CT DIAGNOSIS OF STURGE-WEBER SYNDROME : ONE CASE REPORT

Sirirat Kittiwongsopon
Pradit Chaiyabud
Puripakorn Pakdirat
Ratchaburi Hospital

ABSTRACT :

Kittiwongsopon S, Chaiyabud P, Pakdirat P. CT Diagnosis of Sturge - Weber Syndrome : One Case Report. (Region 7 Medical Journal 1994 ; 3 : 263-269.)

Department of Radiology, Department of Surgery, Department of Anatomical Pathology, Ratchaburi Hospital, Ratchaburi, Thailand.

A 12 - year - old male patient with headache and congenital facial port wine stain was presented, the report describes the classic features and CT findings of the Sturge - Weber syndrome, emphasizing the importance of CT as a useful diagnostic procedure.

บทคัดย่อ :

ศิริรัตน์ กิตติวงศ์โสภณ, ประดิษฐ์ ไชยบุตร, ภูริปกรณ์ ภักดีรัตน์. CT Diagnosis of Sturge - Weber Syndrome : One Case report. (วารสารแพทย์เขต 7 2537 ; 3 : 263-269.)

กลุ่มงานรังสีวิทยา, กลุ่มงานศัลยกรรม, กลุ่มงานพยาธิวิทยากายวิภาค, ร.พ. ราชบุรี.

ผู้ป่วยเด็กชายไทย อายุ 12 ปี มีปานแดงที่ใบหน้าตั้งแต่เกิด มาโรงพยาบาลด้วยอาการปวดศีรษะ รายงานฉบับนี้ได้บรรยายลักษณะเฉพาะของ Sturge - Weber syndrome ที่พบและจากการตรวจสมองด้วยเอกซเรย์คอมพิวเตอร์ โดยเน้นความสำคัญของเอกซเรย์ คอมพิวเตอร์ในการวินิจฉัย

Sturge - Weber syndrome, also called encephalotrigeminal angiomatosis, is a congenital neurocutaneous syndrome characterized by facial port wine stain and intracranial angiomatosis. Clinical manifestation with seizure, mental retardation, hemiparesis and visual deficits, which is progressive with worsening, is associated with continued development of calcifications in the vascular defects[1].

Report

The patient was a 12 - year - old thai male who had been diagnosed as having Sturge - Weber syndrome with the triad of classic congenital left facial port wine stain involving the distribution of the opthalmic branch of the trigeminal nerve (fig 1), seizures and gyral calcification at the left parieto-occipital region.

Skull radiography showed no abnormal calcification except hyperpneumatization of sinus change secondary to developmental change in the brain. (fig 2)

Computerized tomography without contrast injection demonstrated left occipitoparietal gyral calcification and ipsilateral temporoparietal hemiatrophy. (fig 3)

Contrast enhanced scan revealed collateral subependymal vessels (fig 4) and focal abnormal vessels at trigone of Lt ventricle, implied venous angiomatosis. (fig 5)

Left occipitoparietal craniectomy was performed, intraoperative findings showed left occipitoparietal leptomeningeal angiomatosis. (fig 6)

Removal of large vascular choroid plexus in the left trigone was done and pathological examination proved to be cavernous hemangioma of the left trigone. (fig 7)

Discussion

Different degrees of cerebral calcification, encephalofacial angiomatosis and seizures characterized the Sturge - Weber syndrome.[2]

Intracranial abnormalities include leptomeningeal angiomatosis with an underlying superficial layer of gyral cortical calcification, angiomatosis malformations of the choroid plexus and ipsilateral atrophy. Abnormal local venous drainage occurs with non functional or absent cortical veins and patent deep veins.[3, 4]

These abnormalitics have been well demonstrated by neuroradiologic studies including skull films, angiography and computerized tomography. The value of contrast MRI for detecting leptomeningeal angiomatosis has recently been described.[5 - 9]

The classic change of Sturge - Weber syndrome, "tramline" type calcification along the cortical gyri are demonstrated by skull radiographs in older children after 2 years of age and adult or earlier by cranial CT.[2, 10]

An infantile form of Sturge - Weber syndrome in young children (usually 2 years and younger) can be seen on MRI. Calcification is not present.[10]

In view of the small calcification, CT has an advantage over MR and conventional plain film of detecting and it can appear as early as 3 months of age.[3, 7, 8, 11, 12]

Calcium deposits may be found in the wall of cerebral vessels, in the perivascular tissue and early within the neurons, these are commonly interpreted

