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 diopters 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, Glees 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.

Adson 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 headache. 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 20, 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 thick-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 “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 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, #202549). 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.