CHIASMAL ARACHNOIDITIS AS A MANIFESTATION OF GENERALIZED ARACHNOIDITIS IN SYSTEMIC VASCULAR DISEASE*†

CLINICO-PATHOLOGICAL REPORT OF TWO CASES

BY

M. OLIVER, A. J. BELLER, AND A. BEHAR

From the Departments of Ophthalmology and Neurosurgery, the Rothschild-Hadassah University Hospital, and the Department of Pathology, Laboratory of Neuropathology, Hebrew University-Hadassah Medical School, Jerusalem, Israel

THE "Syndrome de Balado" (Balado and Satanowsky, 1929), later known as chiasmal arachnoiditis (Ch.A.) has been reported by many authors (Cushing, 1930a, b; Heuer and Vail, 1931; Craig and Lille, 1931; Vincent, Puech, and David, 1931; David, Hartmann, and Hebert, 1936; Bollack, David, and Puech, 1937a; Hausman, 1937; Vail, 1938). Its aetiology and pathogenesis have, however, remained controversial, and there is no agreement whether it is a specific entity or the manifestation of a more generalized disease of the meninges. One reason for this appears to be the lack of histopathological examinations, since the disease is not usually fatal. Only five cases have been reported in the literature with pathological studies (Davis and Haven, 1931; Bollack, David, and Puech, 1937b). Two additional cases are described in this report with clinico-pathological evidence and an attempt has been made to elucidate the pathogenesis of the syndrome.

Case Reports

Case 1, a woman aged 22 years, was admitted to the ophthalmological department of our hospital for the first time in June, 1950, with acute deterioration of the visual acuity of the left eye, ptosis of the left eyelid, and left hemifacial pain of 10 days' duration.

Physical Examination.—There was ptosis of the left upper eyelid and limited movements of the left eyeball due to paresis of all the extra-ocular muscles except the superior oblique and medial rectus. The left pupil was larger than the right with a sluggish reaction to direct light but a good consensual response. Hertel's exophthalmometry showed slight proptosis of the left eye (2 mm.). Visual acuity was 6/6 in the right eye, but was restricted to counting fingers at a distance of 1.5 m. in the left. The right visual field and fundus were normal. The left visual field showed a nasal and central defect (Fig. 1, overleaf) and there was papilloedema of 1 diopter with marked venous congestion. There was hyperalgesia in the distribution of all three divisions of the left trigeminal nerve. Blood pressure was 115/85 mm. Hg, and the general condition of the patient was good.

Laboratory Investigations.—Erythrocyte sedimentation rate 20 mm. in the 1st hr (Westergren), Hb 75 per cent., RBC 4,550,000/cmm., white cell count 8,300/cmm. with a normal differential count. Blood sugar 97 mg. per cent. Urea 32 mg. per cent. Wassermann reaction negative. The urine contained traces of albumin and occasional white blood cells.

X rays of the skull, orbits, optic foramina, sinuses, and chest were all normal.

Left common carotid arteriography was normal.

Progress.—After 3 weeks of treatment with penicillin, the pains in the distribution of the left trigeminal nerve, the ptosis, and the paresis of the extra-ocular muscles disappeared, but the visual acuity and field defect in the left eye remained unchanged. There was regression of the papilloedema, but evidence of optic atrophy was noted for the first time. Follow-up examinations over a period of 8 months revealed only increase of the optic atrophy.

* Received for publication January 26, 1967.

† Address for reprints: M. Oliver, Department of Ophthalmology, Rothschild-Hadassah University Hospital, Jerusalem, Israel.

FIG. 1.—Visual field of the left eye of Case 1. June, 1950.

Second Admission to Hospital.—In February, 1951, she was re-admitted because of several attacks of “total blindness” in the right eye, each of which lasted for several minutes. She also complained of pain in the right eye, especially on looking to the side.

Physical Examination.—As before her general condition was good and the blood pressure was normal. The left eye had not changed since the previous admission, and the visual acuity of the right eye remained 6/6, but the right visual field was now slightly constricted on the temporal side (Fig. 2), and fundus examination revealed venous congestion and a marked blurring of the disc.

FIG. 2.—Visual fields of Case 1. February, 1951.

Laboratory Investigations.—Blood, urine, and x ray examinations showed no change.

Lumbar puncture: pressure—140 mm. water; albumin—12 mg. per cent.; glucose—67 mg. per cent.; NaCl—710 mg. per cent. Pandy and Nonne tests negative. No cells found. Wassermann reaction and Kahn test negative. Goldsol 000000.