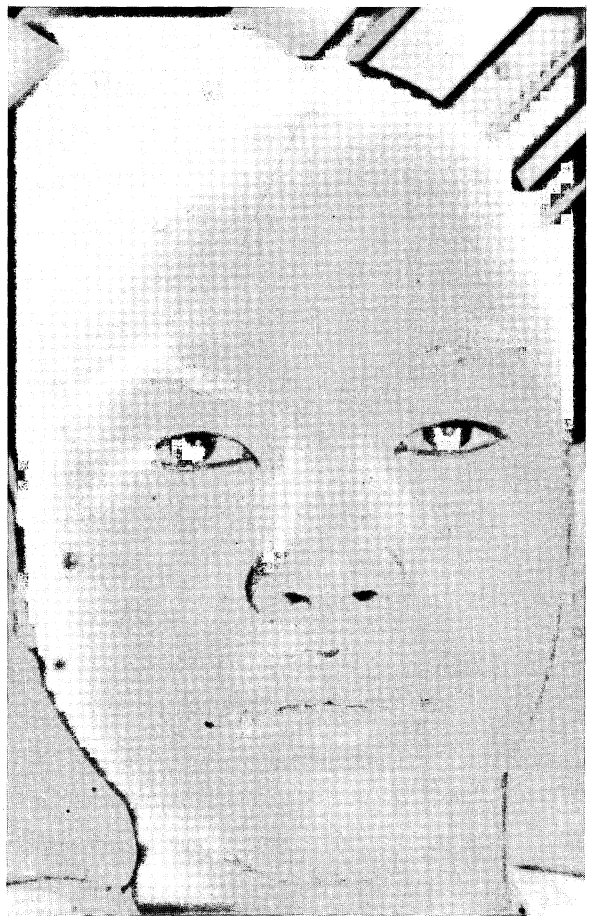


Fig. 1 Characteristic facial port wine stain in the distribution of the first trigeminal nerve on the left face of an older child with Sturge - Weber syndrome.

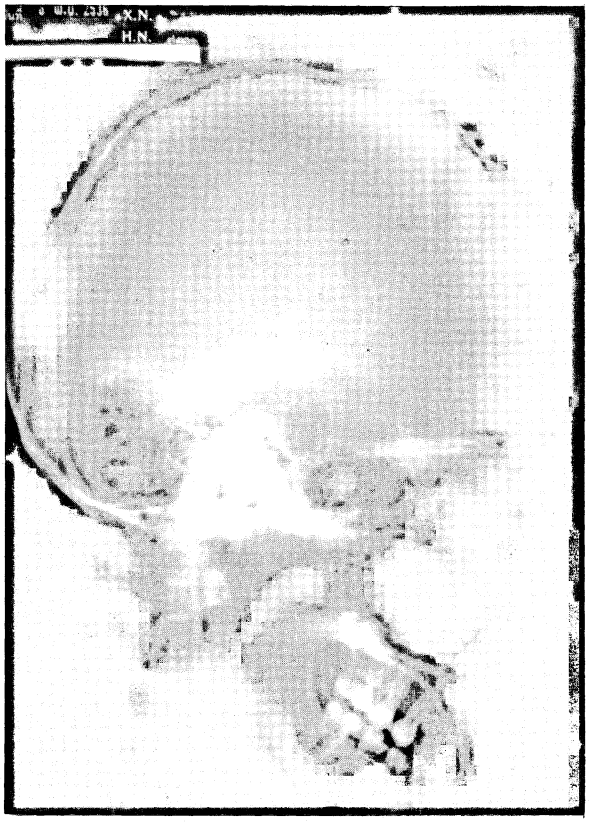


Fig. 2 Lateral skull radiograph, showing hyperpneumatization of upper air sinus, without abnormal cerebral calcification.

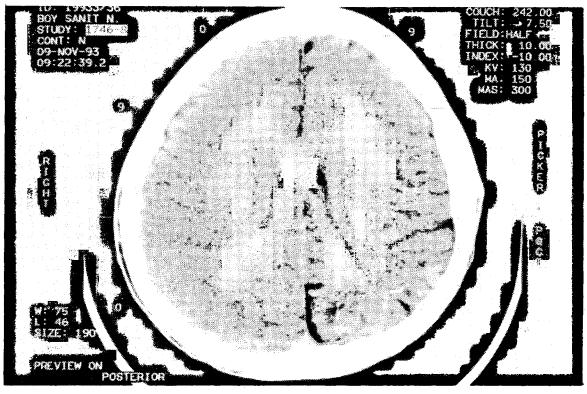


Fig. 3 Non contrast CT, the pathognomonic focal occipitoparietal gyral calcification with brain atrophy is seen ipsilateral to the left facial port wine stain.

