Imaging of the Sturge-Weber Syndrome

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Purpose: The purposes of this article are to illustrate the typical imaging features of eight patients with this syndrome and to discuss the advantage of each imaging modality with a concise review of literatures.

Materials and Methods: We retrospectively reviewed plain skull radiographs (6), computed tomographic (CT) scans (8), magnetic resonance (MR) images (4) and cerebral angiograms (3) of eight patients with Sturge-Weber syndrome. We analyzed the radiographic findings of Sturge-Weber syndrome and compared the findings of CT, MR and angiography.

Results: Plain radiographs showed characteristic gyriform calcifications (3) after 2 years of age. CT scans excellently demonstrated cortical calcifications (5), prominently enhancing choroid plexi (5) and dilated periventricular veins (2). MR revealed dilated deep cerebral veins as tubular or spot-like signal void structures at periventricular areas (3) and showed stripes of cortical enhancement after gadolinium injection (2). Angiography showed dilated tortuous medullary and deep cerebral veins (3) as the collateral pathways of blood shunting. MR was superior to CT in the detection of parenchymal atrophy, venous abnormalities and the extent of angiomatosus involvement. Angiography showed enlarged deep cerebral or medullary veins better than MR imaging.

Conclusion: We think that each imaging modality including CT, MR or angiography has unique advantages in the diagnosis of this syndrome but MR will be used frequently because of its superior ability for the detection of atrophy, vascular abnormalities and direct visualization of leptomeningeal angiomatosis with contrast enhancement.

Index Words: Brain, Sturge-Weber syndrome
Brain, Computed tomography
Brain, Magnetic resonance
Brain, Angiography

INTRODUCTION

Sturge-Weber syndrome is one of the neurocutaneous syndromes characterized by facial vascular nevus and leptomeningeal venular angiomatosis which is responsible not only for various neurologic manifestations such as seizure, dementia, hemiplegia, or hemianopsia but also for characteristic radiologic findings such as brain atrophy, cortical calcifications as the primary or secondary changes of the basic pathology (1). Sturge first described this rare condition in 1879 (2) and Weber demonstrated the characteristic intracranial calcifications in 1922 (3). With the advent of sectional imaging modalities, CT and MR are used more frequently than the plain skull radiography or angiography which used to be the primary diagnostic imaging modalities in the past. The purpose of this article is to illustrate the typical imaging features of eight patients with this syndrome we experienced during the past 5 years at our institution and to discuss the advantages of each imaging modalities with a concise review of the literatures.
SUBJECTS and METHODS

Eight patients with Sturge-Weber syndrome, four males and four females with age ranging from 3 months to 17 years (mean 4 years and 11 months), were included in this study. Seven patients had facial angiomas which were ipsilateral to the hemispheric lesions in five and bilateral in two patients. One patient had no facial angioma but diagnosed as Sturge-Weber syndrome because of the typical imaging features. One patient with bilateral facial nevi had somewhat different cutaneous manifestations from other patients. There were multiple port-wine nevi involving face, trunk and extremities and hemihypertrophy of right upper extremity but no venous varicosities on the affected limb. Plain skull radiography was performed in 6 out of 8 patients. CT images were obtained in all patients with and without contrast enhancement on CT 9800 or 8800 scanners (GE Medical Systems, Milwaukee WI). MR imaging was performed in four patients on a 2.0T or 0.5T unit (Goldstar, Seoul, Korea) but contrast enhancement was done in only two. Cerebral angiography was performed in three patients. We analyzed the radiographic images of eight patients with Sturge-Weber syndrome especially in the points of brain atrophy, cortical calcifications, choroid plexus abnormality, leptomeningeal enhancement pattern, and vascular abnormality. Also we compared CT, MR, and angiography in three patients. The clinical data and radiographic findings including CT scan, MR imaging, angiography are summarized in the Table 1.

RESULTS

Plain skull radiography

Tram-line like gyriform intracranial calcifications were revealed in three out of six patients and located at parietooccipital areas on the lateral view. The age of three patients who showed definite calcifications on the plain radiographs was 2 years, 3 years and 17 years, respectively. Also, atrophy of hemicalvarium was present in two, ipsilateral bone and sinus hypertrophy was seen in one patient.

CT scans

CT scan of a neonate was normal but follow-up scan after 4 months showed increased attenuation of affected lobes and homogeneous enhancement after contrast medium injection (Fig. 1a, b). Definite gyriform cortical calcifications and parenchymal atrophy were demonstrated in five patients on noncontrast CT scans (Fig. 1c). No definite calcifications were detected in three patients but diffuse gyral enhancement

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Fig. 1. (case 7) a. CT findings of a neonate with seizure. Noncontrast CT scan at 2 weeks after birth reveals no definite abnormality. b. Repeated noncontrast CT study 4 months later shows increased attenuation of right frontotemporal lobes (arrows) which is considered due to the presence of microcalcifications at affected lobes. c. Follow-up CT 3 years later. Marked calcifications and atrophy of right hemisphere are evident.

was noted (Fig. 2a). An abnormally large choroid plexus with intense enhancement in the same side with the facial angiomata and intracranial lesions were noted in five patients (Fig. 3). Enlarged deep cerebral vein was identified in two patients as tortuous tubular enhancing structures at the periventricular area (Fig. 3b). Hemicalvarial thickening along the affected lobes was present in one patient.