The electro encephalogram showed diffuse disturbance of cortical activity with a slight disturbance of the diencephalic centres.

Pneumo-encephalography showed a filling defect of the chiasmatic cistern, and the anterior horn of the right lateral ventricle appeared to be flattened.

Right common carotid arteriography was normal.

Operation.—In March, 1951, a left transfrontal exploratory craniotomy was performed to exclude the presence of a space-occupying lesion. The arachnoid membrane was thickened and adhesions were found in the region of the optic nerves and chiasma. No cyst formation was found. The chiasma, optic nerves, and left internal carotid artery were freed from adhesions. The left optic nerve was obviously atrophic. The post-operative recovery was uneventful, but there was no improvement in the left eye. No further attacks of sudden blindness were experienced.

Follow-up.—The patient emigrated to Canada and was admitted to hospital on several occasions during the next 5 years. Between October, 1951, and August, 1953, she developed major convulsive seizures on at least three occasions. In November, 1953, she showed signs of systemic disease with malaise, loss of weight, joint pains, and hypertension (210/110 mm. Hg). In January, 1954, she was admitted for investigation of "hypertensive nephropathy" accompanied by a constant low-grade fever, and polyarteritis nodosa was suspected. One month later she experienced an attack of vertigo, ataxia, nausea, and vomiting accompanied by numbness of the right side of the face and left side of the body. The diagnosis was thrombosis of the right posterior inferior cerebellar artery, probably due to polyarteritis nodosa.

Termination.—In December, 1956, she had a sudden onset of nausea, vertigo, and vomiting followed by coma and she died the following day.

Diagnosis.—Thrombosis of the basilar artery or intrapontine haemorrhage due to polyarteritis nodosa.

Necropsy Findings.—There was marked softening of the midbrain and pons and a large recent haemorrhage was found in the rostral portion of the right pons. Adhesions were found in the middle cranial fossa particularly around the optic nerves, internal carotid arteries, and pituitary stalk. The left optic nerve was atrophic and the left internal carotid artery was drawn upwards and medially by adhesions binding it to the optic nerve and other structures.

Microscopical Examination.—The right basal ganglia showed marked thickening and hyalinization of capillaries and arterioles. The latter showed perivascular infiltration and signs of fibrinoid permeation with aneurysmal dilatation of walls (Fig. 3). In the brain there were multiple milium scars containing scattered lymphocytes, macrophages laden with haemosiderin, reactive microglia and astrocytes (Fig. 4).

FIG. 3.—Cerebral polyarteritis nodosa. Aneurysmal dilatation, fibrinoid permeation, and leucocytic infiltration of the wall of an artery. Haematoxylin and eosin. $\times 110$.

FIG. 4.—Healed milium infarction in cerebrum. Microglial and astroglial reaction replacing necrotic focus. Haematoxylin and eosin. $\times 110$.

The left optic nerve showed total demyelination, marked fibrosis, and much glial tissue (Fig. 5, overleaf). There was also some demyelination involving the upper fibres of the optic chiasma (Fig. 5). The right optic nerve was normal.

The meninges in the chiasmal region were markedly thickened and collagenized, and contained numerous lymphocytes, red blood cells, and haemosiderin-laden macrophages. Medium-sized arteries in the meninges

FIG. 5.—Transverse section through optic nerves at the chiasma, showing atrophy, pallor, and total absence of nerve fibre orientation in left nerve. Haematoxylin and eosin. $\times 8$.

showed marked intimal fibrosis, organized thrombi, and polymorphonuclear and round cell infiltration of the walls, and adjacent leptomeninges. The latter showed diffuse fibrous thickening (Fig. 6).

FIG. 6.—Fibrous adhesive arachnoiditis of cerebrum around leptomeningeal vessels, showing healing and healed polyarteritis nodosa. Haematoxylin and eosin. $\times 44$.

Case 2, a man aged 62 years, was admitted to the medical ward in March, 1965, because of bronchopneumonia. He had been treated for syphilis with salvarsan at the age of 18.

Physical Examination.—Body temperature 39°C . Cyanosis and distention of the neck veins. The patient was markedly dyspnoeic and moist crepitations were present over the right lung. Blood pressure 160/80 mm. Hg.

Neurological Examination.—The knee and ankle reflexes could not be elicited, and speech was dysarthric. There was anisocoria ($R > L$) and Argyll Robertson pupils. Since the patient's behaviour was peculiar, psychological tests were performed and the findings were compatible with general paralysis of the insane.

