Visual Loss Associated with Fusiform Enlargement of the Intracranial Portion of the Internal Carotid Artery

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Our purpose is to discuss a syndrome which is characterized by progressively diminishing visual acuity and peripheral vision in one or both eyes after the age of 30 years. At exploratory craniotomy or necropsy, the terminal segment of the internal carotid artery is found to be enlarged and to displace the ipsilateral optic nerve upward against the superior rim of the optic foramen.

Caramazza described the clinical findings of 2 unconfirmed cases in 1932. Linear sellar calcifications were noted on the skull films, and a correlation with an arteriosclerotic process was suggested. In 1934, Schloffer reported his experience with a 65-year-old woman who complained of a loss of vision in the right eye of 5 years’ duration, a recent visual defect on the left of 6 weeks, plus headache, dizziness, and diplopia. Examination revealed decreased vision and field defects in both eyes. The right disc was atrophic and the left was raised with 2 dipters of papilledema. Because of the apparent Foster Kennedy syndrome a frontal tumor was diagnosed. A right frontal craniotomy was performed and no tumor was found, but the right internal carotid artery was enlarged and compressed a thinned optic nerve upward. The left nerve was not visualized. At post-mortem, it was found that both internal carotid arteries and both optic nerves were equally involved. It was felt that decompression of one or both optic foramina might have been beneficial.

In 1938, Gleez described 3 cases that had come to his attention with the diagnosis of cerebral sclerosis. These patients were not explored, but there were certain features of the Foster Kennedy syndrome and he suggested the possibility of a bulbous carotid artery syndrome.

Adason reported 2 cases in 1941; the main clinical findings were defects in visual acuity and fields. At the time of exploration, the pathological complex under consideration was encountered and the optic foramina were unroofed. In one instance, the defects were not advanced and there was improvement after the operation. The other patient’s defects were extreme and no significant recovery was noted. In 1942, Yaskin and Schlezinger, published a report of a patient with loss of visual acuity associated with optic atrophy in one disc and papilledema in the other. The surgical procedure was limited to an exploration which revealed a bulbous carotid artery and a compressed optic nerve. There was no improvement in vision.

In 1944, Tassman also reported a case with features of the Foster Kennedy syndrome. The optic foramen was decompressed on one side; however, visual acuity deteriorated and the field defects progressed postoperatively. Taptas reported a case with bilateral visual defects and optic atrophy in 1948. Some improvement was reported after operation although no attempt was made to decompress the distorted optic nerves.

Five cases, (4 of which were verified by exploration) were described by Ley in 1950. The surgical procedures varied considerably and there was only one instance of improvement.

During the last few years, the following 4 cases have been seen by various members of
the Division of Neurological Surgery at the Johns Hopkins Hospital.

Clinical Material

Case 1 (Un. Mem. Hosp., #338522). M.L., a 38-year-old white woman, was first seen by an ophthalmologist in February, 1955, and complained of visual loss in the right eye of 8 months' duration. The visual acuity on this side was 20/40, the disc was pale and the visual field was generally constricted. Later, in October, 1955, it was found that only light perception remained on this right side. The visual field was further constricted and the upper nasal quadrant was practically absent. At this time she also complained of right frontal head pain. A carotid arteriogram and a pneumoencephalogram were carried out and were reported to be within normal limits.

She was first seen by a neurosurgeon in December, 1956. At this time she was blind in the right eye. It was found that the right pupil was moderately dilated and did not respond to light directly but did consensually. The right disc was described as chalky white. The left eye was normal in all respects. X-rays of the skull (including views of the optic foramina) were negative. It was presumed that an intracranial neoplasm was present and was impinging upon the right optic nerve.

Operation. On February 29, 1957, a right frontal craniotomy was carried out. There was no neoplasm, but the right optic nerve was found to be flattened and elevated by an enlarged internal carotid artery. This optic nerve was not functioning and was partially divided to provide a better exposure. The artery did not appear thin-walled. The optic chiasm, left optic nerve, and the left internal carotid artery appeared normal.

Postoperative course. An arteriogram was carried out on the right side after the operation and was again normal. The patient was last seen on October 5, 1959, when her condition was essentially unchanged.

Case 2 (J.H.H., #709564). W.K., a 66-year-old white man, was seen at the Johns Hopkins Hospital in 1955 with a defect in peripheral vision. At the onset, he awoke one morning and noted that the lower half of the right field was lost. Later, he felt that visual acuity in both eyes had diminished. There were no other symptoms. He had been seen at the Wilmer Institute about 1 week after the onset of symptoms; the left eye was found to be normal but there was papilledema of the right disc and the lower fields were constricted. Visual acuity however was normal. The field defect was described by Dr. Frank Walsh as obliterating the lower field on the right and coming up a bit above the horizontal plane especially on the nasal side. The right carotid pulse was greater than the left. The remainder of the neurological examination was negative. Blood pressure was 140/85 and the pulse 74. A systolic murmur was heard and various skeletal deformities of rheumatoid arthritis were recorded.

