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SUDDEN BLINDNESS IN CRANIAL ARTERITIS*

BY

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THAT vision may be affected during an attack of cranial arteritis is well recognized, but that loss of sight may herald the disease is less well appreciated. The purpose of this paper is to describe the visual symptoms that may occur at any stage and to describe the treatment of the ocular disturbance, which may, if begun early enough, prove successful in restoring sight and averting permanent blindness.

Usually one of the main scalp arteries is first involved, the process then spreading in random fashion to the others, to the main carotid trunks, and very constantly to the branches to the maxillae. Each artery remains inflamed for several days, during which it is extremely painful. The vessels involved appear as solid cords and may be completely thrombosed or the lumen may be reduced to a small slit. A sub-acute inflammatory process is present in the media, which, together with the internal elastica, may become necrotic. The vasa-vasorum are occluded and the intima is often grossly thickened, so that the calibre of the artery is reduced and peripheral ischaemic damage ensues. The chronic process spreads longitudinally, the vessel wall is infiltrated with lymphocytes, and a giant cell reaction may be excited. The disease only occurs in the older age groups and has not been seen under the age of 55 years.

Eye complications occur in about half of the cases. Birkhead, Wagener, and Shick (1957) describe 55 patients with cranial arteritis, 56 per cent. of whom had ocular involvement, and in 36·4 per cent. there was loss of sight of varying degree. Five of their cases were bilaterally blind and six unilaterally blind. Disturbance of vision is usually the result of occlusion of the blood vessels to the retina or optic nerve, though in some cases the oculomotor mechanisms are damaged. Patients blinded by the illness can be placed into one or two groups: those with ischaemia of the retina and those with ischaemia of the optic nerve. In the latter the clinical picture is either one of papilloedema or of retrobulbar neuritis and the differential diagnosis may be difficult. The pattern of visual field loss in cranial arteritis has not previously been stressed and the central scotomata seen with demyelinating conditions of the optic nerves have rarely been encountered.

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Meadows (1954) depicted the fields in four of his patients, in whom only small peripheral sectors were preserved.

In a number of cases eye symptoms and not headache first cause the patient to seek medical aid. Thus Parsons-Smith (1952) described fifteen cases in which the disease had presented with various signs or symptoms which included papilloedema, thrombosis of the central retinal artery or its branches, macular haemorrhages and exudates, ocular pain, and diplopia. What perplexes the physician is that these vascular catastrophes may occur in vessels which are, temporarily, the only ones to be involved, and widespread arteritis may not even be considered. This important variant from the usual clinical picture of the elderly patient with an exquisitely tender scalp who later suddenly loses the sight of one or both eyes was exemplified in the sixth case described by Birkhead and others (1956). The patient was an octogenarian who, following a cold, suddenly lost the vision in the right eye and 2 days later in the left eye. He had no pain and the temporal arteries appeared to be quite normal. Biopsy of the right temporal artery, however, revealed temporal (cranial) arteritis. In the past many such cases may have been diagnosed as senile bilateral retrobulbar neuritis.

When the blood supply to the eye is involved the attack is sudden and vicious and permanent blindness is to be expected. Meadows (1954) described twelve patients with temporal arteritis who received no special treatment after sudden bilateral visual failure: eight were left completely blind and the remaining four had only hand movements or counting fingers in one portion of one or both eyes. He referred to *post-mortem* evidence of arteritis and occlusion of the central retinal artery early in its course, before it had entered the optic nerve. He suggested that the visual loss which is accompanied by little change in the fundus oculi, apart from blurring of the optic discs, was due to involvement of the short ciliary arteries as they took part in the formation of the arterial circle of Zinn surrounding the nerve head. Marked recovery of vision occurred in only one of his cases, and in this patient vision improved from counting fingers to 6/18 in one eye, but the other eye remained blind. Meadows commented that spontaneous recovery from the painful effects of temporal arteritis may be expected, but that any improvement in vision is rare.

Anticoagulant therapy had been found effective in certain cases of occlusive disease of the retina by Duff, Falls, and Linman (1951) and a patient whose sight was affected in both eyes by cranial arteritis recovered the sight of one eye after a course of heparin (Parsons-Smith, 1952). Significantly better results have been achieved since the introduction of the adrenocorticotrophic hormone (ACTH) and the corticosteroids, though even with these preparations some published results have been disappointing. Bennett (1956) surveyed the response to hormone therapy in six of his own cases and in twenty cases reported in the literature, and he concluded that the prognosis for recovery of vision was significantly better in cases treated with cortisone.

