SURGERY FOR WEST’S SYNDROME

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Many cases of West’s syndrome are inoperable, because there is no clear epileptic focus. In some cases, however, there is a structural abnormality which can be surgically removed. Dr. Hoffman discusses his experience with surgery for cortical dysplasia in West’s syndrome.

INTRODUCTION

West’s syndrome or “infantile spasms” is named after a physician by the name of West, who wrote a letter to Lancet in 1841 in which he described a “peculiar form of convulsion” associated with mental retardation as seen in his son. Clinically, West’s syndrome consists of a triad of myoclonic spasms, hypsarrhythmic EEG and mental retardation.

PATHOLOGY

The structural abnormality in symptomatic infantile spasms may be quite small or may be extensive involving an entire hemisphere. The usual abnormality is cortical dysplasia. A dysembryoplastic neuroepithelial tumor, however, which is a mixture of malformation and tumor, can also give rise to the abnormality. Patients with hemimeganencephaly, in which one hemisphere is diffusely enlarged with disorganized cortical laminae, can also present with infantile spasms. Patients with focal tubers and with a variety of tumors and cysts also present with infantile spasms.¹,²,³,⁴,⁵

Cortical dysplasia, however, is the commonest condition that gives rise to infantile spasms. Patients with cortical dysplasia benefit from surgical excision of the area of malformation.⁶ In cortical dysplasia, there is a cortical laminar disorganization. There may be heterotopic neurons in the white matter, and there can be subpial granular cells. There
also may be glioneural nodules in the subarachnoid space. The neurons may be enlarged and there can be "balloon cell" changes very similar to the balloon cell changes that one sees in tuberous sclerosis. Some of the patients with cortical dysplasia have lissencephaly, polymicrogyria and schizencephaly.

Focal cortical dysplasia is commonest in the central insular region. The irritative zone is usually much more extensive than the visible structural lesion. The dysplastic lesions can be visible or invisible on routine MRI. Occasionally they can be seen on CT and they are visible with PET scanning. Echoplaner MRI will usually show the lesion. The area of cortical dysplasia has an intrinsic epileptogenicity. The patients with balloon cells do not do as well as those without balloon cells.

SURGERY

Frameless stereotaxy has been very useful in defining the anatomical boundaries of focal cortical dysplasia. Electroctography is also useful in defining the electrical abnormality in these patients. Similarly, patients with dysembryoplastic neuroepithelial tumors, other tumors, cysts and focal tubers also benefit from frameless stereotaxy in defining the lesion and helping remove the entire lesion.

When cortical dysplasia involves one hemisphere—and the other hemisphere is normal—one can do a hemispherectomy. The type of hemispherectomy that I favor consists of a decorticating in which one peels off the cortex, while leaving the white matter. If one gets into the ventricle, this is repaired with surgical and fibrin glue. The calvarium (skull) over the side of the hemispherectomy is then morcellated to reduce its size so that there is very little space present between the residual hemisphere and the overlying skull.

Using this technique, we have never seen hydrocephalus, and the functional results are much better. Frequently hand movement and even finger movement is preserved.

RESULTS: THE TORONTO EXPERIENCE

Among 11 patients with symptomatic infantile spasms and cortical dysplasia who underwent surgery, none was older than eight months when their infantile spasms began. All 11 patients had focal cortical dysplasia. In four of these 11 patients a second operation had to be done to remove residual cortical dysplasia.

Five of the 11 patients are now completely free of seizures. Two patients have an occasional seizure. Two patients have only nocturnal seizures, and one patient is improved but still has an ongoing seizure problem.

DISCUSSION

Patients with poorly controlled symptomatic infantile spasms face a possible risk of Lennox-Gastaut syndrome, with intractable seizures and mental retardation. With

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<th>Table 1. Indications for Surgery in West's Syndrome</th>
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<td>- presence of a structural abnormality, such as cortical dysplasia</td>
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