The past medical history was not pertinent. The family history revealed that the patient's father had died from a stroke; 2 brothers and 1 sister had died with heart disease; and 1 sister had died with a renal disorder. Urinalysis showed 1+ albumin and 3–4 white blood cells per high power field. The peripheral blood counts and the serum electrolytes were normal. The chest film was clear. Initially, it was thought that a right carotid arteriogram indicated some enlargement of the intracranial portion of the internal carotid artery. However, in retrospect these films appear normal. A pneumoencephalogram showed only a slight increase in the amount of air over the convexity of the frontal regions. The cerebrospinal fluid protein was 14 mg, per cent, and the complement fixation and colloidal gold tests were negative.

Operation. A right frontal craniotomy was carried out. The right optic nerve appeared flattened and pushed upward by a large, sclerotic carotid artery. The nerve however was said not to appear "tight in the optic foramen." The chiasm and the left optic nerve appeared normal. There was no evidence of a neoplasm and a decompression was not performed.

Postoperatively the papilledema subsided and the (right) disc became pale. Visual acuity remained unchanged.

Case 3 (BCH, #262549). This 55-year-old colored man noted "silver dots" before his eyes and then a sudden loss of vision on the right side. This occurred on April 3, 1959, as he was working as a laborer. He was seen at the Baltimore Eye, Ear and Throat Hospital on the following day where a marked loss of vision in the right eye was recorded. His condition remained unchanged for about 4 to 5 weeks. At the end of this time the patient developed polydipsia and polyuria. He lost approximately 25 lbs. in the next 4 weeks. His physician first recorded glycosuria 2 weeks before his admission. He was placed on a diabetic diet and given tolbutamide at that time. On June 11, 1959, focal seizures of the left side of the face appeared; these persisted for 2 to 5 minutes, recurred every 15–20 minutes and were preceded by numbness of the face. They did not respond to anticonvulsant medication; he was therefore admitted to Baltimore City Hospital on June 15, 1959.

Examination. He was apathetic but orientated. The right pupil was larger than the left and a slight weakness in the left side of the face was
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The right disc was pale and the vessels were sclerotic. He could only distinguish large objects on the right side. Visual acuity was also diminished on the left side; however, this loss was in part accounted for by the presence of a cataract. Blood pressure was 170/100. Four positive sugar was found in the urine. Fasting blood sugar was 300 mg. per cent; the blood picture and the serum electrolytes were normal. The cerebrospinal fluid protein was 34 mg. per cent. The chest and skull films were negative. A right carotid arteriogram was carried out and appeared normal save for a diffuse enlargement of the whole internal carotid artery and the first part of the middle cerebral artery.

Operation. On June 25, 1959, the chiasm was examined via a right frontal approach. The right internal carotid artery was fusiformly enlarged and the right optic nerve was markedly displaced. This nerve was particularly thin. The left optic nerve was not adequately visualized; a decompression was not performed. In this instance, it appeared that the nerve compression was significant but that it was only one of many manifestations of involvement of the carotid system. The patient's general condition did not improve and he died on June 29, 1959.

Autopsy. The brain, the optic nerves and chiasm, and the intracranial portion of the internal carotid arteries were photographed and are depicted in Fig. 1. The walls of the carotid vessels were extremely thick; this seemed quite clearly to be an atheromatous process. There was no indication of an aneurysmal dilatation. We do not have sections of these vessels; but slides of their branches and the basilar vessels, and the aorta and renal and pulmonary vessels indicate very marked arteriosclerosis.

Discussion

If one analyses the verified cases of Adson, Ley, Schloffer, Taptas, Tassman, Yasckin and Schlezinger and ours, several features are prominent.

1. Age of Onset. The onset of symptoms in 6 cases was in the 4th decade of age; 2 began in the 5th, 3 in the 6th, and 3 in the 7th. Ley's analysis may be somewhat misleading in that he recorded the age at the time of exploration rather than at the time of onset.

2. Interval from Onset to Exploration. Eight patients were explored within the 1st year after onset of symptoms, 2 within the 2nd year, 2 in the 5th, 1 in the 6th, and 1 in the 7th. There does not appear to be a consistent relationship between the duration of the symptoms and the severity of the visual defects.

3. Sex Incidence. In the 14 cases under consideration there were 8 men and 6 women.

4. Initial Complaints. Six of the 14 patients complained of progressive loss of vision in one eye followed later by loss in the
other eye, 2 of only unilateral visual loss, 2 of simultaneous bilateral loss, and 2 of field defects. Headaches were noted in 7 cases, dizziness in 2, diplopia in 1, paresthesiae in 1, ataxia in 1, and vasomotor changes in 1.

