Neoplastiform Xanthomatous Granulomas of Choroid Plexus in a Child Affected by Hand-Schuller-Christian Disease

Case Report

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Diabetes insipidus, exophthalmos, and osteolytic lesions are the most common clinical findings in Hand-Schuller-Christian (HSC) disease. Involvement of the central nervous system has occasionally been found and is usually a late occurrence in the course of the disease.3,5,6,9–11,13,15,16,18–21

We are reporting the surgical removal of huge xanthomatous granulomas involving the choroid plexus of both lateral ventricles in a child with apparent Hand-Schuller-Christian disease.

Case Report

G.P., an 8-year-old boy, was admitted to the neurosurgical service of the Ospedale Civico in Palermo on March 10, 1965, because of headache and vomiting. Since the age of 4 years he had been under intermittent pituitary treatment for polydypsia and polyuria. When 6 years old, he developed right-sided focal seizures.

Examination. The patient was alert and cooperative and appeared in good general condition. Neurological findings included paleness of the optic discs with slight edema of the edges, and right-sided hemiparesis with positive pyramidal signs. Under Pituitrin treatment, the urinary output was 3 to 3½ liters in 24 hours.

Laboratory Examination. Urinalysis was normal except for a specific gravity of 1004. Fasting blood sugar was 74 mg% and the blood urea nitrogen 44 mg%. X-rays of the skull revealed separation of the cranial sutures and an increase in the convolutional markings. An electroencephalogram indicated clear-cut changes over the left temporoparietal area, suggesting a space-occupying lesion.

On March 19, frequent epileptic seizures occurred, characterized by conjugate deviation of the head and the eyes to the left and tonic-clonic movements of the left side of the face, spreading to the left side of the body. An accentuation of the right-sided motor deficit was observed. The fits could not be controlled by administration of barbiturates and osmotic solutions (20% Mannitol). The right lateral ventricle was tapped. Cerebrospinal fluid was drained under an increased pressure; following this procedure, the seizures ceased. A ventriculogram (20 cc air) on the same day showed a marked shift of the ventricular system to the right and an elevation of the left temporal horn. A left carotid angiogram showed slight deviation of the midline vessels to the right and upward and medial displacement of the middle cerebral artery. The anterior choroidal artery appeared slightly larger than normal. An avascular, possibly cystic, tumor in the temporal area was diagnosed.

First Operation. On the same day, a large left temporal craniotomy was performed under general anesthesia. The temporal lobe seemed increased in size, but on gentle palpation it felt soft. Through a vertical incision 5 cm from the tip of the temporal lobe a huge cyst was encountered at a depth of 6 mm. It contained yellow fluid and was oval and measured about 10 ¥ 7 ¥ 4 cm in its anteroposterior, vertical, and horizontal diameters. The wall of the cyst had a smooth, white to grayish lining. Over the medial aspect underlying the wall of the cyst, a yellow-colored tissue was noticed. The cyst lining was removed by suction, revealing the outer surface of a large hard mass located in the cavity of the temporal horn and extending to the ventricular trigone (Fig. 1). The posterior pole of the tumor was attached to the choroid plexus, which had to be ligated and divided in order to remove the mass.

Postoperative Course. In a few days the

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boy regained full motor power in his limbs, and his postoperative course was uneventful. As soon as the histological findings became known, the following laboratory determinations were made: blood cholesterol, total 112 mg%; cholesterol esters, 54 mg%; ketosteroids, 0.118 mg% (3.54 mg in 24 hours); blood electrolytes, Na 137 mEq, K 4.51 mEq, Ca 5.03 mEq per liter. X-rays of the chest and of the whole skeleton did not reveal any osteolytic lesions. On April 15, 1965, the boy went home. A right facial weakness was his only neurological deficit. Under Pituitrin treatment he eliminated 2500 cc of urine in 24 hours. Concentration remained quite low (S.G. 1005).

On July 22, 1965, after a volley of epileptic seizures involving the left side of the body, the boy had to be hospitalized again. An EEG indicated epileptogenic activity in the left temporoparietal area. Pneumencephalography showed a filling defect due to a mass situated in the trigone and extending to the body and to the temporal horn of the right lateral ventricle. Right carotid angiography indicated an elevation of the middle cerebral artery; the midline vessels were in normal position. Surgical intervention was refused.

On September 13, 1965, the patient was hospitalized for the third time. No significant neurological changes were recorded except for increased paleness of the optic discs and spontaneous vertical nystagmus. Visual acuity remained normal. An EEG control test failed to reveal the presence of the mass in the right lateral ventricle.

Second Operation. On September 13, 1965, a right temporoparietal craniotomy was performed under general anesthesia. The brain was not bulging but felt rather hard. A 4 cm long incision was made in the posterior parietal area, and at a depth of about 4 cm the lateral ventricle was entered. It appeared occupied by a sulphur-yellow, hard, well-encapsulated mass, not adherent to but touching the ependymal wall. The tumor was removed en bloc, after dividing the choroid plexus to which it was intimately connected. Separation from the ventricular wall could be obtained by gentle retraction and by suction of the ependyma as necessary. Since the mass spread well into the temporal horn and into the body of the ventricle, it had the shape of a horseshoe.

Postoperative Course. Recovery was uneventful until the third day when the boy suddenly became blind. The pupils were midratic and areflexic, whereas the fundi remained unchanged. Angiography ruled out surgical complications, such as intracerebral or subdural bleeding. With the blindness, the urinary output increased to 5 liters despite adequate hormone treatment. Administration of large quantities of cortisone and ACTH failed to improve the patient’s disability. On October 30, 1965, the patient, still blind, was discharged from the hospital.

He was seen again 5 months later; his general condition appeared poor, with loss of weight and marked paleness. Although treatment with Pituitrin had been discontinued, the daily urinary output was now 2 liters. Blood cholesterol and 17-ketosteroids were still normal. Loss of sight remained complete and there was now clear-cut optic atrophy. Mental confusion, amnesia, and nervous tantrums were reported.

Gross Pathological Study. The endoventricular masses were comparable in shape, size, and color (Fig. 2). Each resembled a sausage, 9 cm long and 3 to 4 cm in diameter. The surface was irregular with several bright yellow humps. When sectioned longitudinally, the mass felt hard and firm (Fig. 3). The internal surface was uneven due to numerous sulphur-yellow nodules 4 to 7 mm in diameter protruding from the yellow-grey tissue.

Microscopic Study. The histological findings in the two tumors were identical. There were marked dissimilarities between the sulphur-yellow nodules and the surrounding grey tissue. The grey tissue contained nests of eosinophiles, lymphocytes, plasma cells, histiocytes, and fibroblasts, surrounded by bands of connective tissue. Some areas were sclerotic and poor in cells; others were rich with fibroblasts (Fig. 4).

The sulphur-yellow nodules contained areas in which there were only foam cells; in other areas the foam cells were mingled with eosinophiles, lymphocytes, plasma cells, and neutrophiles (Fig. 5). The different areas were separated by hyaline collagen fibers. The foam cells had a central or paracentral nucleus that was small and hyperchromatic (Fig. 6): the large cytoplasmic space contained many vacuoles and fine granules.