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Tc-99m MIBI SPECT AND CT FINDINGS IN STURGE-WEBER SYNDROME WITH CLINICAL DETERIORATION: A CASE REPORT

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ABSTRACT

Introduction: The SWS, also called encephalo-trigeminal angiomatosis, is an extremely rare neurocutaneous disorder. The most frequent neurological manifestations include intractable seizures, focal deficits, headaches, and developmental disorders. Although the angioma progression is very rare, it may cause alteration of seizures type, frequency and extension.

Case presentation: We report a 25-year young male admitted to the hospital with increased frequency of focal seizures during the last six months. A large port-wine stain presented on the right forehead and upper eyelid of his face. On neurological examination he had mild left-sided facial palsy and hemiparesis. EEG found a focus of epileptic activity in the left temporal region. CT scans showed extended bilateral hyperdense lesions in the temporal and occipital regions. Tc-99m MIBI brain SPECT demonstrated lack of radioisotope uptake. Medical history revealed onset of generalized seizures at the age of six months. Primary CT scans and clinical findings were interpreted as cerebral angiomas with symptomatic epilepsy. After standard antiepileptic treatment a temporary seizures control was achieved.

Conclusion: We suggest that Tc-99m MIBI SPECT and CT neuroimaging may be useful for the detection and differential diagnosis of brain lesions in SWS presenting with clinical deterioration.

Key words: SWS, angiomas, calcifications, intractable seizures, brain SPECT, CT

INTRODUCTION

The SWS is a rare neurocutaneous disorder with leptomeningeal angiomas (Las) and cutaneous angiomas of the face, known as port-wine stains (5, 6, 9, 10). Evidences exist that SWS is caused by residual embryonal blood vessels which form the angioma of the leptomeninges, face, and ipsilateral eye (1, 3, 13, 14).

The neurologic manifestations depend on the location of the Las, predominantly in the parietal and occipital regions and their secondary effects on the surrounding brain tissue (3, 5, 9, 14). The most frequent symptoms include intractable seizures, focal deficits (hemiparesis and hemianopsia), headache, and developmental abnormalities (learning disability, developmental delay and mental retardation) (1, 3, 7, 8, 12, 13).

On structural neuroimaging (CT/MRI) unilateral angiomas are more common and usually present as static lesions (1, 9, 14, 15). The progressive angiomas and/or calcifications may cause change into focal seizures with secondary generalization and increase of their frequency and duration (2, 3, 6, 12). Although it is very rare, in patients with clinical deterioration both structural and functional neuroimaging are recommended (5, 7, 9, 13, 15). Recently, Tc-99m MIBI brain single photon emission computed tomography (SPECT) may be useful in detection and differential diagnosis of neoplastic and non-neoplastic cerebral lesions associated with intractable seizures (4, 11, 16).

CASE PRESENTATION

A 25-years young male was admitted to the hospital with increased frequency and duration of focal seizures with secondary generalization during the last six months. A large port-wine stain birthmark presented on the right forehead and upper eyelid of his face (Pic. 1). On neurological examination he had mild left-sided facial palsy and hemiparesis. EEG revealed a focus of epileptic activity in the left temporal region (fig. 1). CT scans showed bilateral contrast-enhancing hyperdense lesions in the temporal and occipital regions, corresponding to angioma progression and calcifications (fig. 2). Tc-99m MIBI brain SPECT demonstrated lack of radioisotope uptake (fig. 3). A symptomatic add-on therapy with levetiracetam was initiated and patient remained clinically stable for the next twelve months.

Medical history revealed onset of generalized seizures

at the age of six months. Later-on he presented with focal sensory-motor seizures. Primary brain CT scans and clinical findings were interpreted as cerebral angiomas with symptomatic epilepsy. The patient underwent treatment with standard antiepileptic drugs and temporary seizures control was achieved.

DISCUSSION

According to the previous observations, the typical patients with SWS present at birth with facial angiomas (1, 3, 7, 14). However, not all children with facial angiomas have central nervous system involvement (CNS) which raises certain diagnostic and prognostic concerns. In correspondence to Roach scale classification (6, 10, 13), we identify our patient as an example of complete SWS (Type I) with affected both facial and CNS areas.

Based on the literature review, the incidence of epilepsy in patients with SWS is 75-90% (1, 3, 8, 10, 12). About 75% had onset during the first year of life, 86% before age 2 years, and 95% before 5 years. They occur in about 71% of patients with unilateral and 87% with bilateral disease. Accordingly, our patient has an onset of focal seizures during the first year of his life in relation to the bilateral location of cerebral angiomas.

