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The Sturge-Weber syndrome: correlation between the clinical status and radiological CT and MRI findings

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Abstract. In the Sturge-Weber syndrome facial venous and leptomeningeal angiomas are associated. We studied 14 consecutive cases with clinical and radiological evaluations [computed tomography (CT) and magnetic resonance imaging (MRI)]. Radiological studies demonstrated the extent and patency of the leptomeningeal angiomatous malformation, the degree of parenchymal atrophy, the presumed ischemic changes affecting the gray and white matter, the presence and extent of cortical calcifications, the prominence of the choroid plexus, the parenchymal venous anomalies, and the diploic prominence in the affected side. The white matter alterations, being greater, the extent of lobar involvement, and the degree of parenchymal atrophy all correlated with the patient's clinical status being poorer. MRI is more efficient in the detection of the radiological findings related to the clinical status: seizure control, degree of psychomotor development and hemiparesis. Therefore, MRI is the imaging modality of choice in the diagnosis of these patients.

Key words: Sturge-Weber syndrome – Computed tomography – Comparative studies – Magnetic resonance – Comparative studies

Introduction

The Sturge-Weber syndrome (SWS) is a neurocutaneous syndrome characterized by leptomeningeal angiomatosis (LMA) and an ipsilateral facial nevus in the areas of the ophthalmic division of the trigeminal nerve. Clinical findings include seizures, contralateral hemiparesis, progressive mental retardation, and glaucoma. Computed tomography (CT) and magnetic resonance imaging (MRI) are the diagnostic imaging modalities usually employed in this syndrome [1–3, 6, 8].

In general, there is little correlation between the imaging findings and clinical status in neurocutaneous syndromes [4]. In the present study we investigated the relationship between the imaging findings (CT and MRI) and clinical status in SWS. We also attempted to establish an imaging algorithm at the time of diagnosis and follow-up studies of the syndrome.

Patients and methods

Fourteen consecutive children (9 male and 5 female) were studied. Their ages varied between 2 and 21 years (mean 10 years). The clinical alterations were evaluated in all cases, with particular emphasis on the presence of neurological deficiencies, hemiparesis, and convulsions. The education level and the intelligence quotient (IQ) using the Terman-Merrill or Weschler scale were also evaluated. A normal child was taken to be one with an education level in agreement with the patient's age and/or an IQ of over 90. Patients were considered to be borderline when they required support and/or presented an IQ of between 70 and 90. Finally, mental retardation was defined as IQ values of under 70.

CT was done in all 14 cases (General Electric 9800 and Toshiba TCT-600HQ) with and without IV contrast agents. A bolus of contrast medium was given by hand (2–3 ml/kg of 300 mg I/ml solution). Two successive studies were made in all cases, separated by an average of 5 years (interval, 1.5–10 years). MRI was performed in 11 children with a 0.5 T superconductive unit (Philips Gyroscan S5). MRI studies were performed within 1 year of the second CT examination. All patients had a sagittal short TR/short TE spin-echo (SE) sequence (TR = 505 ms/TE = 30 ms) and a transverse SE long TR/double-echo sequence (TR = 2200–2500 ms/TE = 50, 100 ms). In 8 cases a gadolinium (Gd-DTPA)-enhanced T1-weighted gradient-echo (GRE) study (TR = 569 ms/TE = 16 ms/90°) was performed in the transverse and coronal plane. In five instances a STIR sequence (TR = 1500 ms/TI = 100 ms/TE = 44 ms) in the coronal plane was also used.

In both CT and MRI, a systematic evaluation was made of the degree of atrophy, the presence of calcification, contrast medium enhancement, gray and white matter alterations, choroid plexus anomalies, diploic enlargement, and the presence of venous anomalies. These abnormalities were correlated with the clinical status by a visual grading scale.