Whitfield, Cook, Evans, and Rudd (1953) found improvement in five of their ten cases who had lost their vision for periods of 10 days or less, but in only one of their cases was cortisone given on the day that bilateral blindness commenced. She was a woman aged 66 who had been admitted to hospital because of the severity of her cranial arteritis and whose vision in each eye suddenly deteriorated to counting fingers. Cortisone was given immediately and her vision was saved, so that 6 months later the visual acuity was 6/6 in the right eye and 6/18 in the left, though the fields in both eyes were considerably constricted. Caccamise (1956) treated with a 10-day course of intravenous ACTH a patient with temporal arteritis who had been blind in both eyes for 2 days. Although progress was slow the visual acuity 4 months later in the right eye was 20/30 for distance and J.4 at 14″ with +2·5 D sph. There was only vague perception of light with the left eye. It now seems likely that the variability in the response depends both upon the length of the interval between the onset of blindness and the first dose of the hormone, and also upon the site of the arterial lesion.

Birkhead and others (1957) compared the results before and after the introduction of the corticosteroids. Since the use of cortisone the number of those bilaterally blind on discharge from hospital was no greater than the number of those bilaterally blind on admission and there were only two additional blind eyes. In contrast, in the groups seen before the advent of adrenal corticosteroid therapy, there was a 3-fold increase in the number of patients who suffered bilateral blindness during the illness and an increase in the number of blind eyes, from 16 to 24, during course of treatment. Among patients seen before hormone treatment became available, five who were blind in one eye at the time of admission became blind in the remaining eye whilst under observation, but there were no similar occurrences among patients who received hormone therapy. On the basis of their study of 250 cases in the literature, bilateral blindness can be expected in 22 per cent. of cases of temporal arteritis; it had occurred in 17 per cent. of their patients in the Mayo Clinic not treated with cortisone and in only 9 per cent. since the corticosteroids had been used.

Material

Fifty consecutive patients with temporal arteritis are reviewed in this paper. They were seen at the Western Ophthalmic Hospital over a period of 5 years and ACTH was available if vision failed. In many patients the diagnosis was not difficult, the illness running the usual course, but atypical clinical pictures were encountered. Difficulty with ancillary methods of investigation, such as the erythrocyte sedimentation rate and results of biopsy, became apparent. Too much weight should not be attached to the erythrocyte sedimentation rate, which is usually raised in the syndrome, since near to normal figures may be obtained in the earliest stages and the rise may occur later. Not infrequently, a piece of artery is examined at biopsy which shows no specific abnormality, but when a more peripheral portion of the same artery is examined typical changes are seen.

Now that the clinical features of the syndrome are so well recognized, it is not considered necessary to prove the diagnosis by biopsy, and in no case should appropriate treatment be delayed pending such investigation.

Corticotrophin (ACTH) is considered the most suitable therapeutic hormone, because patients who are affected with this disease fall into the highest age groups. With ACTH the physiological integrity of the adrenal cortex is preserved, and it is easier to wean elderly patients from the preparation than it is to withdraw the corticosteroids which produce adrenal suppression. No difficulties have been encountered in administering it and there have been no side-effects in this group. It is true that the strength of various batches of the preparation may vary, but lately it has become more stable and the recently produced brands are safe and reliable. The treatment should be continued for 21 to 28 days, during which time the patient should be in hospital or confined to his home. For the first 2 days ACTH was given intravenously, 25 units being given each day. For the third and fourth days, 50 units were given intramuscularly every 6 hours, and after that the dose was reduced to 20 units intramuscularly twice daily. During the final week the dose was gradually reduced. Care must be taken with new preparations that they are suitable for intravenous injection. In emergency, it is recommended that 5 mg. prednisolone be given by mouth three times daily, with an initial dose of 15 mg., switching over to the ACTH as soon as possible. In one of the cases described below, heparin was given synchronously with ACTH.

