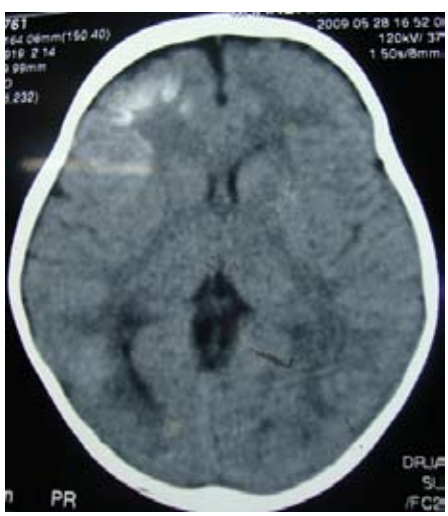
SPOT DIAGNOSIS (IMAGE GALLERY)



NEUROCUTANEOUS SYNDROME

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A one year old developmentally normal girl was brought with fever and left focal convulsions. Her blood counts, electrolytes, calcium, blood sugar, urea, creatinine, cerebrospinal fluid analysis were all normal. Detailed ophthalmological and fundus examination was normal. She had multiple episodes of convulsions in the hospital and was controlled with cabamazepine and phenytoin. Three hemangioma were seen over right upper eye lid, forehead and lateral side of skin over the nose. She had no motor deficits.

What is the diagnosis?

Sturge-Weber syndrome. It is characterized by a facial capillary hemangioma involving the periorbital area, forehead or scalp, a venous angioma of the leptomeninges and a choroidal angioma. The leptomeningeal angioma typically involves the parieto-occipital region or the entire hemisphere. Neuroradiological findings are leptomeningeal enhancement along with signs of cortical atrophy and calcifications. In our patient the CT brain shows cortical calcifications involving the right frontal and parietal lobes. Initially children will have normal development, later often experience neurological and developmental deterioration in association with the onset of seizures and consequently suffer a high rate of disability. Neither the size nor distribution of the facial haemangioma appears to correlate with the neurological features. The majority of children develop seizures in the first two years of life and with the onset of seizures below the age of six months being associated with intractability and a poor outcome. The current medical treatment is anticonvulsant treatment and anti-platelet treatment with prophylactic low dose aspirin. The rationale for the use of aspirin is that venous stasis may lead to thrombus formation.

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