Ophthalmological Examination.—The visual acuity of the right eye was 6/15 with -0.75 D sph., and that of the left eye 6/9 with -1 D sph. The right visual field showed an altitudinal and upper nasal quadrant defect, and some concentric constriction was found in the left eye (Fig. 7). Both fundi showed pallor of the optic discs.

FIG. 7.—Visual fields of Case 2. April, 1965.

Laboratory Investigations.—Erythrocyte sedimentation rate 76 mm. in the 1st hr (Westergren); Hb 10.8 mg. per cent.; haematocrit 31 per cent.; white blood cell count 12,000/cmm.; blood urea 45 mg. per cent.; blood glucose 85 mg. per cent. Cryoglobulins and cold agglutinins were not found. The Wassermann reaction was positive at a titre of 0.2 units. The urine contained traces of albumin.

Chest x ray showed moderate enlargement of the heart shadow and aorta, and a massive infiltrate at the apex of the right upper lobe.

The cerebrospinal fluid showed normal pressure, glucose 76 mg. per cent., protein 63 mg. per cent., goldsol—negative.

Diagnosis.—Bronchopneumonia, tertiary syphilis (tabes dorsalis, general paralysis of the insane), and primary optic atrophy due to syphilis.

Treatment.—The patient was treated with penicillin and discharged from hospital after 10 days.

Second Admission to Hospital.—10 months later he was re-admitted with signs of severe congestive heart failure. The ophthalmological and neurological findings were the same as before. The chest x ray showed a marked enlargement of the heart and an aneurysm of the upper thoracic aorta was diagnosed.

Termination.—The patient appeared to improve after treatment with digoxin, diuretics, and antibiotics, but on the fourth day after admission he suddenly lost consciousness and died within a few minutes. An electrocardiogram immediately before death showed ventricular fibrillation.

Necropsy Findings.—There was severe generalized arteriosclerosis involving the heart, aorta, and kidneys. Tertiary syphilis was confirmed by the presence of an aneurysm in the upper thoracic part of the aorta with histological evidence of a syphilitic mesaortitis. A generalized patchy arachnoiditis was seen over the convex surface and base of the brain encroaching on the optic chiasma and optic nerves (Fig. 8, overleaf).

Microscopical Examination.—The meninges overlying the optic chiasma and optic nerves showed a chronic non-specific inflammatory reaction with proliferation of connective tissue (Fig. 9, overleaf). Both optic nerves showed a marked thickening of the intraneural fibrous septi. Of special interest was a sectoral atrophy with complete demyelination of the right optic nerve (Fig. 10, overleaf). The arachnoid overlying this sector was not thicker than that overlying the rest of the nerve. Transverse sections at various levels of the spinal cord showed selective demyelination of the posterior columns (Fig. 11, overleaf).

FIG. 8.—Fibrous adhesive optic arachnoiditis contiguous with arachnoiditis on basal surface of pons and interpuncular fossa.

FIG. 9.—Fibrous adhesive arachnoiditis of extracranial portion of left optic nerve. Petersen's stain. $\times 52$.

Discussion

In the first case reported above, generalized vascular disease involving the brain and leptomeninges had caused a fibrous thickening of the latter, especially in the chiasmal region. In the second case the changes in the chiasmal region were also part of a generalized patchy arachnoiditis. Bollack and others (1937) considered Ch.A. to be a localized form of arachnoiditis similar to that found in the posterior fossa and in the spine and not necessarily preceded by a generalized meningeal affection (Heuer and Vail, 1931). The autopsy findings,

FIG. 10.—Transverse section of right optic nerve, showing complete demyelination and sectoral atrophy. The latter is surrounded by nerve bundles showing myelin pallor. No arachnoid thickening is present over this particular area of the nerve. Woelcke's stain for myelin sheaths. $\times 44$.

FIG. 11.—Transverse section of spinal cord, showing selective demyelination of tracts of Goll and Burdach. Woelcke's stain for myelin sheaths. $\times 10$.

however, tend to support the assumption that the syndrome is an expression of a generalized disease of the arachnoid, with clinical symptoms and signs referable to the opto-chiasmal region as one of its manifestations.