The fifty patients were all over the age of 55. There were twenty male and thirty female patients. In fifteen of the fifty cases, there was evidence of severe hypertensive heart disease, the blood-pressure being above an arbitrary figure of 200 mm. systolic and 110 mm. diastolic. In every case, there was eventually no doubt on clinical grounds that the patient was suffering from a diffuse inflammatory process which involved the scalp arteries, and in the majority other cranial arteries were also involved. In no case did the diagnosis depend upon a biopsy study. The presenting symptoms and signs are tabulated below:

Severe headache	11
Artery—Central retinal occlusion	6
Retinal branch occlusion	1
Haemorrhages and exudates	4
Optic nerve—Papilloedema	7
Retrobulbar neuritis	3
Optic atrophy	2
Sixth nerve palsy	5
Ocular pain	5
Cerebral lesion—Field defects	4
Central retinal vein branch thrombosis ..	2

ACTH was used in twelve of the cases, and cortisone was given in one other, this preparation having been started elsewhere when vision had failed. These thirteen cases are reported separately, since different clinical features emerge from each which may prove of value in diagnosis and treatment.

Report of Cases Treated with ACTH

Three distinct clinical groups of blindness have been treated with ACTH: five patients who presented with evidence of retinal artery disease, three with retrobulbar neuritis, and five with papilloedema.

(1) Retinal Artery Disease

Case 1, a 68-year-old female, was seen on August 2, 1956, with a thrombosis of the right central retinal artery. She had lost the sight of the right eye for 7 days. Visual acuity was 6/5 with the left eye and the right was blind. The right disc was ill-defined, with a patch of exudate below and confluent with it. There were scattered white areas around the vessels in the central area. The left fundus showed mild arteriosclerotic changes. Blood pressure 210/100. Erythrocyte sedimentation rate 70 mm./hr. She had had severe headache and local pain in the scalp when combing her hair. Eating had been difficult because of pain in the jaw, and the right carotid artery was tender to palpation. She was given a 2-week course of ACTH with no improvement in vision. On August 26, 1956, when she had been without the hormone for 10 days, the sight of the left eye suddenly failed. She then had no perception of light with the right eye and counting fingers only with the left. The left disc was blurred. The erythrocyte sedimentation rate was 25 mm./hr. ACTH was recommenced within a few hours, and continued for one month, and recovery of vision was rapid. On October 3, 1956, the visual acuity in the left eye had improved to 6/12 and J.6, though the field was very constricted; 5 months later, the visual acuity in the left eye had improved to 6/9 and J.4 corrected, but the visual field was unchanged (Fig. 1). The right eye was blind. Both discs had become atrophic.

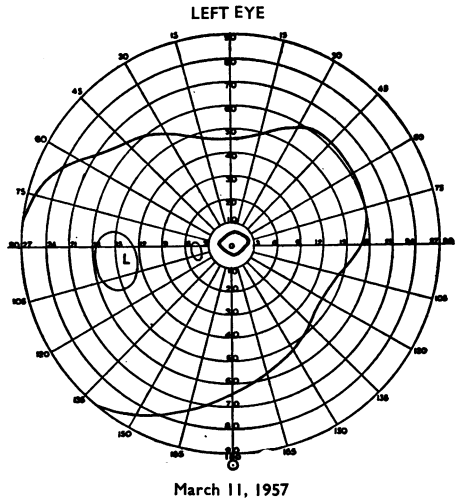

FIG. 1.—Case 1. Visual field in the left eye constricted to less than 10°. Visual acuity 6/12. 5/330 White.

The first patient with cranial arteritis, blind in one eye as a result of a central retinal artery occlusion, subsequently lost the sight in the remaining eye because of an ischaemic lesion within the optic nerve. Immediate administration of ACTH restored vision in the second eye to be involved.

Case 2, an 81-year-old female, was seen on April 25, 1955, with a thrombosis of the right central retinal artery. She had lost the sight of this eye 14 days previously. Visual acuity was hand movements in the right eye, and 6/24 in the left. The right disc was atrophic. Blood pressure 160/85. For a month, she had suffered from agonizing pain

in her temples and had not been able to eat owing to local pain in the upper jaw. She was warned to report immediately should any symptoms develop in her left eye, and was seen regularly because the pain in her temples was unusually severe. 18 days later she woke to find she was blind, the left central retinal artery having become occluded. She was admitted the same day and was given her first dose of ACTH intravenously. Visual acuity was then no perception of light in the right eye, and counting fingers in the left. 4 days later the visual acuity in the left eye had improved to 6/18, the following day to 6/12, and on the tenth day of treatment to 6/9. 5 months later, the visual acuity was hand movements in the right eye and 6/9 and J.1 with glasses in the left. 2 years later, she reported that she had been living an active life, reading books and newspapers. The left disc then showed some slight pallor, and the left temporal artery was not pulsating, whilst the right was normal.

