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GIANT CELL (TEMPORAL) ARTERITIS

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GIANT CELL ARTERITIS is a wide-spread inflammatory disease of the arteries, occurring in elderly patients. The most striking signs and symptoms of this disorder in its classic form result from the involvement of cranial arteries. Since it was originally defined as a clinical entity by Horton in 1932, however, the concept of this disease has changed considerably.

Giant cell arteritis was initially regarded as a benign, self-limited disease affecting primarily the temporal arteries. As early as 1941, however, Gilmour clearly demonstrated the diffuse nature of the arteritis and introduced the less restrictive title of giant cell arteritis. In 1951, Cardell reviewed the 27 fatal cases which had been reported and added another of his own, emphasizing the diffuse and potentially fatal nature of the disease. More recently Paulley and Hughes reviewed their 75 cases and observed that "giant cell arteritis once believed to run a benign course, kills or maims a number of those afflicted, and when in remission, it may only sleep". They stressed the protean and potentially serious nature of the disease, and emphasized the possibility of late, transient, or lack of involvement of the temporal arteries. A brief review of four cases will illustrate some of the important features of the disease, and its treatment.

CASE 1 (K. F.) — Age 72, admitted 9/19/50, discharged 10/24/50. A 72 year old woman was admitted to the hospital because of blindness of the left eye of three days duration. For four days prior to the onset of the blindness, there were intermittent episodes of loss of vision in the same eye. She had had persistent head pains for one month.

On examination the temporal vessels were prominent, tortuous and tender. There was blindness of the left eye and the optic disc was pale. Transient blindness occurred in the right eye on two occasions after admission, without subsequent persistent loss of vision.

With no specific therapy the head pain subsided over a period of four months. The left eye remained blind and the optic disc was atrophic. The temporal arteries persisted as hard cords.

Comment: This patient represents the commonly recognized "classic" picture of temporal arteritis in its self-limited form, complicated by optic atrophy with blindness.

CASE 2 (B. W.) — 82, admitted 10/23/56, discharged 11/3/56. This 82 year old man was admitted to the hospital one day following the abrupt onset of blurred vision in the right eye. For three weeks prior to his admission, there was persistent pain over the left side of the head.

On examination the temporal vessels were prominent, irregular and tender, especially on the left. The right optic disc was blurred, and on the following day, when the vision was reduced to counting fingers, the disc was indistinct and slightly elevated with some

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small hemorrhages superiorly. The visual fields showed a large superior altitudinal central scotoma on the right. On the following day he was started on Prednisone, 60 mg. daily. The left temporal artery was resected and showed typical changes of giant cell arteritis (Figure 1).

The patient's headache was promptly relieved following the resection and institution of steroid therapy, and his vision gradually improved to 20/30 with a residual small superior altitudinal scotoma on the right.

Comment:

This patient's course was quite similar to that of the preceding patient, but illustrates the possible restoration of vision with prompt steroid therapy.

CASE 3 (M. B.) — 77, admitted, 8/1/58, discharged 9/11/58. On admission to the hospital, this 77 year old woman complained of generalized weakness, aches and pains in the neck, shoulders and extremities, with diffuse aching soreness and tenderness of the head. The illness began four months prior to admission, with shooting pains in the head, face and ears. She became markedly anorexic, lost 15 lbs., and felt weak and fatigued. There were periods of confusion of variable degree.

One month prior to admission she began having drenching sweats in association with the appearance of tender, elevated, red nodules over the forehead.

On examination, the patient appeared chronically ill. She was dull and somewhat depressed. There were dime-sized erythematous nodules along the course of the temporal arteries, particularly on the right. The right temporal artery was firm and acutely tender, with decreased pulsation. The optic discs and fields were normal. The remainder of the neurologic examination was not remarkable.

The sedimentation rate was 40 mm/hr. and the hemoglobin 10.5 gms. There was a daily afternoon temperature rise, to about 101.6. The temporal artery was resected and showed typical changes of temporal arteritis.

She was given two injections of Hydrocortisone 100 mg. four hours apart in the evening, and on the following day felt markedly improved, with relief of the aching soreness in the head. The erythematous nodules over the forehead and the fever subsided in approximately 48 hours. Hydrocortisone 160 mg. was continued daily for one month.

