

## Infantile Sturge-Weber Syndrome with Hypointense White Matter on T2-weighted MR images

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The implication of hypointense white matter on T2-weighted MR images in infants with Sturge-Weber syndrome is a subject of recent controversy. We report a case of infantile Sturge-Weber syndrome with decreased white matter signal on T2-weighted MR images. This hypointensity may reflect accelerated myelination and/or increased deoxy-hemoglobin in capillaries and dilated deep medullary veins.

**Key words :** Sturge-Weber syndrome, myelination, MR imaging

### Introduction

The accelerated myelination pattern seen in infants with Sturge-Weber syndrome (SWS) is recently a controversial subject. We know of seven previously reported cases of infantile SWS with hypointense white matter on T2-weighted sequences (1-4). We describe herein an infant with SWS, who presented hypointense white matter of the affected hemisphere on T2-weighted image.

### Case report

A newborn boy, born at term with asphyxia, was examined because of a seizure disorder and extensive port-wine nevi involving the left side of the face, left upper extremity and anterior chest, and lower extremities. Noncontrast CT, obtained at 1 month of age, showed slightly hyperdense left cerebral white matter but no cortical calcification (Fig. 1A). Postcontrast CT, performed on the same day, showed an enlarged, intensely enhancing left choroid plexus and gyral enhancement of the left cerebral hemisphere (Fig. 1B, C). A diagnosis of SWS with Klippel-Trenaunay syndrome was made. The initial MR image was obtained at the age of 2 months. On T2-weighted images, the signal intensity of the left cerebral hemisphere was lower than the right. On T1-weighted images, the intensity of the left cerebral hemisphere was greater than the right (Fig. 2). These findings were most apparent in the central part of the centrum semiovale and precentral and postcentral gyri. On follow-up MR using

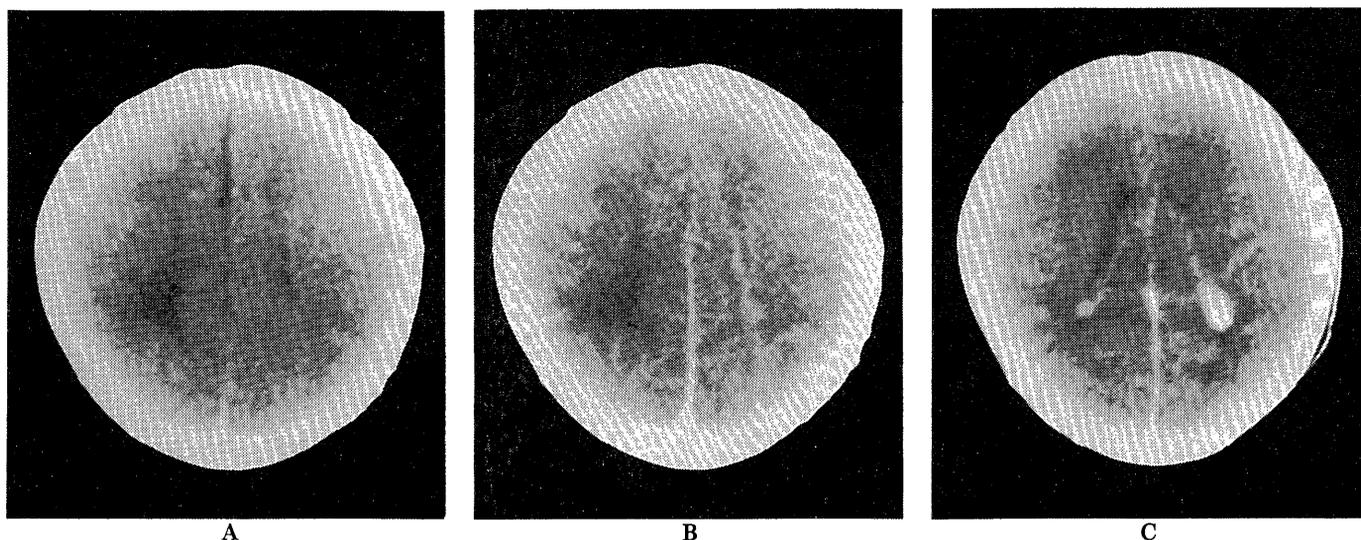
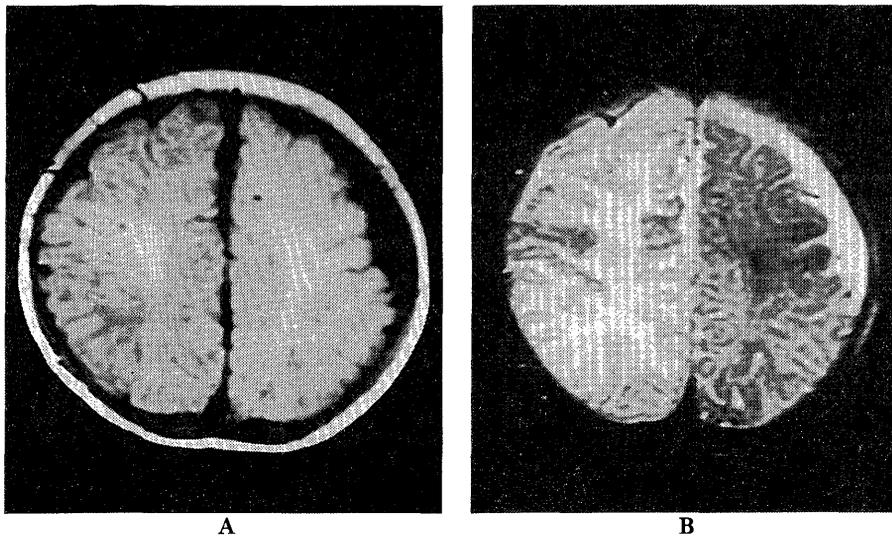
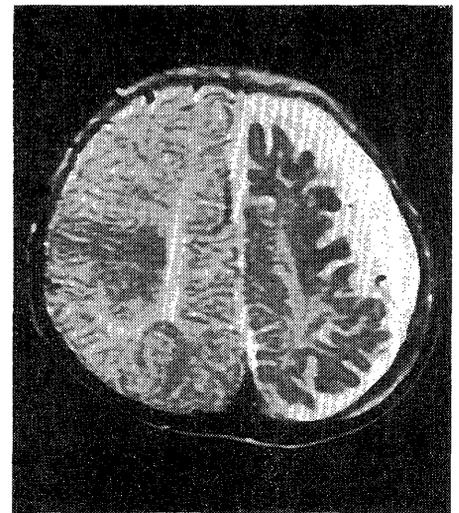


