Histiocytosis X in the Optic Chiasm of an Adult with Hypopituitarism*

Case Report

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HISTIOCYTOSIS X (nonlipid eosinophilic granuloma, lipoid granulomatosis, xanthomatosis, reticuloendotheliosis, reticulosis, Hand-Schüller-Christian disease) is a well-known cause of cranial lesions, exophthalmos, visual loss, diabetes insipidus, and other hypothalamic-pituitary disturbances in children. Visceral and osseous involvement has been reported occasionally in adults with a seeming propensity for the pituitary-hypothalamic region. But an apparently isolated histiocytic granuloma mimicking a suprasellar tumor in an adult is extremely rare. We wish to report such an occurrence and to demonstrate the ultrastructure of the lesion which originated in the optic chiasm.

Case Report

The patient, a 34-year-old man, was admitted to St. Louis University Hospital on February 6, 1967, with a complaint of pain and dim vision in the left eye for 4 weeks. He had been struck over his left eye in an auto accident 1½ years earlier.

He had served in the army from age 19 to 21 and noted nothing abnormal until the age of 25 when he began to gain weight and lose libido. In the next 9 years, he went from 200 to 340 pounds. He shaved less frequently, and began to drink and urinate much more. He continued work as a metal inspector until his admission.

Examination. Vital signs were normal, height 73 inches, weight 335 lbs. The patient had a moon face, dry cracking skin, scanty body hair, female escutcheon, small soft testes, and extreme obesity of the trunk and extremities with no cutaneous striae. There was also bitemporal hemianopsia, extremely poor visual acuity on the left, and marked bilateral optic atrophy (Fig. 1). The patient's reflexes were not characteristic of hypothyroidism. The left eye was tender but not exophthalmic or inflamed.

X-rays of the skull, chest, and optic foramina were normal, with no areas of rarefaction of bone or changes in the sella turcica.

Laboratory examination showed a hematoctrit of 37%, a normal white blood cell count, and nonreactive tests for syphilis; the urinalysis was normal.

The generalized obesity was interpreted as due to an increased appetite and large consumption of food over many years. No signs of diabetes mellitus were present. An oral glucose tolerance test performed with 100 gm of glucose revealed a minimal elevation in blood sugar. During his hospital course the average daily output of urine was between 6 and 10 liters. The maximal specific gravity observed after 12 hours of water deprivation was 1.005. The intramuscular administration of 1.25 units of Pitressin tannate decreased urine output to 3.6 liters per day, and the specific gravity rose to 1.015. This confirmed the presence of diabetes insipidus. A fasting plasma growth hormone

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was 10.5 m\(\mu\)g/ml. Hypoglycemia induced by intravenous administration of 8 units of insulin produced no elevation in growth hormone levels. Serum calcium was 4.6 mEq/liter, phosphorus 2.7 mEq/liter, alkaline phosphatase 1.6 S.U., and urinary hydroxyproline 38 mg/24 hrs. These data indicate an impairment of growth hormone secretion.

Clinical and laboratory evaluation of thyroid function revealed a mild secondary hypothyroidism. The protein-bound iodine was 3.3 \(\mu\)g%; the TBI 0.74, the radioactive iodine uptake at 6 hrs, 5%; at 24 hrs, 15%; and cholesterol 233 mg\%. There was a 25% response of radioactive iodine uptake following administration of 10 units of thyroid-stimulating hormone per day, for 4 days. Adrenal function studies revealed 17 ketosteroids 10.5 mg/24 hrs, 17-OHcorticoids 4.2 mg/24 hrs, normal serum electrolytes: sodium 150 mEq/liter, potassium 4.1 mEq/Cl\(_2\) 110 mEq/liter. Adrenal stimulation with 80 units of ACTHAR-Gel per day for 3 days increased the urinary output of 17-OHcorticoids fourfold. Oral administration of Metoprine (750 mg per 6 hrs) was not followed by an elevation of 17-OHcorticoids. These studies indicate a mild adrenal insufficiency due to impaired adrenocorticotrophin secretion. Hypogonadism was manifested by definitive changes in secondary sexual characteristics as well as functional impairment. Low urinary follicle-stimulating hormone (below 6 units) suggests that this hypogonadism was of secondary origin. Absence of gynecomastia or galactorrhea pointed out no impairment of prolactin secretion.

The endocrine-metabolic picture indicated incomplete panhypopituitarism and diabetes insipidus.

Operation. On February 24, a right frontal craniotomy was done for exploration of the chiasm. The right optic nerve had a brownish hue and was followed to its commissure. The left optic nerve was replaced by a tuberous, shaggy, brown mass which was incised, and biopsied.

The visible tumor appeared to be confined to the left optic nerve and chiasm; the right optic nerve appeared normal. The most likely gross diagnosis was optic glioma.

Postoperative Course. The patient had no light perception in the left eye. Vision in the right eye was unchanged. Pitressin was required for 2 weeks. He was maintained on cortisone acetate 25 mg per day, sodium levothyroxine 0.3 mg per day, and fluoxymesterone 10 mg per day. He received a 2700 r midline dose of radiation therapy over a 3-week period and is presently at work.

Histological Examination. There were numerous granulomas scattered in a matrix composed of many fine astrocytic fibers, a few large-bodied astrocytes, and moderate numbers of myelinated axons (in paraffin sections of the surgical specimen stained with hematoxylin and eosin). The cells within the granulomas were for the most part loosely arranged, small to medium in size, and polygonal or stellate in outline. Cytoplasmic vacuoles were absent. In the peripheral and central portions of several of the granulomas, large numbers of eosinophils were present. Nearby blood vessels were cuffed by mononuclear cells, mainly lymphocytes (Fig. 2).

Electronmicroscopic Examination. These elements were shown by electronmicroscopy in greater detail. After careful searching, a few histiocytes were found with characteris-