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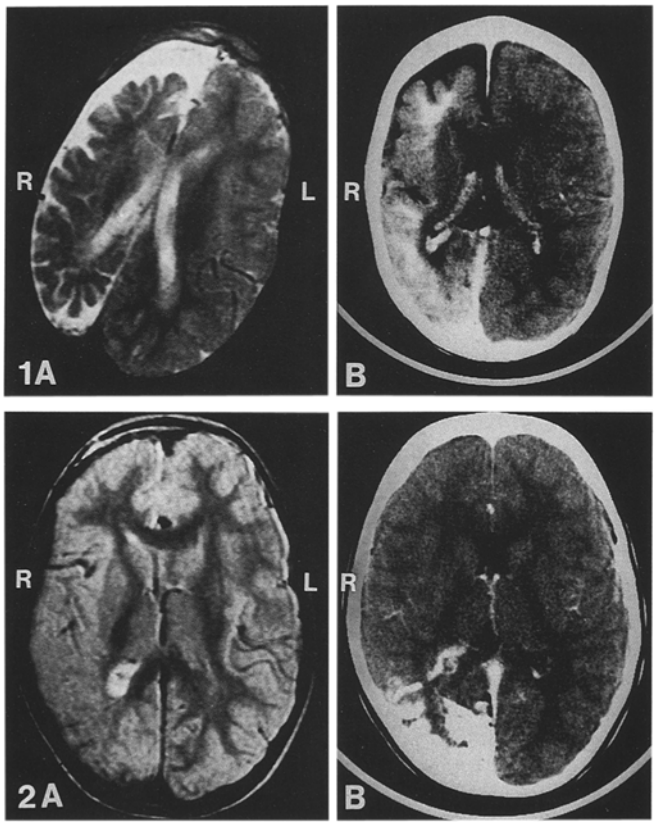

Fig. 1 A, B. A 2-year-old patient with right facial angioma. Focal seizures began at 3 months old and are present despite medical treatment. There is borderline psychomotor delay and left hemiparesis. **A** T2WI (SE 2200/100) shows marked hemispheric atrophy. White matter changes are not demonstrated. **B** CT demonstrates diffuse parenchymal calcification in the right hemisphere

Fig. 2 A, B. A 12-year-old patient with right facial angioma. Focal seizures began at 4 months, but for 4 years the patient has been free of them. Hemiparesis is discrete and psychomotor development borderline. **A** PDWI (SE 2200/50) shows right occipital atrophy with the white matter being isointense to the cortex. Note also the prominent homolateral choroid plexus. **B** CT shows occipital calcification, making it impossible to demonstrate volume loss and white matter changes

Results

All cases were sporadic, except for one probably dominant autosomal case – the mother also presented the syndrome but was not included in the present study.

All patients presented the characteristic facial angioma. This was ipsilateral to the cerebral involvement in 12 cases, bilateral in 1 – with left LMA – and contralateral to the LMA in another patient. Psychomotor retardation or a diminished IQ was observed in 3 patients; 6 were considered to be borderline cases, and 5 were normal. All patients presented convulsions. In 10 cases (71%) the epileptic crisis began within the first year of life the rest varying between 18 months and 16 years. Crisis control was good in 9 patients – all either without psychomotor retardation or borderline – and poor in 5 – 3 with psychomotor retardation. Six patients revealed hemiparesis contralateral to the LMA.

Table 1. Correlation between clinical and radiological findings. PsR, Psychomotor development; PCC, poor control of epileptic crisis; HP, hemiparesis

	CT	MRI	PsR	PCC	HP
No. of cases	14	11	8	5	6
Brain atrophy, discrete	0	8	3	1	0
Brain atrophy, severe	6	6	5	4	6
White matter changes	0	9	6	5	5
Gray matter changes	0	6	3	2	3

The extent of cerebral involvement was best defined by MRI (Figs. 1, 2). CT incorrectly showed the extension in one patient, in whom it failed to detect the involvement of a temporal lobe seen with MRI. The involvement was occipital in two, temporo-occipital in four, parieto-occipital in one, hemispheric in three, and frontoparietotemporal in one patient. Cerebral lobar atrophy could be equally well evaluated with CT and MRI. However, when there was no coexistent prominence of sulci, CT failed to detect the true degree of volume loss (Fig. 2). In all cases cerebral cortical calcifications were demonstrated by CT, although MRI failed to show them in 35% of cases. The white and gray matter changes were only detected on MRI (Figs. 1, 2). This finding was best appreciated on SE images with PDWI and STIR sequences. After contrast administration, cortical enhancement was noted in all cases with MRI, but CT failed to show it in 15% of cases. The choroid plexus was enlarged with respect to the contralateral plexus in all patients. This finding was best appreciated on enhanced examinations with either CT or MRI. Abnormalities in venous development, consisting of prominant deep, subependymal veins and venous angiomas, were best seen with MRI in five patients. Prominent diploës with respect to their contralateral parts were better visualized by MRI than by CT in nine children.