The second patient was already under observation when the second central retinal artery became involved. ACTH was given within a few hours and she recovered the vision in this eye.

Case 3, a 78-year-old female, was seen on April 4, 1956, with haemorrhages scattered over both fundi. There was no swelling of the discs. She had lost the sight of both eyes for 2 weeks and visual acuity in each eye was perception of light only. For a month she had endured "excruciating" headache, an abnormally severe sore throat, and severe pain in the jaws. Her daughter had noted swollen blood vessels on the temples and she could not comb her mother's hair because the scalp was so tender. The patient refused to come into hospital, and was given 40 units ACTH daily, as an out-patient. 7 days later she had lost her pain and "the curtain had slowly lifted from the left eye". The vision with the right eye improved to hand movements and with the left to counting fingers so that she was able to see her way around her home. ACTH was given for 5 weeks, and the haemorrhages disappeared; the discs were only slightly pale, but no further improvement in vision took place. Even so, she was able to read the headlines of a newspaper with the left eye and could help with cleaning in the home, the daughter noticing on one occasion that her mother had seen some cigarette ash on the carpet and had brushed it up.

From the fundus appearances alone, this patient showed signs of gross hypertension with scattered haemorrhages. There was a delay of 2 weeks between the onset of blindness and starting ACTH therapy and only a slight degree of vision was recovered.

Case 4, a 75-year-old female, was seen on May 17, 1956. She had developed cranial arteritis in June, 1955, when, because of the severity of the pain, she had been placed on cortisone in another hospital with good effect. A month later, unfortunately, she forgot to renew the prescription, and within a few days had lost the sight of the left eye. A further supply of cortisone was obtained, but there was no recovery of vision. When seen in May, 1956, she had mild generalized arteriosclerosis. Blood pressure 170/85. Visual acuity was 6/60 in the right eye and no perception of light in the left, and the left disc was pale. She was then on 25 mg. cortisone acetate three times a day, and every time this dosage was reduced she thought that the vision in the right eye deteriorated, so she will have to continue with it indefinitely.

A patient taking cortisone accidentally omitted the tablets and produced an occlusion of the left central retinal artery. It has not since been possible to wean this patient from cortisone.

Case 5, a 68-year-old male, was seen on April 13, 1953, with a history of gradual visual failure in both eyes. He had extensive bilateral retinal haemorrhages and the arteries

were attenuated. In addition, he had a left-sided homonymous hemianopia which dated back to a cerebral insult sustained 18 months previously. The visual acuity in both eyes was 6/9. Blood pressure 220/120. He had active temporal arteritis and on this account was seen frequently and warned to report immediately should his vision deteriorate. His symptoms of cranial arteritis worsened and on May 11, 1953, vision failed in both eyes; the visual acuity was 6/24 in the right eye and no perception of light in the left. ACTH was begun the same day; after 3 days there was some recovery of vision, and 10 days later the visual acuity in both eyes was 6/9, but the visual fields were severely constricted. We were not able to follow this patient's further progress.

Although this man had evidence of retinal disease and when vision failed in both eyes the loss was unequal, other features were noted whilst he was recovering which suggested that the arteritis had also affected the blood supply to the visual cortex. At all events, the response to ACTH was fairly satisfactory.

(2) RETROBULBAR NEURITIS

Case 6, a 73-year-old male, was seen on November 29, 1956, with pallor of both optic discs and visual acuity of counting fingers with each eye. Blood pressure 175/100. The vision with both eyes had deteriorated suddenly 2 months before he had first attended hospital on October 3. As he was then an extremely heavy smoker, he was advised to give up cigarettes, but the visual acuity continued to deteriorate. The visual fields showed large central and paracentral scotomata (Fig. 2). His eyeballs became very tender and he developed such severe pain in his scalp that he could not brush his hair. He lost his appetite and could not masticate because of the pain in the jaw. The erythrocyte sedimentation rate was 27 mm./hr. In spite of the length of the history, it was thought that ACTH should be tried, and he was admitted to hospital on December 3, 1956. ACTH was delayed until December 10 because of an intercurrent respiratory infection, but 4 days after starting the treatment the visual acuity had improved to 6/60 in both eyes, and 18 days later, it was 6/60 with the right eye and 6/36 in the left. A month later, it had improved to 6/24 in both eyes, and by February 2, 1957, he achieved 6/9 in both eyes corrected, the fields were full, and the scotomata had cleared.

