of patients with no other associated condition. Whether this group constitutes a separate entity or is associated with clinically inapparent GCA remains a matter for speculation at the present time.

Eye involvement in polymyalgia rheumatica. Since the greatest danger in patients with GCA is loss of vision, which occurs in 40% of the cases, it is most important to consider whether the diagnosis of PMR implies a similar risk.

This question was examined in a combined group of 49 patients with pure PMR (13, 15, 67-69).

TABLE XI

VISUAL DISTURBANCES IN POLYMYALGIA RHEUMATICA

Biopsy	Number of Patients	Per Cent With Eye Symptoms
Positive	15	53
Normal	34	6

In 15 of the 49 patients, temporal artery biopsy was positive and 53 per cent had visual disturbances. In the remaining 34 with normal temporal artery biopsies the incidence of visual abnormalities was only 6 per cent.

In general, there is consensus in the literature that in patients with PMR and no clinical or biopsy evidence of arteritis the risk of blindness is small.

Management. The treatment of patients with PMR or GCA is dictated by two considerations: the danger of blindness and the long, protracted course of the disease.

In view of the high incidence of visual involvement in patients with positive biopsies and the fact that a large group of patients will demonstrate lesions of arteritis in the absence of clinical manifestations indicating arterial involvement, patients with PMR should have temporal artery

biopsies whenever possible. Temporal arteriography may be useful to localize the involved areas; otherwise, an attempt should be made to remove about 1 cm length of artery.

The symptoms of PMR respond well to anti-inflammatory agents such as butazolidine or small doses of Prednisone (10 mg daily). On the other hand, there is abundant evidence that such measures are unable to stop the onset of blindness or other symptoms of GCA (18, 28) while higher doses are well known to prevent the onset of visual manifestations.

Birkhead et al (61) compared 55 patients with GCA treated with corticosteroids and 53 diagnosed prior to the availability of such agents. On admission to the Mayo Clinic there were 16 blind eyes among the first group of patients. On discharge, after institution of treatment, the number of blind eyes had only increased to 18. In the untreated group, there were 16 blind eyes on admission and 24 on discharge.

At the present time the recommended initial dosage is 40 to 60 mg Prednisolone daily. This dose should be maintained for about two to three weeks or until the clinical symptoms have completely disappeared and the ESR is less than 20 mm/hr by the Westergren method. Then, prednisone should be tapered slowly to an average maintenance dose of 7.5 mg to 15 mg daily. In general, this amount is enough to maintain the patients in a symptom-free state.

It should be kept in mind that a number of patients with pure PMR and negative biopsies have developed symptoms of GCA including visual loss two years after the onset of myalgias and systemic manifestations. Therefore, these patients should be followed very closely and the prednisone dose should be immediately increased if muscle and systemic symptoms reappear, if there is an elevation of the ESR or if the patient complains of severe headaches even in the absence of local signs of temporal artery involvement.

The response to corticosteroid therapy is usually dramatic, with complete disappearance of the muscle and systemic symptomatology within 1 or 2 days.

It has been the experience of most authors that the disease uniformly recurs if corticosteroids are stopped after less than 6 months of therapy. Therefore, treatment should be continued for two years and attempts at complete steroid withdrawal should be made every 2 to 3 months. After two years of treatment, Fauchald (24) reported a recurrence rate of 18%.

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