No temporal changes were noted on the serial CT scans in 9 of the 14 children. In the other 5 there was a decrease in the degree of cortical contrast enhancement; 3 had also an increase in calcification and 2 had an increase in atrophy. In no case did the choroid plexus change.

The correlation between clinical and relevant radiological findings is shown in Table 1. The extent of LMA, the degree of atrophy and white matter alterations were related to control of the seizures and the degree of hemiparesis and psychomotor development. However, there was no correlation with alterations in the gray matter signal intensity, the prominence of the choroid plexus, parenchymal venous anomalies, or the degree of cortical calcifications.

Discussion

Sturge-Weber syndrome (SWS) is a pediatric neurocutaneous syndrome. The clinical status is characterized by epilepsy, generally commencing before the first year of life, hemiparesis and psychomotor retardation. However,

a few may have no clinical manifestations and not even the syndrome's characteristic facial angioma [5].

The extent of cerebral involvement is usually well defined by both CT and MRI. However, due to its greater contrast resolution and absence of bone artifacts, MRI with contrast enhancement facilitates LMA visualization in the peripheral areas that are more difficult to evaluate with CT (e.g., the temporal lobe). Characteristically, cerebral involvement is occipital and ipsilateral to the facial angioma, with greater or lesser participation of the other lobes. Associated involvement includes the temporal, parietal, and frontal regions in order of decreasing frequency. Cerebral atrophy is usually present in the evolution of these patients. Meningeal and cortical involvement was seen as an area of increased enhancement following Gd-DTPA administration in the same localization as the area of high signal intensity observed with SE long TR/medium TE and STIR sequences. We believe [3] that this area includes the LMA [6] and also the first cortical layer affected by gliosis secondary to chronic hypoxia and/or epilepsy proper [1, 7]. Inside this layer, a hypointense calcified ribbon can be seen. Areas of increased signal also appear in the adjacent white matter with T2-weighted and STIR sequences. This hyperintensity makes the white matter appear as intense as the gray matter if not more so. It is possibly secondary to ischemia-related gliosis [1, 3, 7]. The choroid plexus is prominent at the atrial level in these patients. In longitudinal CT studies, no changes in appearance of the plexus were observed over time. In older children with severe involvement, brain atrophy is associated with a thickening and prominence of the diploë. Venous alterations are also common. The surface venous drainage of the affected lobes is shunted to the deep venous system via collateral vessels in a subependymal, periventricular or transparenchymal location [1].

Correlation with seizure control, the degree of psychomotor development, and hemiparesis is reasonably well established as a function of the extent of LMA, the degree of atrophy, and white matter alterations. Patients with these alterations showed a poorer clinical status, with more epileptic crises and less success in controlling them medically, the presence of hemiparesis, and a more severe psychomotor delay. No clinical correlation was observed in the present study with the remaining alterations evaluated with radiological methods.

CT demonstrates cortical calcifications better than MRI. However, MRI clearly showed most of the findings associated with SWS (LMA malformation, parenchymal atrophy, ischemic changes in the affected gray and white matter, prominent choroid plexus, venous anomalies and diploic prominence). MRI, with and without paramagnetic contrast material, is more efficient in detection of the findings related to the clinical status (white matter alterations, extent of lobar involvement, and brain atrophy). Therefore, MRI, done with T2-weigthed spin-echo and T1-weighted sequences after Gd-DTPA administration, should be used in radiological evaluation of the SWS. With these MRI sequences, all of the parameters correlated with clinical status can be observed. Follow-up imaging studies are not justified, although a new MRI study seems reasonable before pediatric discharge to evaluate the final degree of involvement. MR angiography would probably add no additional clinically relevant information.

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