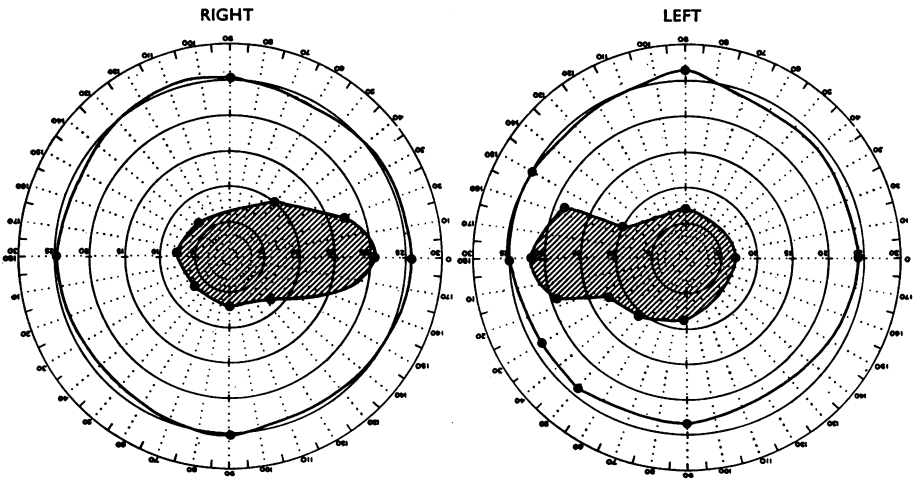

FIG. 2.—Case 6, showing large bilateral, central, and paracentral scotomata. Visual acuity 1/60 in the right eye and 1/60 in the left. 5 mm. White.

It is difficult to understand the underlying pathology in this remarkable case. There was a 17-week interval between the loss of sight and the commencement of the ACTH therapy, yet there was a good response to the injections. It is noteworthy that he had been an extremely heavy smoker and this fact may have produced spasm superimposed upon the organic narrowing produced by the arteritis.

Case 7, a 58-year-old female, was seen on February 23, 1953, with a swollen left optic disc and a history suggestive of retrobulbar neuritis. She had suddenly lost the sight of the left eye 10 days previously; visual acuity was 6/9 in the right eye, the left eye was blind. Blood pressure 190/100. Because the left temporal and occipital arteries were extremely tender to palpation, the former being swollen and thrombosed, she was warned to report immediately should any unusual symptoms develop in the right eye. 22 days after her first symptom the right eye became painful and 2 days later she was blind. She was admitted to hospital immediately. The visual acuity was hand movements in the right eye, and no perception of light in the left. Within a few hours she was given ACTH intravenously. The response to this was at first only very gradual, but on the third morning she had improved so that the visual acuity was 6/9 with the right eye, and hand movements in the left. The ACTH was continued for 3 weeks and the headache subsided. On April 27, 1953, the visual acuity was 6/9 and J.4 corrected in the right eye and counting fingers in the left. She suffered from occasional headaches throughout that summer, but by October 15, 1953, these had ceased and the visual acuity in the right eye had improved to 6/9 and J.1 corrected. 4 months later, the right disc appeared normal and the visual acuity was 6/6; the left disc was very pale and the visual acuity 6/36. Since then, the right eye has remained normal and the left has slowly improved to 6/24 on August 10, 1954, and 6/12 on March 22, 1955. She reported on October 24, 1957, that since her last visit she had remained in perfect health. The visual acuity in the right eye was 6/6 and the fundus was normal. In the left eye she could manage 6/9, but the disc was very pale. The only other clinical abnormality was a non-pulsating left temporal artery.

This, another remarkable case, presented with typical unilateral retrobulbar neuritis. Although this was associated with cranial arteritis, it was not at the time thought that ACTH could be of any benefit as vision had already been lost for 10 days. The patient was warned, however, and when the second eye failed, ACTH was given within a few hours. The expected improvement in vision took place in this eye, and at the same time, most surprisingly, the vision was also recovered in the first eye to be involved, the interval between blindness in the eye and the first dose of ACTH being over 3 weeks.

