Surgical Treatment of Sturge-Weber Syndrome

—Case Report—

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Abstract

It is well known that when Sturge-Weber syndrome manifests with seizures in early infancy, hemiparesis develops early, seizures become intractable, and motor weakness and mental retardation are progressive. In North America and Europe, early surgical intervention is recommended in such cases. However, neurosurgical management of Sturge-Weber syndrome has not been reported in Japan. The authors describe a 4-month-old boy with Sturge-Weber syndrome accompanied by intractable seizures who was successfully treated by a two-stage hemispherectomy. Two years postoperatively he remains free of seizures and is active, although his psychomotor development is moderately retarded. Surgical treatment of Sturge-Weber syndrome, including long-term results, is discussed in detail.

Key words: Sturge-Weber syndrome, phacomatosis, hemispherectomy, infantile hemiplegia

Introduction

Sturge-Weber syndrome exhibits a broad spectrum of clinical and neurological manifestations from its onset.^{5,10,11,18,21)} Studies of its natural history have revealed that one third²¹⁾ to one half¹⁸⁾ of patients with this syndrome are severely disabled. The problems these patients encounter are generally attributed to chronic seizures and cerebral ischemia followed by cerebral atrophy due to extensive cerebral angiomas.^{1,10,18)} A number of authors^{1,4,10,18)} have noted that patients with Sturge-Weber syndrome who exhibit convulsions in early infancy develop hemiparesis relatively early, suffer intractable seizures, and display progressive motor weakness and mental retardation.

Surgical treatment of Sturge-Weber syndrome was first performed in 1933. The procedure consisted of electrocoagulation of a cerebral venous angioma following craniotomy under local anesthesia.⁴⁾ For control of seizures and prevention of the later development of progressive neurological deficiency, surgical intervention within 3 months¹⁾ to 1 year¹⁰⁾ of age is now recommended in North America and Europe. However, neurosurgery for this syndrome

has not been addressed in the Japanese medical literature. We treated a young patient with Sturge-Weber syndrome by hemispherectomy, and we discuss our results and those of others.

Case Report

A 4-month-old boy was noticed, at birth, to have a port-wine nevus on the left forehead (Fig. 1). At 3 months of age he began to have seizures, with twitching of the right side of the face progressing to the right arm and leg. He continued to have occasional seizures after the initiation of anticonvulsant therapy, and also developed slight right hemiparesis. Neither buphthalmos nor glaucoma were noted on ophthalmological examination. Neurological examination revealed mild motor dysfunction of the right extremities.

Electroencephalography revealed asymmetry with a smaller number of spindles and lower amplitude in the left hemisphere. There were also paroxysmal sharp waves in the left middle temporal and occipital areas. Sensory and visual evoked potentials were diminished on the left side. In the left brainstem, the waves I-V of auditory evoked brainstem potentials were attenuated. A plain x-ray disclosed no calcification. A left carotid angiogram showed slight

Received July 20, 1987; Accepted July 22, 1988

Fig. 1 The patient at 4 months of age. Note the port-wine nevus on the left forehead.

Fig. 3 Intraoperative photograph showing extensive leptomeningeal angiomas in the left frontotemporal cortex and dilatation of the subarachnoid space in the temporal region.

Fig. 2 CT scan demonstrating a calcified lesion in the left occipitoparietal cortex which was not well enhanced by the contrast medium. Dilatation of the sulcal impressions in bilateral frontal lobes is also evident.

opacification of the superficial Sylvian veins but no abnormal vasculature. Computed tomography (CT) demonstrated a calcified lesion in the left occipitoparietal cortex, which was only minimally enhanced by contrast medium, and dilatation of the sulcal impressions in the bilateral frontal lobes (Fig. 2).

A two-stage left hemispherectomy was undertaken. First, a left occipitoparietal craniotomy was performed when the patient was 6 months old, at which time we were unwilling to remove the motor

cortex. The calcified vascular lesion in the left occipitoparietal region was excised with a CO₂ laser. Leptomeningeal angiomas and dilatation of the subarachnoid space due to cerebral atrophy were observed in the left frontotemporal region (Fig. 3), although they had not been visualized on the pre-operative CT scan.

