Non-Tumor Causes of the Foster Kennedy Syndrome*

NORMAN J. SCHATZ, M.D.,† AND J. LAWTON SMITH, M.D.

Department of Ophthalmology, University of Miami School of Medicine, Miami, Florida

The combination of ipsilateral optic atrophy and contralateral papilledema was emphasized as a syndrome characteristic of basal frontal tumor by Foster Kennedy in 1911. The purpose of this report is to alert neurologists and neurosurgeons to certain pitfalls in the diagnosis of the Foster Kennedy syndrome by illustrating nonsurgical causes of similar ophthalmoscopic findings.

Report of Six Cases

Case 1. This 64-year-old white woman awoke on the morning of May 1, 1965, with a dark spot in front of her right eye. Her physician stated that she had a ruptured retinal blood vessel. Vision remained unchanged until 4 months later when she noted loss of vision in her left eye. There was no pain or headache. She was admitted to another hospital where examination showed atrophy of the right optic disc and swelling of the left. Because of the ophthalmoscopic picture of a Foster Kennedy syndrome, and in spite of negative contrast studies, an exploratory craniotomy was performed, which was negative.

Examination. The patient was seen for the first time in this office 1 month postoperatively on October 21, 1965. The corrected vision was 20/25 in the right eye and 20/200 in the left eye. The pupils were sluggishly. Visual fields showed a superior altitudinal defect in the right eye and a large central scotoma in the left eye (Fig. 1). Ophthalmoscopy revealed altitudinal optic atrophy below in the right eye (Fig. 2). The left disc was slightly swollen, but pale, with a small hemorrhage above the disc, and narrowed vessels (Fig. 3).

Comment. This patient with loss of vision had optic atrophy in the right eye and a swollen disc in the left. A basal frontal mass was excluded by negative contrast studies and subsequent negative exploratory craniotomy. The diagnosis was ischemic optic neuritis (pseudopapillitis vasculaires). She now shows bilateral optic atrophy, and her vision has improved to 20/15 in the right eye and 20/100 in the left.

Case 2. This 54-year-old white man was admitted in 1963 to the Coral Gables Veterans Administration Hospital complaining of blurred vision in the right eye. He had had poor vision in the left eye for many years. Corrected acuity was 20/30 in the right and 20/200 in the left eye. The right disc was swollen with one to two small hemorrhages seen at the nerve head margin. The left disc revealed old optic atrophy. Visual field examination disclosed an enlarged blind spot in the right eye and a dense central scotoma breaking out through an altitudinal defect in the left eye. The eyes were white. During the next few weeks of observation, vision in the right eye began to deteriorate. Because of the possibility of a Foster Kennedy syndrome, bilateral carotid arteriograms were done, and later exploratory craniotomy was performed. No mass lesion was found intracranially.

Examination. The patient was first seen in this office several months later when recalled for complete re-evaluation. At that time a meticulous history revealed that he had been raised in a brothel in Baltimore, and that at 9 years of age he had been treated with heavy metal injections for several months. Biomicroscopy revealed two or three small ghost vessels in the upper temporal cornea of the left eye. Dilated indirect ophthalmoscopy revealed evidence of old scarring and pigmentation in the peripheral fundus in both eyes. Both discs were now pale with altitudinal atrophy, particularly in the right eye. Serum was submitted to the Venereal Disease Research Laboratory in Atlanta, Georgia, which revealed the Treponema pallidum immobilization test (TPI).
Fig. 1. Visual fields of Case 1. Peripheral fields show the nerve fiber bundle defect. Central fields show an altitudinal hemianopia in the right eye with 20/25 vision. Vision in the left eye was 20/100 with a central scotoma.

to be nonreactive, but the fluorescent antibody absorption test (FTA-ABS) was reactive. The patient was treated with 20 million units of penicillin.

Comment. This 54-year-old man developed optic neuritis in the left eye at about 30 years of age, which left him with poor vision and optic atrophy in that eye. When seen more than 20 years later he complained of decreased vision in the right eye. Visual acuity was 20/30 in the right eye, and the disc was swollen and had peripapillary hemorrhage. As the Foster Kennedy syndrome could not be excluded, exploratory craniotomy was performed which was negative for a mass lesion. Later, a more careful history
led to the diagnosis of optic neuritis in congenital syphilis.