Case 8, a 74-year-old female, was seen on May 13, 1957, with a history suggestive of left-sided retrobulbar neuritis, the vision having suddenly deteriorated 7 days previously. She gave a history of a recent sensory epileptic attack involving the right side of the body. The tendon reflexes were slightly increased on that side and, as the left carotid artery was not pulsating as well as the right, it seemed likely that the symptoms in the left eye and those down the right side of the body arose from a disturbance within the left carotid tree. The visual acuity in the right eye was 6/6 corrected, and in the left 6/60 corrected. The left optic disc was pale and there was no significant abnormality in the visual fields. The erythrocyte sedimentation rate was 80 mm./hr. Blood pressure 180/90.

The patient was kept under observation and a month later developed symptoms of diffuse cranial arteritis. She could not comb her hair because of pain or put her head on the pillow. The left occipital artery was swollen and exquisitely tender. Although she had been warned about the possibility of visual loss, she did not return immediately when this occurred in both eyes. She was very inaccurate and it is not known precisely how

long she had been blind, but the period was at least a fortnight, the right eye having failed slightly before the left. A course of ACTH was begun on June 18, 1957, 43 days after the first symptom had been noticed in her left eye. The visual acuity was then counting fingers in both eyes, but the only abnormality in the fundi was a pale left optic disc. 9 days later there was some slight improvement to 6/60 in the right eye and counting fingers in the left. A month later, the acuity in both eyes was 6/60, and a year later, it was 6/36 in both eyes.

This case has some interesting features: the retrobulbar neuritis preceded the symptoms of cranial arteritis by a month; the disease recurred in the eyes so that vision was lost in both; there was certainly response to ACTH in spite of an interval of about 2 weeks between the onset of blindness and starting treatment. A fourth remarkable point is the extraordinary degree of composure that these patients exhibit when they lose their sight, a feature that has been stressed by other authors.

(3) Papilloedema

Case 9, a 67-year-old female, was seen on April 12, 1955, with right-sided papilloedema, with beading of the veins and small disc haemorrhages. The left disc was slightly blurred. She had lost the sight of the right eye for 7 days and the left eye had failed that morning. The visual acuity in the right eye was no perception of light, and in the left counting fingers. Erythrocyte sedimentation rate 76 mm./hr. Blood pressure 138/70. She had severe symptoms of temporal arteritis, and ACTH together with heparin was commenced immediately. The visual acuity in the left eye slowly improved, so that 12 days later she could read J.8, after another 3 days J.4, and 4 days after that J.2 and 6/9 with corrections. Meantime the visual acuity in the right eye improved to hand movements. The improvement which took place in the visual field of the left eye is shown in Fig. 3. 6 months later she could appreciate hand movements with the right eye and in the left the visual acuity was 6/9 and J.1. Both optic discs had then become very pale and for some unaccountable reason her blood pressure had risen to 250/120 on August 12, 1957.

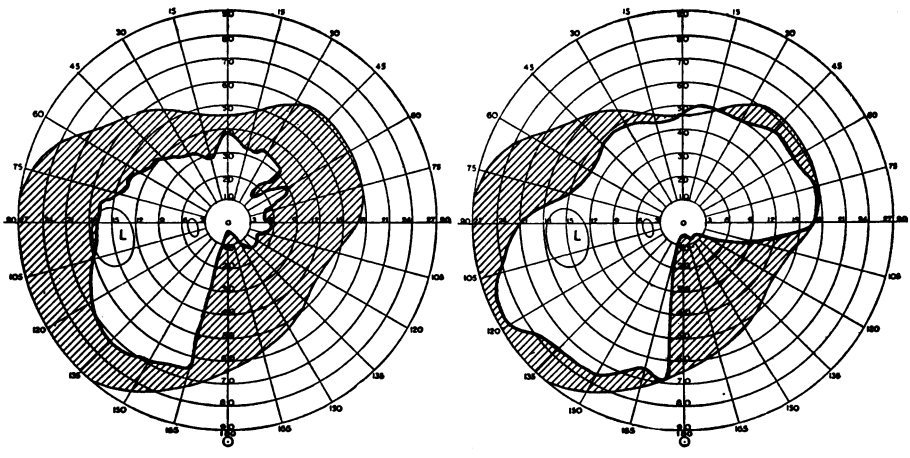

FIG. 3.—Left eye of Case 9.

(*i*) Visual acuity counting fingers. 20/330 White. (*ii*) Visual acuity 6/9. 3/330 White.

This case was already under observation with acute unilateral papilloedema and haemorrhages when the second eye was affected. ACTH was given within a few hours and the vision was recovered.