**MR imaging**
Spin-echo pulse sequence MR imaging was performed in four patients. It was not possible to detect calcifications in spin-echo MR imaging but mild atrophy of the affected lobes which was indistinct on CT scans was demonstrated clearly on T2 weighted axial images in two patients (Fig. 2b). The parenchymal venous abnormalities which were not detected on CT scans appeared as tubular or spot-like signal void structures at periventricular or subependymal areas in three patients (Fig. 3b). Images obtained after contrast enhancement showed stripes of cortical enhancement covering the affected lobes in two patients (Fig. 4). Abnormal low signal intensity of the white matter of the affected hemisphere was present in one infant on T2 weighted images.

**Angiography**
Angiography revealed markedly enlarged deep medullary veins which were not demonstrated on CT scans or MR images in two patients (Fig. 2c, d). The number of superficial cortical veins in involved areas decreased when compared with the contralateral side in all three patients. Tortuous dilated deep cerebral veins including internal cerebral veins, balsal veins of Rosenthal were noted in three patients. Faint or nonvisualization of superior sagittal sinus was noted in two patients on the angiogram of the involved side. Dense capillary blush in some portion of the affected cortex was noted in two patients.

**DISCUSSION**

Characteristic intracranial calcifications on plain skull radiographs may not appear within the first two years of life. Because similar gyriform calcifications have been described in various other conditions including infarction, purulent meningitis, viral encephalitis, tuberous sclerosis, it is not a pathognomonic feature of this disease (4-7). Atrophy of hemicalvarium, ipsilateral bone and sinus hypertrophy may be present as a compensatory response to the volume loss of the brain parenchyma.

CT scan may be normal especially in infants or shows subtle changes such as mild brain atrophy, increased attenuation of the affected lobes on the noncontrast scan, diffuse gyral enhancement after contrast medium injection, but progression of mineralization and volume loss of the affected lobes are evident on follow-up CT studies (Fig. 1) (8). CT often detects calcifications before the age of 1 year, as early as 4 months after birth in our series (Fig. 1), and is more sensitive in demonstrating the characteristic cortical calcifications than spin-echo MR imaging. Pathologically, calcification is located in the atrophied cortex beneath the leptomeningeal angiomata and is thought to be related to chronic tissue hypoxia (9). However, on the basis of the pathologic studies, the extent of parenchymal abnormalities including infarction, gliosis, demyelination is known to occur in wider regions than the obviously calcified or atrophied parenchyma revealed by CT (10). An abnormal large choroid plexus in the same side with the
Fig. 2. (case 1) a. Enhanced CT scan shows diffuse gyral enhancement in the right parietooccipital areas and prominent, well enhancing choroid plexus (arrow) but no definite calcifications on noncontrast scans (not shown).
b. Axial T2 weighted MR image (0.5T spin echo TR/TE 3000/100) shows sulcal widening of the right frontotemporal area and multiple tubular signal void structures (arrows) at the periventricular area.
c. Lateral angiogram at mid-venous phase demonstrates fine numerous dilated medullary veins draining to the ventricular ependyma and tortuous dilated internal cerebral vein (arrows).
d. Convergence of dilated medullary veins to the periventricular area and enlarged thalamostriate vein (arrows) are well visualized on the anteroposterior view at late venous phase.

Fig. 3. (case 5) a. Noncontrast CT scan demonstrated brain atrophy, prominent choroid plexus (arrow) within lateral ventricle, and calcifications of the left cerebral hemisphere.
b. With contrast medium injection, prominent enhancing choroid plexus (arrow) within the lateral ventricle is evident and enlarged subependymal veins (arrow head) are also observed.

Fig. 4. (case 2) Contrast enhanced T1 weighted axial MR images (0.5T spin echo TR/TE 600/40) shows diffuse enhancement with a stripe pattern following convolutions of left cerebral hemisphere. This cortical enhancement is thought to represent the extent of leptomeningeal angiomatosis of this patient.
facial angioma and intracranial lesions represent angiomatic involvement of the choroid plexus itself and are better observed after administration of contrast agent(11).