**Case 3.** This 17-year-old girl had no visual complaints until February, 1962, when she fell from a tree and fractured her right shoulder; she was not unconscious and had no eye complaints. Three days later, aching pain was noted in the left eye followed shortly by precipitous loss of all vision in this eye. Skull x-rays and lumbar puncture at that time were normal. Steroids were given, and the vision improved. One week later, vision became hazy in the right eye but cleared in a few days. Subsequent recovery in both eyes was complete.

She remained well until 2 years later, when a sudden loss of vision occurred in the right eye with swelling of the optic nerve. There were no associated neurological complaints. With steroid treatment, vision improved in 3 weeks.

She had been unable to abduct the right eye since birth.

**Examination.** The first of a series of examinations, in December, 1964, showed the visual acuity to be 20/15 in both eyes. There was a typical Duane's retraction syndrome on the right. The visual fields were full. Ophthalmoscopy showed optic nerve pallor in both eyes. The patient was next seen a year later, 1 month after having had an episode of visual loss in the right eye. At that time visual acuity was 20/20 in both eyes. On April 4, 1966, pain developed behind the right eye, and vision was reduced to finger counting at 1 to 2 feet eccentrically. The left eye vision was 20/15. Ophthalmoscopy revealed a definitely swollen right disc; the left disc was pale and atrophic as before. At this time she had the ophthalmoscopic picture of the Foster Kennedy syndrome.

**Comment.** This 17-year-old girl had recurrent optic neuritis, three bouts on the right and one on the left. The ophthalmoscopic picture was that of a Foster Kennedy syndrome, but the clinical picture was not that of neoplasm. The diagnosis was optic neuritis.

**Case 4.** This 17-year-old girl had noted poor vision in the left eye for about 5 years. There were no headaches and no associated neurological complaints. Two years before the onset of the visual loss, she had been dragged from an amusement ride but did not become unconscious.

**Examination.** Visual acuity was 20/20 in the right eye and 16/200 in the left eye. Eye movements were normal. The left pupil was slightly larger than the right and showed a definite Marcus Gunn pupillary phenomenon; visual fields were full in the right eye. In the left eye there was a dense central scotoma breaking out of the blind spot with...
a definite upper altitudinal component. Ophthalmoscopy of the right eye was normal but for “pseudoneuritis” of the optic disc. The left fundus revealed an atrophic disc with narrowed arterioles. Indirect ophthalmoscopy disclosed the peripheral changes of traumatic chorioretinitis.

Comment. The presence of a slightly blurred disc with good vision in the right eye and definite optic atrophy on the left with a central scotoma raised the question of the Foster Kennedy syndrome in this case. The changes in the peripheral fundus and the stable course after follow-up established this as traumatic peripheral chorioretinitis due to optic nerve injury from a fall 7 years earlier.

Case 5. This 8-year-old boy was seen as an emergency because visual loss and optic atrophy of the left eye were noted at a routine examination. The birth weight and development were normal. His motor development was normal, but he had had some difficulty in school. A routine examination 2 years earlier had shown vision to be normal in both eyes. Ten months before we saw him he had been struck by a car and thrown up on the windshield. He did not become unconscious and sustained only abrasions over the lower trunk; hospitalization was not required.

Family history revealed that an older brother had been institutionalized with mental retardation and seizures, and a paternal uncle died at 3 years of age from a convulsive disorder.

Examination. The visual acuity was 20/15 in the right eye and 5/200 in the left. The visual fields were full in the right eye, but there was a dense central scotoma on the left. Ophthalmoscopy showed that the right eye was normal except for slight blurring of the disc. The left disc showed definite optic atrophy. In the periphery there were old, discrete chorioidal scars.

Comment. The ophthalmological picture in this case was that of a Foster Kennedy syndrome. The differential diagnosis was between traumatic optic atrophy or an intracranial lesion.1 Skulx x-rays and brain scan were normal. The boy was followed for 3 years and showed no change. The clinical diagnosis of traumatic optic atrophy and choroiditis was made. The slight blurring of the right disc was interpreted as pseudopapilledema.