After the first operation the seizures continued. A CT scan obtained when the patient was 11 months of age disclosed a contrast-enhanced lesion in the left frontotemporal region in which the angiomas had been found during surgery (Fig. 4).

When the child was 12 months old the second operation was performed. Through a left frontotemporal craniotomy the left frontal and temporal cortices containing the angioma were removed. Figure 5 is the postoperative CT scan, showing the completed left hemispherectomy.

Postoperatively the hemiparesis temporarily worsened, but then motor function in the right extremities recovered. At 3 years of age, his psychomotor development was retarded (motor function, 1.3 year; mental function, 1 year) and right finger dexterity was very poor. However, he can walk and run, and is able to speak a few words. His seizures disappeared after the second operation.

Pathological examination showed extensive leptomeningeal angiomas (Fig. 6). Calcifications were noted in the subendothelial space of the angioma and in the perivascular and intraparenchymal spaces of the gray and white matter. Also observed were degeneration and loss of neurons and reactive gliosis. He is being followed.

Fig. 4 CT scan obtained after the first operation (age, 11 months). Note the absent left occipitoparietal cortex and the contrast-enhanced lesion in the frontotemporal region.

Fig. 5 CT scan taken after the second operation, when upon completion of the left hemispherectomy.

Discussion

I. Natural history of Sturge-Weber syndrome

The presence of a facial port-wine nevus at birth is a hallmark of Sturge-Weber syndrome.^{1,5,10} However,

Fig. 6 Photomicrographs of frontal cortical tissue. There is extensive leptomenigeal angiomatosis, with calcifications within the angioma and brain parenchyma. Degeneration of neurons and gliosis are also seen. HE stain, $\times 50$ (upper) and $\times 100$ (lower).

the onset, severity, time course, and outcome of the neurological manifestations, which include seizure, hemiparesis, and mental retardation, are quite varied.^{1,5,6,10,11,16,18,21} Little is known of the natural history of the disease. Two long-term follow-up studies^{18,21} disclosed that one third to one half of the patients had done poorly or died. In a Mayo Clinic series comprising 31 cases reported by Peterman *et al.*,¹⁸ 17 patients (55%) were considered well. Of the 10 (32%) who did poorly, five were confined to institutions and five lived at home but were in very poor health as a result of uncontrolled seizures, severe mental retardation, progressive motor impairment, or blindness. Four of the patients (13%) died.

Tsuchida *et al.*²¹ reviewed 41 cases of Sturge-Weber syndrome treated at the Hospital for Sick Children in Toronto, including nine that were treated surgically. The follow-up data demonstrated that eight patients (20%) had only seizures without other neurological deficiency, 12 (29%) had slight to

moderate neurological impairment and epilepsy, and 12 had severe neurological deficiency. Nine cases were lost in the follow-up. In all patients who had severe neurological deficits, the onset of convulsive seizures was within 1 year of life. Ten of these patients were physically disabled, four were incapacitated, and the remaining four had severe mental retardation with moderate hemiparesis.

The well-known fact that tram-truck calcifications are seen on plain skull x-rays of older children, whereas those of infants appear normal,^{1-3,5,6,8,10,14} indicates that the process of brain calcification is progressive in nature. However, examination of surgical specimens has disclosed that the deposition of calcium in Sturge-Weber syndrome begins as early as 1 month of age and progresses overtime.^{1,10,13,18,19} The calcifications are thought to form as a result of diminution of the blood supply to the brain; the resulting hypoxia causes local alterations in pH and cellular damage, leading to gliosis and atrophy of the brain parenchyma.¹⁰ Thus, angiomas may induce not only calcification but also progressive destruction of the cerebral cortex through reduction of the blood supply to the involved hemisphere and consequent atrophy, seizures, and neurological deficits.⁴ Brain hypoxia or ischemia also produces gliosis of the white matter, which may be found in infants as young as 1 month of age and increases in severity. Cortical gliosis, which typically begins after the age of 3 or 4 months, is also progressive.¹⁰

II. Surgical treatment

To date, there have been few reports on surgical intervention in cases of Sturge-Weber disease. Balado was probably the first surgeon who performed electrocoagulation of a cerebral venous angioma, which reduced the frequency of convulsive seizures.⁴ This pioneering effort was followed, over the years, by lobectomies or partial resections.¹

One of the four surgical patients in the series of Peterman *et al.*¹⁸ displayed an apparent increase in intelligence after surgical excision; in addition, seizures and behavioral problems lessened. These investigators postulated that surgical excision of the diseased cerebral cortex offers a good chance for recovery of normal activities and prevention of the progression of neurological complications, if anticonvulsant therapy is ineffective in controlling seizures and the patient has not deteriorated past all hope of improvement.