Fig. 1. CT obtained at 1 month of age. A : Precontrast CT. The density of the left centrum semiovale appears slightly greater than the left. No cortical calcification is seen. B, C : Postcontrast CT demonstrates large, strongly enhancing left choroid plexus and gyral enhancement on the left.



**Fig. 2.** MR images obtained at the age of 2 months.

- A : T1-weighted spin echo image shows hyperintense left cerebral white matter in comparison to the right side.  
 B : On T2-weighted spin echo image, the signal intensity of the left cerebral hemisphere is lower than the right.



**Fig. 3.** T2-weighted MR image obtained at the age of 8 months. The difference in signal intensity between the two hemispheres has become less apparent. Atrophy of the left cerebral hemisphere and a chronic subdural hematoma are seen.

the same T2-weighted sequence obtained at the age of 8 months, the difference in signal intensity of the two hemispheres was less apparent. The left cerebral hemisphere had become markedly atrophic and a chronic subdural hematoma secondary to the hemiatrophy had developed (Fig. 3).

## Discussion

SWS is a neurocutaneous syndrome (phakomatosis) characterized clinically by a facial port-wine nevus, a seizure disorder, and mental deficiency (5). Central nervous system manifestations include a leptomeningeal vascular malformation (angioma) and cerebral atrophy, which are typically ipsilateral to the facial nevus. Calcifications that are known to develop in atrophic cortex deep to the leptomeningeal venous malformation are rarely seen before 2 years of age. The enlarged choroid plexus is thought to be a collateral pathway for venous drainage, and gyral enhancement is attributed to leptomeningeal angiomas (5). The enlarged, intensely enhancing choroid plexus and superficial meningeal enhancement, demonstrated in our case, seem to be important diagnostic findings in infantile SWS patients who have no cortical calcifications.

Patients with SWS usually do not have cutaneous angioma involving the extremities. However, over 40 cases of associated SWS with Klippel-Trenaunay syndrome, similar to our case, have been reported (6). SWS and Klippel-Trenaunay syndrome are probably disembyo-

plasias that differ only in location of the lesion and in severity of involvement (6).

MR findings in cases of SWS have been well described, but reports of infants with SWS are sparse (1-5, 7). Of these reports of infantile cases, only four refer to the signal intensity of the white matter; all of the seven cases in the literature and our patient exhibited hypointense white matter of the affected hemisphere on T2-weighted images (1-4). These findings are seen only in infants with immature myelination pattern and not in older children. While the nature and pathogenesis of the low-intensity white matter of the affected hemisphere on T2-weighted sequences remains unsolved, three explanations have been proposed. One is that the lower intensity may reflect accelerated myelination. Jacoby et al. described a pattern of accelerated myelination affecting the involved hemisphere in two infants with SWS demonstrated by MR (1). Our case is considered to support this theory; MR demonstrated T2- and T1-shortening of the affected white matter; these findings were most apparent in the central portion of the central semiovale, where normal myelination occurs earlier than the surrounding white matter (8). On follow-up MR using the same T2-weighted sequence, the difference in signal intensity of the two hemispheres became less apparent due to the normal myelination in the contralateral hemisphere. In a histological study, Vannucci demonstrated hypermyelinated areas in the perinatal hypoxic-ischemic lesions (9). Because of poor venous drainage in the affected brain in SWS, the cerebral circulation is insufficient (5). Accelerated myelination may be the result of chronic ischemia due to impaired

venous drainage. Another suggestion is that the T2-shortening may be caused by increased deoxyhemoglobin in capillaries and veins (4). Presumably, the increased levels of deoxyhemoglobin is the result of decreased superficial drainage and collateral flow of deoxygenated blood through the enlarged deep medullary vein into the deep venous system (10). The third possibility is the deposition of ferritin or calcium (2). However, pathologic study and CT demonstrated that the calcium deposition was confined to the subcortical white matter, the middle layers of the cerebral cortex (No. 2-4), and the wall of the small intracerebral vessels of the involved hemisphere (5).

In conclusion, intensely enhancing large choroid plexus and gyral enhancement seem to be the diagnostic features in infants with SWS who have no cortical calcifications. Radiologists must be aware of these findings and should not rule out SWS because of the absence of cortical calcification. The T2-shortening of the affected white matter may reflect accelerated myelination or increased deoxyhemoglobin in the capillaries and medullary veins. The true cause of the T2-shortening will remain unknown until further MR and pathologic studies of SWS infants can be conducted on a larger number of patients.

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