5. Examination of Visual Fields. Defects of the superior fields were seen in 7 cases. Two of these had an altitudinal defect in one eye and a dissimilar defect in the other. Of the remainder, 2 were unilateral and 3 bilateral. Bitemporal defects were observed in 1 case, bitemporal scotomata in 1 case, unilateral field constriction in 2 cases, and bilateral field constrictions in 1 case. In 2 instances the fields were not reported.

6. Optic Discs. Three patients had optic atrophy in one eye and papilledema in the other; 1 had unilateral papilledema alone; 3 had unilateral optic atrophy alone. There were 2 instances in which bilateral optic atrophy was found. Three had atrophy in one eye and venous stasis in the other. In 1 instance there was atrophy co-existing with papilledema in both eyes.

7. Other Neurological Signs and Symptoms. Various cranial nerves were involved in 5 cases; these included the 1st, 3rd and 7th nerves. The sensorium was dulled in 1 instance; weakness of an extremity was noted in 1 case and focal seizures in another.

8. Associated Illnesses. Eight patients were suffering from known arteriosclerosis and/or hypertension, 3 had diabetes mellitus.

In brief this condition afflicts adults of either sex who are over 30 years of age, usually with some manifestation of arteriosclerosis. The initial complaint is likely to be diminished vision. The rate of deterioration is such that many patients will have an intracranial exploration within the first 2 years of the illness. The visual acuity and visual fields as well as the optic discs are abnormal on one or both sides.

Etiology. Fusiform dilatations of the carotid arteries have been reported in necropsy material from: (a) cases with apparently no clinical findings, (b) cases where vascular insufficiency and encephalomalacia were prominent, and (c) cases where the major manifestations were apparently due to ischemia and subsequent atrophy of certain of the cranial nerves. In all of these groups an arterio-sclerotic lesion has been demonstrated. In one instance (seen by Ley) an unusual lesion was reported. There was no evidence of a sclerotic process. The vessel was thin, with multiple small dilatations externally and several folds within. No atheromatous deposits were felt or seen. The sections revealed hypoplasia of the muscularis and intimal degeneration. Ley considered this latter finding to be secondary to a primary change in the muscularis. This suggests that (in addition to saccular aneurysms), at least two distinct processes within segments of the terminal internal carotid artery may lead to a significant displacement of the optic nerve. However, the presence of one or more atheromatous deposits as part of a generalized process is of prime importance in the majority of instances.

The negative angiographic findings are of particular interest. Our films were unrevealing (although in one instance the internal carotid artery appeared quite broad throughout its course). These findings suggest that although plaques are commonly seen in this segment, the gross enlargement may be missed entirely.

The mechanical effect seems quite clear. In those cases reported here, the encroachments were marked. The optic nerves in our necropsy specimen (Fig. 1) were, for example, very attenuated. Similarly, the altitudinal field defects may be explained by a pressure effect on the nerve at the foraminar rim. However, compression of or arteriosclerotic lesions within the ophthalmic artery may be contributory, and on the basis of local hypoxic swelling, account for the papilledema. Finally, a single, large atheromatous plaque may exert local pressure on the nerve without significant displacement. Each of these factors is fully covered by Walsh.

Treatment. Exploration of the optic nerves and chiasm (without decompression) was carried out in 7 instances. Of these, 1 patient
was said to have improved; 3 were unchanged; 1 worsened; 2 died in the immediate post-operative period. Unroofing of the optic foramen on the involved side (or sides) was done in 3 cases. One patient improved, 1 was unaltered and in 1 instance the follow-up was too brief for adequate evaluation. In 4 cases the foramen involved was unroofed and the internal carotid artery was ligated in either its intra- or extracranial course. One patient improved and 1 succumbed. Two patients worsened—one with a contralateral hemiplegia. The ligation procedure seems unwarrantable in the majority of cases where there is generalized arteriosclerotic change. The complications noted in the cases reviewed are unduly high. A meaningful evaluation of the influence of decompression of the bone has not been possible because of the small number of cases. This maneuver may also be hazardous in this condition because of the underlying pathology. The deroofing procedure was successful in Adson’s case and, while our position is basically conservative, we believe that it should be given a further trial in selected cases. In many instances, it is likely that with proper recognition, explorations will be rejected.

Summary

1. The findings of 4 patients with surgically verified vascular compression of one or both optic nerves are reported. Bulbous internal carotid arteries have been found and saccular aneurysms excluded.

2. The finding of 10 further patients are summarized from the literature. In one of these a fusiform aneurysmal dilatation (with no sclerotic element) was found.

3. These patients come to the doctor because of a loss of visual acuity in one or both eyes. The optic atrophy and field defects usually observed are often associated with wide-spread arteriosclerotic changes.

4. In our experience carotid arteriograms have not contributed to the diagnosis.

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References