Case 10, a 73-year-old female, was seen on June 21, 1954, with left-sided papilloedema. She had lost the sight in that eye 4 days previously, and had suffered for a month from severe pain in the left occiput and left temple, so much so that it hurt to put her head on a pillow, and the pain had radiated into the jaw and made eating difficult. Both temporal arteries were prominent, pulseless, and extremely tender. The visual acuity in the right eye was 6/9 with some constriction of the visual field, and counting fingers in the left. Erythrocyte sedimentation rate 7 mm./hr. Blood pressure 200/110.

She was warned to return immediately if any disturbance developed in the right eye, and 27 days after her first attendance she woke to find that the sight in the right eye had deteriorated. She was not seen in hospital until the following morning, when the visual acuity was perception of light in the right eye, and the left eye was quite blind. The right disc was swollen. ACTH was begun within 36 hours of the first symptom in the right eye and there was some response. Within a few days, the visual acuity in the right eye had improved to counting fingers, she was able to get about the ward without running into obstructions. 2 months later, she was able to see enough to walk to the town and do her shopping and could just see the headlines of a newspaper. On November 14, 1957, the visual acuity in the right eye was hand movements at 10′, and with a magnifying glass she could just manage N.36. The left eye was quite blind.

This case shows that delay in instituting ACTH, by as little as 36 hours, may make it less effective. Even so, the patient made slight improvement.

Case 11, a 75-year-old male, was seen on July 11, 1957, with right-sided papilloedema and loss of sight 5 days previously. The left eye was not affected. For a month he had suffered from severe headache which he had associated with "sudden swelling of the temple veins". He had lost his appetite and had pain in the right maxilla, throat, and tongue. The right eye was quite blind and the visual acuity was 6/9 in the left eye. Erythrocyte sedimentation rate 81 mm./hr. Blood pressure 185/90. ACTH was started 5 days after the loss of sight and within a week the pain had gone and the swelling of the disc had subsided. For years he had had generalized rheumatism and this was temporarily relieved with ACTH. No recovery of vision took place in the right eye, but the left remained unaffected. On November 18, 1957, he was in good health. Neither temporal artery was pulsating, but there was no tenderness. The right disc was pale, and the eye blind whilst the visual acuity was 6/9 in the left eye.

The institution of ACTH 5 days after the onset of blindness was effective in causing the papilloedema to subside and the rheumatism to abate, but no recovery of vision ensued.

Case 12, an 82-year-old male, was seen on May 17, 1954, with bilateral papilloedema. He had lost the vision of the left eye for 12 days and of the right for 10, and had endured the symptoms of temporal arteritis for a month. The visual acuity in the right eye was hand movements only, and the left was quite blind. Blood pressure 135/80. ACTH was started 2 days later, and though no improvement took place in the left eye, achieved 6/60 after 10 days with the right.

ACTH was started 12 days after blindness and there was some slight recovery.

Case 13, a 76-year-old female, was seen on January 8, 1957, with bilateral papilloedema. She had lost the sight of the left eye for 7 days and that of the right for 4. The visual acuity in the right eye was 6/60, and in the left perception of light. Blood pressure

220/80. Her temporal arteries were swollen and inflamed, she had had difficulty with eating owing to pain in the jaw, and severe pain behind her ears which she had thought was due to mumps. She had not been able to brush her hair because of tenderness over the scalp. ACTH was started on January 10, 1957, when the visual acuity in the right eye was perception of light and the left was quite blind. Only slight improvement resulted from the treatment; 2 months later she could count fingers with the right eye, but the left eye which was the first to be affected remained blind. Even with the slight amount of sight remaining to her she was able to get around her room and her family were surprised how much she was able to see.

In this case of bilateral papilloedema ACTH brought only slight improvement in the eye which had been blind for 4 days, and the other eye which had been blind for a week was not influenced by the treatment.

Discussion

Most symptoms of cranial (temporal) arteritis clear spontaneously, but when the blood supply to the eyes is involved the sequelae are likely to be permanent. In this series of fifty patients with cranial arteritis who attended an eye hospital, the course of the illness extended in most cases over a period of about 3 months by which time the pain had usually subsided, and in cases of diplopia the oculomotor lesion had resolved. No spontaneous improvement was expected, nor did it take place, in patients who had been blinded by the disease and received no specific treatment.