Case 6. This 60-year-old white woman was seen on July 17, 1965, because of sudden loss of vision in her right eye of 8 days’ duration. Some pain in the right temporal region had been present since the onset of the visual loss. There were no other complaints. There was a past history of a goiter since 1946.

Examination. The right eye showed no light perception; acuity in the left eye was 20/30+2. The right pupil was amaurotic and dilated poorly to cocaine; the optic disc was pale and swollen with several flame-shaped and linear hemorrhages around its margin (Fig. 4). The arterioles were moderately narrow. Ophthalmoscopy of the left eye showed two or three hemorrhages between the disc and macula (Fig. 5).

General physical examination was normal except for an enlarged, irregular, multinodular, firm, thyroid gland. Because the sedimentation rate was 31 mm/hr, a right temporal artery biopsy was done, but showed no evidence of arteritis. The patient was treated with 80 mg of prednisone daily without improvement. During hospitalization she was seen in medical and surgical consultation. Because of the pressure on the trachea, the possibility of carcinoma, and the cosmetic appearance, a thyroideectomy was performed on July 26, 1965. On the third day after surgery, tracheal edema resulted in cardiac arrest, and the patient died.

Postmortem examination. A complete postmortem examination, including the brain, was performed. This revealed severe coronary arteriosclerosis with ventricular hypertrophy and myocardial fatty infiltration. There was acute laryngeal edema and pulmonary congestion. Histological examination of the thyroid specimen revealed Hashimoto’s disease.

Neuropathological examination of the brain showed no gross or microscopic abnormalities. Both eyes were examined histologically and were normal except for the optic nerves and adjacent retinae. The right optic nerve head was edematous and showed hemorrhages. There was serous detachment of the retina extending nasally and temporally from the disc to involve the macula.
Non-Tumor Causes of the Foster Kennedy Syndrome

Some gliosis of the nerve fiber layer of the retina was seen (Fig. 6). The blood vessels of the right optic nerve were surprisingly normal. The left eye showed swelling and some gliosis of the optic nerve head. There was hemorrhage of the retina adjacent to the disc temporally (Fig. 7). The optic nerves in cross section appeared normal (Fig. 8).

Comment. This case, to our knowledge, is the first reported histopathological study of ischemic optic neuritis. The funduscopic picture commonly resembles that of the Foster Kennedy syndrome. The pathogenesis of ischemic optic neuritis has not been clearly established. The following evidence suggests that it is due to small vessel disease of the optic nerves: 1) arteriograms performed in several of these cases have shown no notable abnormalities of the major vessels; 2) there is a high incidence of hypertension and diabetes; and 3) the visual field defects and subsequent segmental changes in the optic discs are compatible with focal infarcts in the optic nerves. In this case, the small vessels in the optic nerve heads looked surprisingly normal. There may have been involvement of more proximal vessels in the circle of Zinn, but these were not specifically studied.

Discussion

In their review, Francois and Neetens found 169 cases of the Foster Kennedy syndrome. Fifty-four of these cases, or 32%, were not due to tumors; 115, or 68%, were due to tumors of various locations (Table 1).

Ischemic optic neuritis is the disease most commonly confused with the Foster Kennedy syndrome in our experience. It has received almost no attention in the English literature. Francois, et al., have recognized this disorder as pseudopapillitis vasculaires.

Two such cases reported here were explored because of this difficulty in diagnosis. Ischemic optic neuritis is a disease of later life. Both eyes are usually involved but not simultaneously. The onset is abrupt, and pain is rare. Central vision is often spared, but altitudinal field loss is common. Pallor and choking of the nerve head is common. A history of vascular disease is almost always present.

In making the differential diagnosis between ischemic optic neuritis and tumors presenting the Foster Kennedy syndrome, the following criteria are useful (Table 2):

1. History. Sudden loss of vision with a delay in the involvement of the second eye is a common occurrence in inflammatory and vascular diseases of the optic nerves, while progressive decrease in acuity and field is seen with mass lesions.

2. Visual Fields. Vascular lesions of the optic nerve often produce altitudinal defects, nerve fiber bundle defects, and central sco-
Fig. 6. Case 6. Normal appearing retina with some serous detachment. H.&E., X100.