Following discharge from the hospital, the dosage of Hydrocortisone was gradually reduced and then discontinued at the end of three months. One month later, the patient was re-admitted to the hospital, complaining of excessive fatigue, with pain and stiffness in the back and thighs.

The temporal arteries felt irregular and hardened, but were not tender or inflamed. She again showed a mild daily temperature rise to 100.

The blood hemoglobin was 9.7 gms. Serum electrophoresis showed a total protein of 6.1 (N 6.5-8.3) with albumin 2.35 (N 4.1-5.6), A1 globulin 0.46 (N 0.17-0.37), A2 globulin 0.87 (N 0.34-0.76), B globulin 0.87 (N 0.52-1.01), gamma globulin 1.55 (N 0.61-1.34).

On resumption of steroid therapy (Prednisone 5 mg. t.i.d.) the symptoms again subsided. Five months later, steroid therapy was stopped a second time, and in two weeks she again had a recurrence of headaches, soreness in the right temporal region and leg pains, associated with vomiting for two days. During the course of this acute recurrence she noted impairment of vision to the right and some weakness of the right side of the face. There was no apparent facial weakness when she was seen several days after the onset of this difficulty in the Clinic, but it was felt that she had a homonymous relative decrease in the peripheral right field. Her symptoms again subsided on resumption of Prednisone 10 mg. daily. There was no further recurrence of symptoms when the Prednisone was gradually discontinued again at the end of six months.

Comment:

This patient's illness was marked by prominent systemic complaints with periods of confusion for several months prior to the appearance of obvious temporal artery involvement. There was recurrence of symptoms on interruption of steroid therapy, three and eight months after it was initiated. The second recurrence was associated with transient left cerebral symptoms, possibly on the basis of cerebral ischemia.
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CASE 4 (B. E.) — Admitted 11/12/57, discharged 11/27/57. Eight weeks prior to admission, this 61 year old woman began having left frontal throbbing headaches with a "flu-like" illness. Abrupt loss of vision occurred in the left eye three weeks before admission. During this illness she was anorexic and had a weight loss of 10 lbs.

Two weeks before the onset of symptoms, penicillin had been applied locally after a tooth extraction and the following day, swelling of the left face and lips appeared. This cleared completely in several days with antihistamine therapy. There was no previous history of penicillin sensitivity. On admission the patient complained of persistent left frontal pain, and loss of vision in the left eye.

Neurologic examination revealed an optic neuritis with marked loss of vision on the left. There was some residual vision in the upper nasal quadrant with a large central defect. No definite abnormality of the temporal arteries was noted by several examiners, even though the possibility of temporal arteritis was considered.

An EEG showed a left fronto-temporal delta focus. In view of this finding and the absence of apparent temporal artery disease, an arteriogram was done and this was normal.

The blood hemoglobin was 9.5 gm., the sedimentation rate 45 mm/hr. The spinal fluid protein was 25 mg%. The total serum protein was 6.5 with albumin 27% (N 44.2-61.7), gamma globulin 28% (N 11.3-24.9), beta globulin 14% (N 9.7-16.9), alpha2 23% (N 7.4-13.4) and alpha1 9% (N 3.7-7.3).

The patient was discharged from the hospital on Prednisone 40 mg. daily. When she was seen again, one week later, she was free of head pain and felt well. The optic disc changes were subsiding, but there was no improvement in vision. Prednisone therapy was discontinued at the end of two months, but was promptly reinstituted by her local physician because of the recurrence of head pain, and this was again promptly relieved.

The patient was seen three years later, in Ophthalmology Clinic and was reported to have no significant change in her visual field or acuity. She had had no further recurrence of the difficulty, but the total duration of therapy was not recorded.

Comment:

This patient was of particular interest in view of the absence of apparent involvement of the temporal arteries even after the appearance of optic nerve complications presumably due to ophthalmic arteritis. It is quite possible the temporal arteries may have shown histologic changes; but even without this evidence, her history and response to therapy would appear to leave little doubt regarding the diagnosis. The occurrence of symptoms in this patient within two weeks following an allergic reaction suggested the possibility that the giant cell arteritis may have represented in this case a delayed hypersensitivity phenomenon.