A study of the eleven patients in whom the arterial blood supply to the retina was involved and of the twelve patients who presented with signs of optic nerve disease has shown that no further deterioration of vision took place in those who were treated with ACTH. Indeed, in several cases, a surprisingly good response to the hormone was achieved. In all, 33 blind eyes were encountered. Nine of these were not given ACTH and not one improved. In 24 eyes in which vision had been reduced to counting fingers or less, a course of ACTH was prescribed. Sight was restored in eleven of these eyes, and in a further four sufficient sight was regained to enable the patients to get around in the home and look after themselves. No patient who received ACTH progressed to complete bilateral blindness.

It is apparent that the interval between the onset of blindness and the first dose of ACTH should be as short as possible. Every hour of delay worsens the prognosis and the condition must be regarded as an acute emergency. Study of the literature shows that, when poor results followed the use of corticosteroids, there had generally been delay in prescribing them. If the hormone can be given within a few hours of the onset of blindness, vision is likely to be regained. In this series, six eyes treated within 12 hours with ACTH all recovered. In the seventh case, one of papilloedema, there was a delay of 36 hours and recovery was only slight. This urgency of prescribing ACTH applies equally to each of the three clinical groups, though unexpectedly good results were achieved in those with retrobulbar neuritis in spite of prolonged delay.

An important clinical feature has emerged from this study, namely that ocular symptoms may antedate general cranial arteritis by weeks or months. The patient reported by Birkhead and others (1957), who had lost the sight of both eyes suddenly and had had no pain yet showed biopsy evidence of temporal arteritis, has already been referred to. Five patients in the present series developed eye symptoms before the onset of the cranial arteritis. Two patients had 6th nerve palsies. In one the arteritis developed 3 weeks later, whilst in the other there was an interval of 6 months. In a case of right-sided papilloedema in a man of 66, general arteritis developed a month later. In a woman of 77, who sustained a thrombosis of the right central retinal vein on January 28, 1953, the eye became extremely painful 2 months later and after a further month the classical picture of cranial arteritis developed. The visual acuity when first seen was hand movements only, but after 3 months she became quite blind. In Case 8 the retrobulbar neuritis had antedated the generalized inflammatory condition by a month.

The annual incidence in the County of Middlesex of bilateral blindness of sudden onset due to vascular disease in patients over the age of 55 years is about seven per million. Many of these cases are due to cranial arteritis, but the exact proportion is not known. Bearing in mind that ocular symptoms can occur as an isolated manifestation of cranial arteritis, though general symptoms of this will appear later, we have made a control study of a group of elderly people who suffered from acute vascular lesions of the eye. They were observed over the same 5-year period as the cases of cranial arteritis. Thirty patients over the age of 60 years had had an occlusive insult within one central retinal artery. None of them showed any other evidence of local disease of the eye or orbit. There was one significant difference of medical interest between the two groups of cases. In those with cranial arteritis, 30 per cent. had co-existing severe hypertensive heart disease, and in the control group 66 per cent. suffered from gross hypertension. Each control case was reviewed after a period of 2 years, but not one developed symptoms of cranial arteritis nor did any one develop a vascular catastrophe in the opposite eye.

Since it may be impossible to differentiate within a few hours of the onset of blindness between the effects of cranial arteritis and those of isolated occlusive incidents in the blood supply of the eye, and since it has been shown that ACTH produces dramatic results if given immediately after an insult in cases of temporal arteritis, it is now recommended that all such elderly patients should be given this hormone at once, even if only one eye has been involved. If ACTH is not available, prednisolone 15 mg. should be given immediately and the patient sent into a special centre as soon as possible. If there has been no improvement after a week of ACTH therapy, the drug can be discontinued, as the ocular lesion is then more likely to be an isolated one and not part of the diffuse syndrome of cranial arteritis. It is neither practical nor advisable to give the hormone in cranial arteritis

before visual symptoms appear, but it should be held in reserve for the moment when the blood supply to the eye becomes impaired.

Summary

(1) Cranial arteritis may present with central retinal artery occlusion, retrobulbar neuritis, or papilloedema.

(2) Each of these conditions is likely to leave the affected eye blind if untreated.

(3) ACTH given on the same day as blindness has restored sight in every eye so treated.

(4) ACTH may prove effective after a longer period of blindness in cases of retrobulbar neuritis.

(5) It is recommended that all occlusive incidents in the blood supply to the retina or optic nerve in elderly people who have no other evidence of ocular disease be treated with a course of ACTH.

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