Spin-echo MR is inferior to CT in demonstrating the characteristic intracranial calcifications and shows foci of low signals in the regions of very heavy calcification but recently MR with gradient echo acquisition has proved to be more sensitive than the spin echo MR or even the CT for the detection of diffuse fine parenchymal calcifications(12, 13). MR imaging is better in showing the degree of parenchymal atrophy and venous abnormalities than CT(Fig. 2)(13-15). Noncontrast MR alone underestimates the extent of intracranial disease significantly. Therefore contrast enhancement is necessary for the complete MR evaluation of patients(12). In our experience and others, contrast enhanced MR imaging shows a stripe of cortical enhancement covering the affected lobes(Fig. 4)(12, 13, 16). This cortical enhancement is considered to include the leptomeningeal angioma itself and the first cortical layer affected by gliosis(16, 17). Direct visualization of leptomeningeal angioma is important in order to ascertain the diagnosis when CT is normal especially in infants, or to know the exact extent of the lesions. Contrast enhanced MR imaging well correlates with the pathologic findings in the distribution of leptomeningeal angiomatosis(16). Jacoby et al. proposed accelerated myelination in early Sturge-Weber syndrome as the explanation for the abnormal signal intensities of white matters of the affected hemisphere (18). But the exact causes of abnormal signal intensities of the white matters of affected hemisphere in infants are unclear and MR-pathology correlative studies will be necessary(14).

The basic features of cerebral venous drainage abnormalities in this syndrome are decreased superficial cortical veins in the involved areas. As a result of decreased superficial cortical venous drainage, marked enlargement of deep medullary veins can develop as collateral pathways. Through these routes, centripetal venous flows result in enlarged tortuous deep vein such as internal cerebral veins, basal veins of Rosenthal and their branches(Fig. 2c, d)(9). Angiography demonstrated these venous abnormalities more clearly than others but these findings are not pathogonomic for Sturge-Weber syndrome because those findings may occur with loss of cortical veins from any other cause.

One patients in our series(case 2) showed typical radiographic features of Sturge-Weber syndrome but cutaneous manifestations of the patient are somewhat different from other patients, showing some features of Klippel-Trenaunay-Weber syndrome which is characterized by the classical triad of cutaneous portwine nevus, osseous and soft tissue hypertrophy, and venous varicosities of the affected body part. This rare syndrome can occur in combination with one of the neurocutaneous syndromes, especially with Sturge-Weber syndrome(20).

The relationship between intracranial lesions and facial angioma in this syndrome is variable, usually ipsilateral but bilateral or even contralateral relationship is possible(21). Sturge-Weber syndrome without facial angioma has been reported(22-24) and in this rare situation, the diagnosis depend solely on the characteristic findings of radiologic studies, especially CT scans or MR imaging, as in case 3 of our series.

In conclusion, we think that each imaging modality including CT, MR, or angiography has the unique advantages in the diagnosis of the syndrome but MR is more useful than others because of its superior ability for the detection of parenchymal atrophy, vascular abnormalities and direct visualization of the leptomeningeal angioma with contrast enhancement. Diagnostic value of angiography appears to be less important and might be replaced by noninvasive contrast enhanced CT scans or MR imaging.

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Sturge-Weber Syndrome의 영상진단

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목적: 본 논문의 목적은 Sturge-Weber 중후군의 특징적인 방사선학적 소견 및 각 영상진단방법, 즉 두부 전산화 단층촬영(CT), 자기공명영상(MR), 뇌혈관조영술간의 차이점을 파악하고자 하는데 있다.

대상 및 방법: Sturge-Weber 중후군 환자 8명에서 시행한 단순 두개골촬영 6예, CT 8예, MR 4예, 뇌혈관조영술 3예를 두개강내 석회화, 뇌실질의 위축, 맥락막층의 이상유무, 뇌혈관 이상유무 및 비정상적인 조영증강을 중심으로 후향적으로 분석하였고 이중 3명에서 CT, MR, 뇌혈관조영술의 소견을 비교하였다.

결과: 단순촬영은 특징적인 뇌피질 형태의 투게강내 석회화(3)를 보일수있으나 2세 이전에는 잘 보이지 않았다. CT에서는 뇌피질의 석회화(5), 비후된 맥락막층의 조영증강(5), 확장된 심부 뇌정맥(2)이 보였으며 MR에서는 확장된 심부 뇌정맥이 측뇌실 주변에서 무신호강도로 보이고(3) 조영증강후 병변부위에 얇은 맥락막조영증강이 뇌피질을 따라 보였다(2). 뇌혈관조영술에서는 뇌수질정맥 및 심부뇌정맥이 확장된 쌍(3)을 보였다. MR은 CT와 비교했을때 뇌피질의 위축, 뇌혈관이상 및 연뇌막 혈관중증의 발견에 있어서 우수하였고 뇌혈관조영술은 뇌수질정맥 및 심부뇌정맥이 확장된 소견을 MR이나 CT보다 잘 보여주었다.

결론: CT, MR, 뇌혈관조영술은 이 질환의 영상진단에 있어서 각각 장점이 있으나 특히 MR은 뇌혈관이상 및 조영증강되는 연뇌막 혈관중증을 잘 보여줌으로써 가장 우수한영상진단방법으로 생각된다.