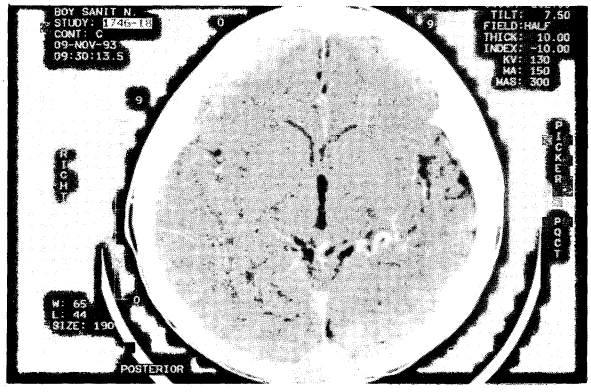


Fig. 4 Contrast enhanced scan, showing prominent collateral subependymal vein.

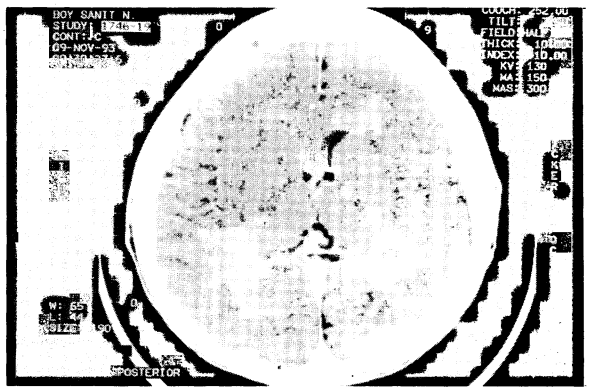


Fig. 5 Contrast enhanced CT, showing glomus of enhancing abnormal vessels in left trigone.

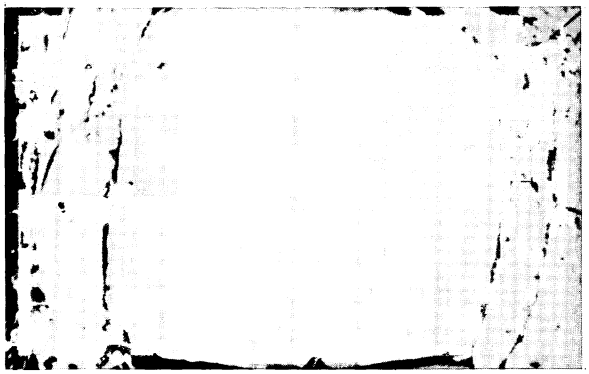
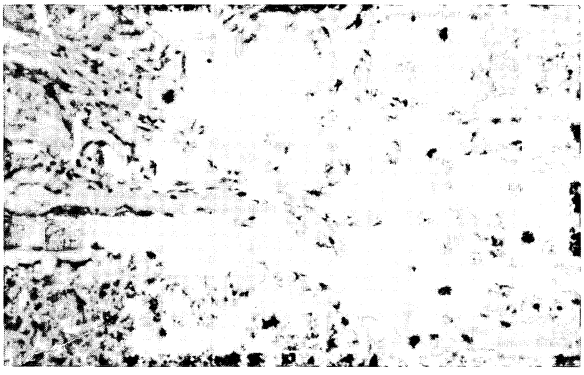


Fig 6. Intraoperative findings : left occipitoparietal leptomeningeal angiomatosis.



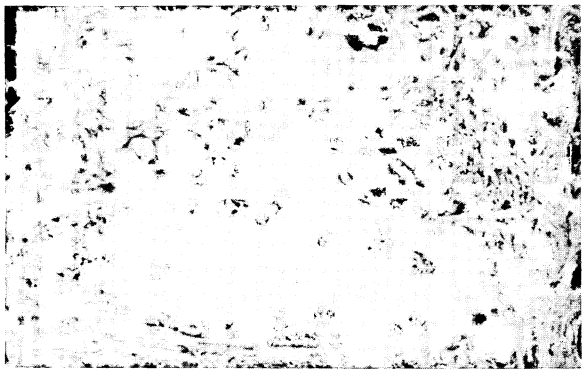


Fig. 7 Pathological specimen of abnormal vessels in the left trigone reveals cavernous hemangioma with calcospherules.

as secondary phenomena of dystrophic origin. Report of light microscopy and ultrastructural studies of Sturge - Weber syndrome suggested the role of vascular and other factors, such as anoxia, necrosis of cerebral tissue and calcium - ion concentration acts as secondary factors.[13]

Some observations can be made on the basics of this case about CT findings.

First gyral calcification in this patient was detected by CT only because it was below the capacity of conventional plain film.