CLINICAL PICTURE

Giant cell arteritis is a disease of elderly patients which occurs most commonly past the age of 60. There are at least two cases in the literature however, in which the disease is reported in young women, but the diagnosis is open to question in at least one. Gilmour's patient was a 23 year old woman with a clinical picture more consistent with the diagnosis of pulseless disease (Takayasu's disease, aortic arch syndrome). The pathology of the temporal artery in Meyer's 22 year old patient was not typical and suggests the possibility that this may have represented some other form of arteritis.

Although the onset, course and involvement in giant cell arteritis are varied, pain in the head, neck or face, associated with tenderness in the region of the involved temporal occipital or facial arteries, is usually a prominent feature of the illness, initially or at sometime during the course of the disease. Systemic symptoms including fever, anorexia, weakness, weight loss and pains in the joints and muscles
often appear in association with the head pain and in many cases may precede it for a month or more.

The head pain is often described as “sharp or shooting” with more persistent aching and soreness in the head or scalp. It may fluctuate markedly in character and severity and in some cases may be insignificant or obscured by other, more dramatic symptoms. Visible or palpable changes in the affected arteries, usually appear in association with the pain. Diffuse redness and swelling or erythematous nodules may be seen in the skin overlying the involved vessels. The artery is usually tender, thickened, and often nodular and tortuous. In some patients however, pain, scalp tenderness and other symptoms may occur without obvious changes in the cranial arteries, and Fisher commented recently on the importance of careful palpation for compressibility of the cranial vessels, with biopsy of suspected vessels for possible confirmation of the presence of giant cell arteritis.

Blindness, the most common serious complication of the disease, occurs in over 50 per cent of the patients. Loss of vision usually occurs from within a few days up to ten months after the onset of symptoms, but it may be an initial symptom. In some patients transient episodes of blindness occur with or without subsequent loss of vision (Case 1). Such episodes may be confused with similar disturbances which occur in carotid artery occlusive disease. The blindness usually occurs abruptly in one eye and may be followed by involvement of the other eye within days or weeks. If blindness is not complete, altitudinal and sector defects are most common, but any form of field loss may result with involvement of the retina, optic nerves or chiasm. The visual loss is most often associated with an ischemic optic neuropathy (Case 2), due to arteritis of ophthalmic artery and its branches. Retrolublar neuritis (Case 1) and central retinal artery occlusion are less frequent causes. Before steroid therapy, giant cell arteritis resulted in blindness in one or both eyes of approximately 50 per cent of the patients. Blindness has been reported following the institution of steroid therapy, but Hollenhorst reported that only one of their patients lost vision after the onset of therapy. Improvement in vision may occur following institution of therapy (Case 2), and it is likely that subsequent involvement of the second eye will be prevented if the period of therapy is sufficiently prolonged.

Ocular motor paralysis and diplopia without demonstrable palsy occur in about 15 per cent of the patients. This complication usually appears prior to the onset of visual loss and the paralysis is apt to fluctuate in character. Ptosis is the most common finding, but any of the ocular muscles may be affected. Recovery often occurs in days, but palsies may persist for months, and there may be permanent residual impairment. As in the case of blindness, diplopia and ocular muscle palsy may appear in the absence of gross evidence of arteritis, simulating in this case the picture seen in diabetic ophthalmoplegias.

Cerebral infarction is not a common complication but may occur during the acute phase of the disease. Cranial arteritis occurs in patients in the age group prone to develop arteriosclerotic and vascular lesions unrelated to the disease. It has been demonstrated, however, on autopsy in some fatal cases, that giant cell arteritis of the internal carotid and vertebral vessels may result in thrombosis and cerebral
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infarction.\textsuperscript{15,6} Multiple areas of cerebral infarction may also be caused by embolization from mural thrombi resulting from arteritic coronary artery occlusion.\textsuperscript{15} Involvement of the terminal cerebral vessels is not reported in these cases, but Crompton\textsuperscript{2} recently reported a fatal case with evidence of arteritis not only in the vertebrals, but also in a branch of the posterior cerebral, as well as in small arteries in the roof of the fourth ventricle and in the leptomeninges of the lower medulla. McCormick\textsuperscript{9} reported giant cell arteritis involving small meningeal and intracerebral vessels with severe cerebral involvement in two patients, but the diagnosis was questionable in the patient presenting the most striking small vessel changes.