Fig. 7. Case 6. Left optic nerve head showing swelling and superficial hemorrhage. H.&E., X150.

Fig. 8. Case 6. Cross section of the right optic nerve with normal central artery and vein. H.&E., X60.
TABLE 1

Causes of the Foster Kennedy syndrome in 169 cases (from Francois and Neetens)

<table>
<thead>
<tr>
<th>Cause</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>NON-TUMOR (92%)</td>
<td>(54 total)</td>
</tr>
<tr>
<td>Arteriosclerosis and hypertensive cardiovascular disease</td>
<td>17</td>
</tr>
<tr>
<td>Optochiasmatic arachnoiditis</td>
<td>16</td>
</tr>
<tr>
<td>Internal carotid aneurysms</td>
<td>3</td>
</tr>
<tr>
<td>Pressure by internal carotid</td>
<td>3</td>
</tr>
<tr>
<td>Trauma with meningitis</td>
<td>3</td>
</tr>
<tr>
<td>Vascular, allergic &amp; endocrine</td>
<td>3</td>
</tr>
<tr>
<td>Syphilis</td>
<td>2</td>
</tr>
<tr>
<td>Internal hydrocephalus</td>
<td>2</td>
</tr>
<tr>
<td>Paget's disease</td>
<td>1</td>
</tr>
<tr>
<td>Undetermined</td>
<td>4</td>
</tr>
</tbody>
</table>

TUMOR & LOCATION (68%) (115 total)

<table>
<thead>
<tr>
<th>Location</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frontal</td>
<td>50</td>
</tr>
<tr>
<td>Olfactory groove</td>
<td>19</td>
</tr>
<tr>
<td>Sphenoid wing meningioma</td>
<td>17</td>
</tr>
<tr>
<td>Para or suprasellar</td>
<td>10</td>
</tr>
<tr>
<td>Optic nerve and orbit</td>
<td>5</td>
</tr>
<tr>
<td>Intracerebral</td>
<td>9</td>
</tr>
<tr>
<td>Cerebellar or intratumoral</td>
<td>4</td>
</tr>
<tr>
<td>Occipital</td>
<td>1</td>
</tr>
</tbody>
</table>

tomas. It is important to recall that optic neuritis can occur with normal acuity. There may be 20/20 vision with a swollen nerve head, as in papilledema, but instead of an enlarged blind spot, the field defects described above will be found.2

3. Ophthalmoscopy. Vascular disease often shows optic atrophy in a segmental area which correlates well with the visual field loss. The optic atrophy seen in tumors is often homogenous and more frequently involves the temporal area of the disc. The swollen disc in optic neuritis seldom reaches the same degree of swelling as that in papilledema. In ischemic optic neuritis, the disc is often pale and swollen with streak or linear hemorrhages and narrowed vessels, indicating the basic ischemic nature of the disease. Biomicroscopy may show cells in the vitreous. In tumors, a Marcus Gunn pupil will be present on the side of the optic atrophy. In ischemic neuritis, the Marcus Gunn pupil will be found on the side of the swollen nerve head.

4. Neurological Examination. This is often normal in ischemic neuritis, and the patient may give a history of previous stroke or transient attacks. In basal frontal tumors, the sense of smell may be lost on the side of the optic atrophy; contralateral hemiplegia may be present, as well as signs of frontal lobe damage.

Summary

The most common clinical condition to be confused with the Foster Kennedy syndrome is ischemic optic neuritis or vascular pseudopapillitis. We have discussed the clinical features and differential diagnosis of this syndrome and have reported cases of occult trauma, optic neuritis, and syphilis, each of which presented a similar ophthalmological picture. These cases emphasize the importance of recognizing that the Foster Kennedy syndrome is not always caused by a tumor.

References


TABLE 2

Differential diagnosis of ischemic neuritis versus basal frontal mass

<table>
<thead>
<tr>
<th>Clinical Data</th>
<th>Ischemic Neuritis</th>
<th>Basal Frontal Mass</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Sudden visual loss. Delayed visual loss in second eye.</td>
<td>Insidious loss of vision.</td>
</tr>
</tbody>
</table>