Recently, numerous studies suggest that seizures result from cortical irritability caused by cerebral angioma and/or calcifications, through mechanisms of hypoxia, ischemia, and gliosis (1, 3, 9). We suppose that the same factors contribute to epileptogenesis in our patient with bilateral angiomas and subjacent calcifications. Although the results from the previous studies are not equivocal (1, 5, 10, 12), we support the opinion that the early onset of seizures prior to age two and the bilateral angiomas with adjacent calcifications more often suggest development of refractory epilepsy.

Evidence exist that radiological imaging plays an important role in assessment of intracranial lesions (1, 2, 7, 9, 13, 15). Accordingly, CT scanning may show tram-track calcifications, cortical atrophy, enlarged choroid plexus, and contrast enhancement. Our CT findings (fig. 3) are equivocal with these previous observations.

Sometimes CT and MRI differential diagnosis is difficult, so a new functional technique SPECT is a promising alternative (4, 11, 16). In our patient a brain SPECT was performed after i.v. injection of 20 mCi Tc-99m MIBI. Although CT scans reveal progression of angiomas and calcifications, the lack of tracer uptake excludes malignant transformation of cerebral lesions (fig. 4).

CONCLUSION

Although the progression of angiomas and calcifications is uncommon in SWS, our findings note that in cases with clinical deterioration a neuroimaging is recommended. We suggest that Tc-99m MIBI brain SPECT and CT may be useful for the differential diagnosis and follow-up of progressive brain lesions associated with refractory seizures.

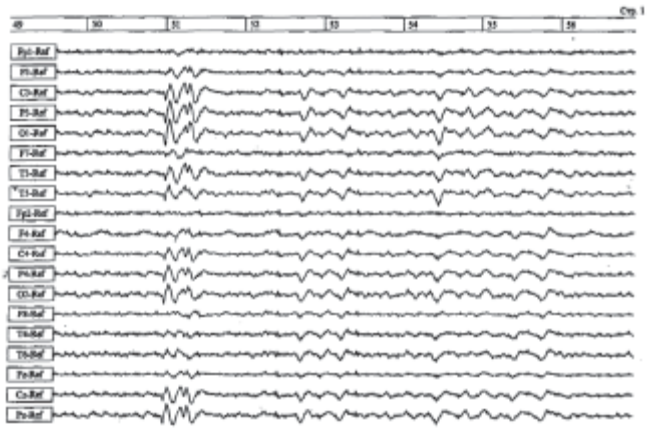

Fig. 1. EEG reveals a focus of epileptic activity in the left temporal region.

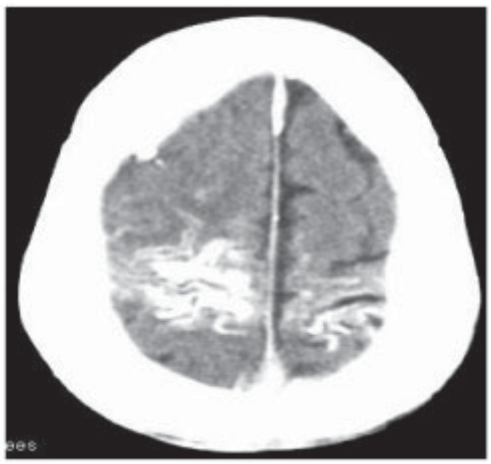

Fig. 2. CT scan shows bilateral hyperdense lesions in the temporal and occipital regions.

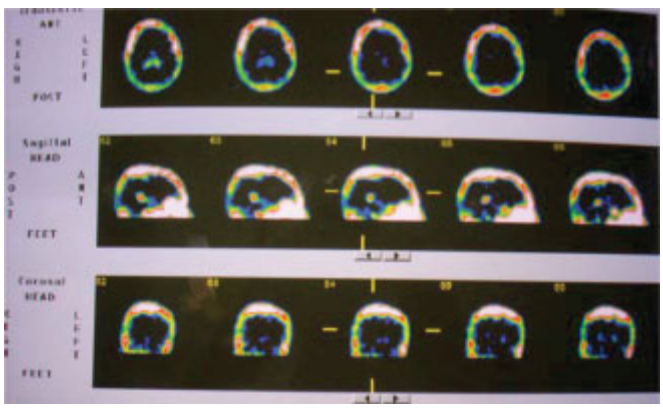

Fig. 3. Tc-99m MIBI brain SPECT demonstrates lack of radioisotope uptake.

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