Case Reports

Hodgkin's Disease Involving Optic Nerve and Brain

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The occurrence of Hodgkin's disease with involvement of optic nerves and chiasm has not been reported in the literature reviewed by the authors. Presence of Hodgkin's disease in the brain and in association with so-called multifocal leukoencephalopathy is well documented.

The following case of Hodgkin's disease was diagnosed at autopsy after clinical and operative findings revealed evidence of a mass intrinsically involving the optic nerves and chiasm. This case was followed from the time of apparent onset of symptomatology to its termination by the same group of physicians.

Case Report

A 56-year-old white male was admitted to General Rose Memorial Hospital on Aug. 27, 1960 for investigation of decreased visual acuity, low-grade fever and loss of weight commencing July 15, 1960 with an upper respiratory infection. Pharyngitis and elevation of temperature to 103°F. subsided in 2 to 3 days, although malaise and asthenia persisted. Decreased visual acuity in the right eye appeared 10 days after onset of the illness. An ophthalmologist reported slight edema of the right optic disc with "some vascular and inflammatory changes of the right retina." Treatment with prednisolone (Prednison) resulted in no improvement.

The remainder of the history was not remarkable except that his wife was somewhat concerned about his slightly inappropriate behavior of about 6 months' duration.

Examination. He was well developed, well nourished and in no acute distress. Temperature was 99.2°F., respiratory rate 22, and pulse rate 80 per min. Blood pressure was 112/67. Skin was normal in appearance and there was no adenopathy. Thorax, lungs, heart, abdomen and rectum showed no abnormalities.

There was slight inappropriateness of behavior. There was no aphasia, he was well oriented and presented no defect in memory. Positive neurological abnormalities were confined to the eyes. The right optic disc was edematous and hemorrhages were noted in contiguity with the vessels. He was unable to detect light or movement of hands with this eye. The right pupil was dilated and nonreactive directly or consensually. The optic fundus on the left was not remarkable, but there was blindness of the entire temporal visual field to confrontation. He could count fingers at a distance of 2 feet with this eye. The left pupil was large and reacted sluggishly to light, and extraocular movements were full with no nystagmus.

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Laboratory studies demonstrated a polymorphonuclear leukocytosis with reticulocytosis, low count of platelets and normal hemoglobin, hematocrit and erythrocyte sedimentation rate. V.D.R.L., L.E. preparations and febrile and heterophile agglutinations were normal. Urinalysis and liver-function studies gave normal results. The spinal-fluid manometric pressures were normal and there was no pleocytosis, but the fluid contained 170 mg. per cent of protein.

Roentgen-ray studies of skull, chest and gastrointestinal tract were not remarkable, and bilateral carotid arteriography revealed minimal elevation of the proximal anterior cerebral arteries.

Operation. Bifrontal craniotomy was done on Sept. 3, 1960. Examination of the brain, meninges and blood vessels by the neurosurgeon and pathologist revealed no abnormalities. The right optic nerve was about twice normal in size with even greater enlargement appearing in the chiasm. There was some yellow discoloration of these structures. Biopsy was not done at the time of operation in hopes of preserving remaining vision by appropriate treatment of "edema or tumor" following operation.

FIG. 1. Photograph of brain. Note enlargement of right optic nerve and chiasm.
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Postoperative Course. Treatment included prednisolone and tetracycline. He was afebrile for 1 week. Spikes in temperature and dementia then appeared, and the medications were discontinued. He became more disoriented and demented with cachexia, and nuchal rigidity developed on Oct. 1, 1960. On lumbar puncture the cerebrospinal fluid contained one polymorphonuclear cell, sugar 19 mg. per cent, and protein 85 mg. per cent. Spinal-fluid electrophoresis was within normal limits.

Cultures of blood and bone marrow were negative and examination of the bone marrow revealed decreased thrombocytic formation.

He expired on the 43rd hospital day.

Postmortem Examination. The brain revealed slightly yellow discoloration beneath the frontal lobes with normal-appearing convolutions and sulci. The left leptomeninges were slightly thickened. The optic nerves were enlarged, particularly on the right, with diffuse swelling extending into the chiasm, optic tracts and hypothalamus (Fig. 1).

Gross sectioning revealed hemorrhagic cortical patches, 3–5 mm. in diameter, in the left frontal, occipital and right temporal lobes with adjoining gray-red softening of underlying white matter. There was a left parietal area of corticomedullary softening about 1.0 cm. in diameter. The white matter of both parietal lobes showed barely visible gray streaks. The thalami, basal ganglia, peduncles, brain stem and cerebellum were unremarkable grossly.

Incidental findings at autopsy included coronary arteriosclerosis with recent intramural hemorrhage of the descending left coronary branch. There was a large thrombosed aneurysm of the abdominal aorta.

The spleen weighed 290 gm. and showed numerous prominent malpighian corpuscles. Peripancreatic lymph nodes were enlarged to 0.5 cm. in diameter and were white to pearly gray on cut section.

Microscopically, typical findings of Hodgkin’s disease were observed in peripancreatic lymph nodes, spleen, brain, and leptomeninges as well as in optic nerves, chiasm (Figs. 2 and 3), tracts and hypothalamus.

The microscopic finding of severe granulomatous meningoencephalitis was in marked contrast to the apparently normal gross appearance of the brain at operation and autopsy.

White matter underlying the most involved sites of cortical involvement revealed barely visible recent demyelination.

Discussion

The occurrence of visual disturbances in previously reported cases of Hodgkin’s disease has been attributed to involvement of optic cortex. Involvement of the optic nerves, chiasm and tracts in this case is unique. The clinical finding of absent vision on the right with a left temporal hemianopia led to the assumption that the right optic nerve and chiasm were involved. The elevated spinal-fluid protein and minimally elevated

Fig. 2. Photomicrograph (low power) of optic chiasm showing replacement of normal neuronal fibers by neoplastic cells.