The clinical and pathological evidence favours a vascular factor in the pathogenesis. In Case 1 Ch.A. was the presenting symptom of polyarteritis nodosa which was diagnosed 3 years later. There was a close time relationship between those neurological signs obviously based on vascular events and those due to Ch.A. Many cases of collagen diseases are known

to present neurological signs years before the clinical picture is clear and the diagnosis established. On the other hand we are unaware of any reported cases in which the presenting symptom of polyarteritis nodosa was Ch.A. The affection of the optic pathways might have resulted from a vascular obliterative process due to polyarteritis nodosa or to strangulation of blood vessels traversing the arachnoid, thickened by the periarterial inflammation. This in itself may have been caused by haemorrhage within the arachnoid as a result of polyarteritis nodosa (Fig. 6). In Case 2 there was no gummatous infiltration of the optic nerves but there was some evidence that the pathogenesis of the Ch.A. was related to endarteritis. The sectoral demyelination of the optic nerve (Fig. 10) was more likely to be due to an obliterative endarteritis than to a strangulating effect of the thickened arachnoid, since the arachnoid overlying this sector was not thicker than that over the rest of the nerve. This assumption is supported by the fact that the small vessels of the optic nerve originate partly in the meninges and enter the nerve in a radial fashion (Hayreh, 1963).

These two cases favour the theory that vascular factors contribute to the pathogenesis of Ch.A. (Taptas and Dimopoulos, 1949; Dickmann, Cramer, and Kaplan, 1951).

The value of surgical treatment in suspected cases is controversial. Improvement in vision ranges from 28 per cent. (Bollack and others, 1937c) to 47 per cent. (Dickmann and others, 1951). However, despite the best surgical techniques, many patients still deteriorate. There are no clear criteria concerning indications for surgery.

It seems that many factors influence the outcome of surgical treatment. These two cases illustrate both the effects of strangulation of the optic nerve and of parenchymatous damage. The first patient was operated upon 8 months after signs of the disease first appeared and at that time the left optic nerve was already severely damaged, whereas the right fundus showed only incipient papilloedema and venous congestion. She had at that stage several paroxysms of complete blindness in the right eye; since operation relieved her complaints and there was an objective improvement in the right fundus, it is likely that the right optic nerve had been strangulated by the fibrous adhesions which were found at the operation. It is clear that parenchymatous damage, as found in the left optic nerve of the first patient, or as manifested by the sectoral defect in the second, is irreversible and that surgery is of no benefit.

The difficulty of predicting the prognosis after operation is related to the fact that there is no definite method of determining clinically whether the field loss, papilloedema, or optic atrophy are due to fibrous strangulation or direct parenchymatous damage.

Summary

A clinico-pathological report of two cases of chiasmal arachnoiditis is presented; in the first the condition was a manifestation of polyarteritis nodosa and in the second it was a part of meningovascular syphilis.

The histopathological findings emphasize the importance of vascular factors in the pathogenesis of this syndrome, and explain the unpredictable result of surgical intervention.

We are grateful to Dr. G. Mathieson and the Montreal Neurological Institute for enabling us to study the histopathological preparations of Case 1.

REFERENCES

- BALADO, M., and SATANOWSKY, P. (1929). *Arch. argent. Neurol.*, **4**, 71.
- BOLLACK, J., DAVID, M., and PUECH, P. (1937a). "Les arachnoïdites optochiasmatiques". Masson, Paris.
- (1937b). *Ibid.*, p. 59.
- (1937c). *Ibid.*, p. 248.
- CRAIG, W. M., and LILLE, W. I. (1931). *Arch. Ophthal. (Chicago)*, **5**, 558.
- CUSHING, H. (1930a). In "XIII Concilium Ophthalmologicum, Amsterdam—den Haag, 1929", vol. 3, p. 97.
- (1930b). *Arch. Ophthal. (Chicago)*, **3**, 505, 704.
- DAVID, M., HARTMANN, E., and HÉBERT, E. (1936). *Bull. Soc. ophtal. Paris*, p. 789.
- DAVIS, L., and HAVEN, H. A. (1931). *J. nerv. ment. Dis.*, **73**, 129, 286.
- DICKMANN, G. H., CRAMER, F. K., and KAPLAN, A. D. (1951). *J. Neurosurg.*, **8**, 355.
- HAUSMAN, L. (1937). *Arch. Neurol. Psychiat. (Chicago)*, **37**, 929.
- HAYREH, S. S. (1963). *Brit. J. Ophthal.*, **47**, 651.
- HEUER, G. J., and VAIL, D. T. (1931). *Arch. Ophthal. (Chicago)*, **5**, 334.
- TAPTAS, J. M., and DIMOPOULOS, T. (1949). "Arachnoïdites optochiasmatiques et maladie neurovasculaire". Masson, Paris.
- VAIL, D. (1938). *Arch. Ophthal. (Chicago)*, **20**, 384.
- VINCENT, C., PUECH, P., and DAVID, M. (1931). *Rev. neurol.*, **1**, 760.