In 1960, Falconer and Rushworth⁴ reported the results of five hemispherectomies. When these children were 3–8 years of age (median, 5 years) follow-up revealed that the seizures had disappeared

and all the patients had improved intellectually. The authors concluded that hemispherectomy may be confidently expected to abolish seizures and produce other tangible improvements, provided that the other hemisphere is structurally normal.

Hoffmann *et al.*¹⁰ reported the results of hemispherectomies in seven infants with this disease (1–10 months of age) performed from 1964 through 1979. Follow-up was from 1 to 13 years (median, 8 years), most patients were seizure-free and taking no anticonvulsants. One child with bilateral facial nevi experienced an occasional seizure, and another who developed hydrocephalus postoperatively, complicated by multiple shunt problems, died. However, all the surviving patients have normal IQs. The significant observations of Hoffmann *et al.* are that the younger the patient was at the time of hemispherectomy, the higher the eventual IQ and that there appears to be an inverse correlation between IQ and the duration of preoperative seizures. They added that if seizures begin in infancy and the disease is unilateral, the entire hemisphere is involved, and hemidecortication must be undertaken early if useful function is to be preserved. Our patient had had uncontrollable seizures for 10 months and the hemispherectomy was completed when he was 12 months old. In this case, therefore, surgery may have been performed too late to achieve and excellent outcome.

The surgical techniques for hemispherectomy have been well described.^{4,9,10,17} Since the pial vascular malformation in Sturge-Weber syndrome causes extensive bleeding during multiple lobectomies, the removal of the hemisphere “en block” is more appropriate. Dissection and obliteration of major vessels during this procedure are not difficult owing to cerebral atrophy. The use of a laser or ultrasonic aspirator may help to minimize intraoperative blood loss.

III. Results of surgery

In our case, posthemispherectomy neurological deficiency was surprisingly minimal. Hoffmann *et al.*¹⁰ observed a low incidence and severity of hemiparesis among children operated on in early infancy: gait was almost normal, arm function was preserved, and hand and finger dexterity improved to some degree. In our patient, although surgery did not entirely prevent psychomotor retardation, the procedure did eliminate his intractable seizures.

A wide range of variability has been observed in terms of functional recovery after hemispherectomy, and there is some controversy concerning the effect of hemispherectomy on infantile hemiplegia. This is attributable partly to differences in methodology and

interpretation of results and partly to the common assumption that the remaining hemisphere is intact, which is often incorrect.²⁰⁾ Although later-developing visual and spatial abilities of right hemidecorticate children have been reported to be severely impaired,¹²⁾ removal of the left hemisphere has rarely resulted in speech disturbance, and the majority of investigators have reported a rise in intellectual level following removal of the entire hemisphere.¹⁵⁾ The remarkable developmental potential of the remaining structures after right hemispherectomy was demonstrated in follow-up studies by Griffith and Davidson.⁷⁾ Also, Smith and Sugar²⁰⁾ followed a 5.5-year-old boy who underwent left hemispherectomy for seizures and found that, 21 years postoperatively, he possessed superior verbal and nonverbal intellectual abilities.

Thus, the results of hemispherectomy for infantile hemiplegia appear consistent with those of hemispherectomy for Sturge-Weber syndrome. The postoperative recovery of function in hemispherectomized patients may reflect compensatory mechanisms, either within the damaged hemisphere or in the opposite, intact hemisphere. The mechanisms involved in this phenomenon are not yet understood and should be studied further.

Acknowledgments

The authors wish to thank Drs. Masaya Segawa and Yoshiko Nomura, Segawa Neurological Clinic for Children, Tokyo, for their collaboration.

This study was partly supported by grants (62-E and 63-A) from the National Center of Neurology and Psychiatry (NCNP) of the Ministry of Health and Welfare, Japan.

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