Mental changes, manifested by confusion, dementia and depression, are relatively common\textsuperscript{13} during the course of the illness, and it is likely that these changes result from ischemia or thrombembolic cerebral disease.

In his review of 75 cases of giant cell arteritis, Paulley,\textsuperscript{11} who prefers the use of the term “arteritis of the aged”, makes a vigorous plea for the recognition of the atypical forms in which the classical symptoms may be lacking or overshadowed by other major problems. In addition to the clinical pictures already described, he adds as modes of presentation cardiac ischemia, anarthritic rheumatism, pain in the ear with vertigo and deafness, vomiting and cachexia, episodes masquerading as meningeal irritations, pyrexia of unknown origin, the aortic arch syndrome and dissecting aneurysms of the aorta in the aged. In his report, he presents cases illustrating these unusual atypical modes of presentation.

Figure 1a

Cross Section of temporal artery (Case 2) showing eccentric intimal proliferation and cellular infiltration predominantly at the junction of the thickened intima and media.

(H & E stain 17 x)
DIAGNOSIS

Biopsy of accessible involved cranial vessels usually reveals a characteristic picture which is diagnostic of this disease, in combination with the varied clinical symptoms described in an elderly patient.

Figure 1b
Segment of the wall of the artery in Fig. 1a showing cellular infiltration of mononuclear cells with multinucleated giant cells in area of infiltration.
(H & E stain 170 x)
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The typical features seen in affected vessels include marked intimal proliferation with obliteration of the lumen, degeneration of the internal elastic layer with giant cells concentrated in this area, degenerative changes in the media, and cellular infiltration predominantly in the media but extending into the intima and adventitia (fig. 1).

In some patients the biopsy may be inconclusive or there may be no apparent involvement of accessible vessels. In these cases a trial of steroid therapy should be given for diagnostic as well as therapeutic purposes. This is preferable to delay of treatment until the appearance of obvious vascular changes which may not occur until serious complications have intervened.

Other laboratory studies, though not diagnostic, provide supportive evidence. The erythrocyte sedimentation rate is usually grossly elevated, anemia with leukocytosis is quite common, and protein electrophoresis is often abnormal with a decrease in albumin and an increase in alpha or beta globulin fractions (Cases 3 and 4). The spinal fluid, although usually normal, may show an elevated protein.

TREATMENT

The response to steroid therapy in giant cell arteritis is usually prompt and often dramatic, particularly in regard to the systemic symptoms and head pain. When therapy is initiated prior to the onset of visual symptoms, the incidence of blindness, as has been noted, may be insignificant, and the recovery of vision may occur in the case of partial visual defects (Case 2). It is likely too, that fatalities which have resulted largely from cerebral and coronary artery occlusion will be less common in adequately treated patients. Hollenhurst recommended a starting dose of 300 mg. of cortisone, or its equivalent, followed by 150 to 200 mg. daily for four to six weeks, with a maintenance dose of 50 to 75 mg. daily for three to four months. Paulley observed that Prednisone 10 mg., given twice daily was usually effective in controlling the disease, while in some instances the dosage had to be increased to 30 mg. three times a day, in the acute stage of the disorder. The maintenance dose is variable but should be adequate to control remediable symptoms completely. More prolonged maintenance therapy for from six to twelve months as suggested by Paulley is advisable, as illustrated by the recurrences experienced by one of our patients (Case 3) when therapy was discontinued at the end of three and again at eight months. When therapy is discontinued prior to the subsidence of the disease process, the symptoms seem to recur within a month and necessitate immediate resumption of treatment. The use of anticoagulants in conjunction with steroid therapy has been suggested, but the satisfactory response obtained with steroids alone suggests that combined therapy is not essential.

SUMMARY

Four cases of giant cell (temporal) arteritis of the aged illustrating some important features of the disease and its treatment are presented with a review of the literature emphasizing the less well recognized aspects of the disorder and the possible absence of typical temporal arteritis. The use of steroids in suspected cases is suggested, and the importance of early and prolonged therapy is stressed.
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