Secondly, gyral calcification alone without characteristic facial port wine stain is non specific finding. It has been described in association with bacterial and viral intracranial infection, irradiation and intrathecal methotrexate for central nervous system leukemia,[3,4] chemical meningitis, secondary to subarachnoid fat from an epidermoid tumor,[15] tuberous sclerosis[16] and celiac disease.[17]

Thirdly, the characteristically Lt occipito - parietal leptomeningeal angiomatosis which was located ipsilateral to the facial angiomatosis was indirect evidence computed tomographically by the gyral calcification and absent to poor enhancement

after contrast enhancement.

The histopathology proved the presence of abnormal vessels in the ipsilateral choroid plexus, which had been earlier demonstrated by computed tomography.

This case we described on plain skull and CT findings is quite similar to other reports in appearance of classic Sturge - Weber syndrome,[11, 12, 18] only occasionally seen with typical manifestation.

Conclusion

Sturge - Weber syndrome is one entity in neurocutaneous syndrome. Although uncommon, it must be considered in patients with facial port wine stain which in the distribution of the opthalmic branch of the trigeminal nerve.

CT is the imaging of choice when this syndrome is suspected in older children and adult with negative conventional plain skull.

In addition, computerized tomography allows an earlier diagnosis since it shows different degrees of cerebral calcification together with intracranial angiomatosis and cerebral atrophy.

Reference

1. Oski FA. Principle and practice of pediatrics. Pennsylvania : J.B. Lippincott company, 1990 : 1946 - 47.
2. Porller AS. Vascular disorder. Dermatologic clinics 1987 ; 5(1) : 246-8.
3. Grossman CB. Magnetic resonance imaging and computed tomography of the head and spine. Baltimore : Williams and Wilkins, 1990 : 232-3.
4. Williams DW, Elster AD. Cranial CT and MRI in the Klippel - Trenaunay - Weber Syndrome. AJNR 1992 ; 13 : 291-4.
5. Benedikt - RA, Brown DC, Walker R, Chaed VN, Mitchell M, Geyer CA. Sturge-Weber syndrome : cranial MR imaging with Gd - DTPA. AJNR 1993 ; 14(2) : 409-15.
6. Marti BL, Menor F, Mulas F. The Sturge - Weber syndrome : correlation between the clinical status and radiological CT and MRI findings. Childs - Nerv - Syst 1993 ; 9(2) : 107-9.
7. Tournut P, Turjman F, Gubal AL, Revol M, Lapras C, Froment JC. MRI in Sturge - Weber Syndrome. J Neuroradiol 1992 ; 19(4) : 285-92.
8. Masenko JJ, Rosenbloom SA, Duchesneau PM, Lanzien CF, Weinstein MA. The Sturge - Weber syndrome : comparison of MR and CT characteristics. AJNR 1990 ; 11(1) : 131-4.
9. Sperner J, Schmauser I, Bitlner R, et al. MR imaging findings in children with Sturge - Weber syndrome. Neuropediatrics 1990 ; 21(3) : 146-52.
10. Siverman FN, Kuhn JP, comps. Caffey's pediatric X - ray diagnosis : an integrated imaging approach. 9 th ed. St Louis : Mosby, 1993.
11. Marti BL, Menor F, Poyatos C, COrtina H. Diagnosis of Sturge - Weber syndrome : comparison of the efficacy of CT and MR imaging in 14 cases. AJR 1992 ; 158(4) : 867-71.
12. Chamberlain MC, Press GA, Hesselink JR. MR imaging and CT in three cases of Sturge - Weber syndrome : prospective comparison AJNR 1989 ; 10 : 491-6.
13. Di TG, Di RC, Ablamondi AL, Caldarelli M, Pocchian M. Light microscopy and ultrastructural studies of Sturge - Weber disease. Childs Brain 1982 ; 9(1) : 23-36.

14. Ketonen L, Koskinicmi ML. Gyriform Calcification after herpes simplex virus encephalitis. J comput Asist Tomoge 1983, 7(6) : 1070-2.
15. Machen BC, Williums JP, Lum GB, Joslyn JN, Silverboard G. Intracranial gyriform Calcification associated with Subarachnoid fat. J comput Tomogr 1986, 10(4) : 385-8.
16. Wilm C, Van WE, Damaerel P, Smet MH, Plets C, Brucher JM. Gyriform calcifications in tuberous sclerosis simulating the appearance of Sturge - Weber syndrome disease. AJNR 1992 ; 13(1) : 295-7.
17. Gobbi C, Bougnet F, Creco L, et al. Coeliac disease, epilepsy and cerebral calcifications. Lancet 1992 ; 340(8817) : 439-43.
18. Paller AS. The Sturge - Weber syndrome. Pediatric Dermatology 1987 ; 4(4) : 300-4.

บริษัท ซี.เอส.กรุ๊ป จำกัด

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