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Case Report

Sturge-Weber Syndrome: About A Case

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Abstract: Sturge-Weber syndrome (SWS) or encephalo-facial angiomatosis, is a syndrome rare congenital neurocutaneous and ocular. He has two types of malformations: capillary congenital facial with flat angioma type and capillaro-venous lepto-meningeal location the more often ipsilateral parieto-occipital. The Neuroimaging, essentially imaging by magnetic resonance (MRI), plays a role important in establishing the diagnosis, ideally before the onset of complications neuro-ocular. We report the case of a patient with motor impairment in whom SWS is suspected based on facial angioma and pharmaco-resistant epilepsy.

Key words: Sturge-Weber, flat angioma,magnetic resonance imaging.

INTRODUCTION

Sturge-Weber syndrome (SWS) or encephalo-facial angiomatosis, is a syndrome rare congenital neurocutaneous and ocular. He has two types of malformations: capillary Congenital facial with flat angioma type and capillaro-venous lepto-meningeal location the more often ipsilateral parieto-occipital. The computed tomography and resonance imaging magnetic are considered the keydiagnosis of this syndrome.

OBSERVATION

We report the case of a 21-year-old male patient followed for drug-resistant epilepsy from an early age with cerebral palsy and who has been unconscious for 2 days.

On clinical examination, we have objectified a flat cutaneous angioma at the level left fronto-orbital. A cerebral CT scan was performed (Figure 1) showing vermicular calcifications and left parieto-occipital cortical-subcortical mound with thickening of the cranial vault opposite and hyperpneumatization of the frontal sinus and left mastoid cells. An MRI is also performed (Figure 2) out highlighting a left cerebral hemiatrophy, a thickening of the left choroid plexus as well as FLAIR hyperintensity and contrast enhancement a few left parieto-occipital cortical furrows associated with voids of gyriform signals on the T2* and SWI sequences. Thus the diagnosis of Sturge Weber syndrome was discussed, and the patient was referred for further care.

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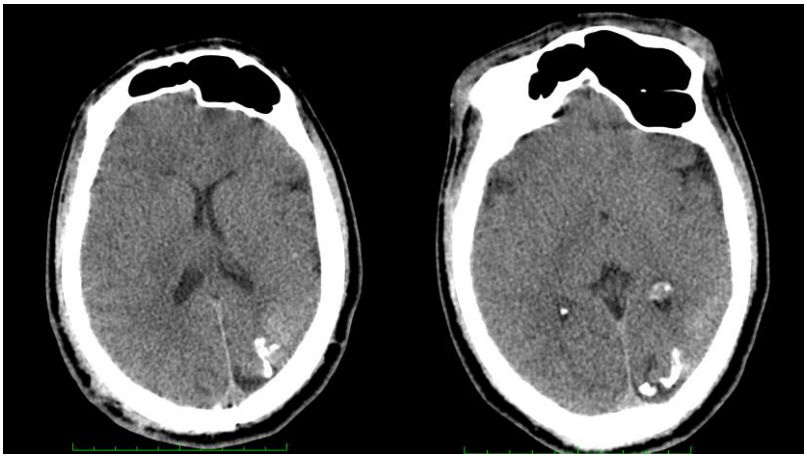

Figure 1: cerebral CT scan without injection of contrast product in axial section showing: left cerebral atrophy, left parieto-occipital calcifications, thickening of the choroid plexus with hyperpneumatization of the left frontal sinus

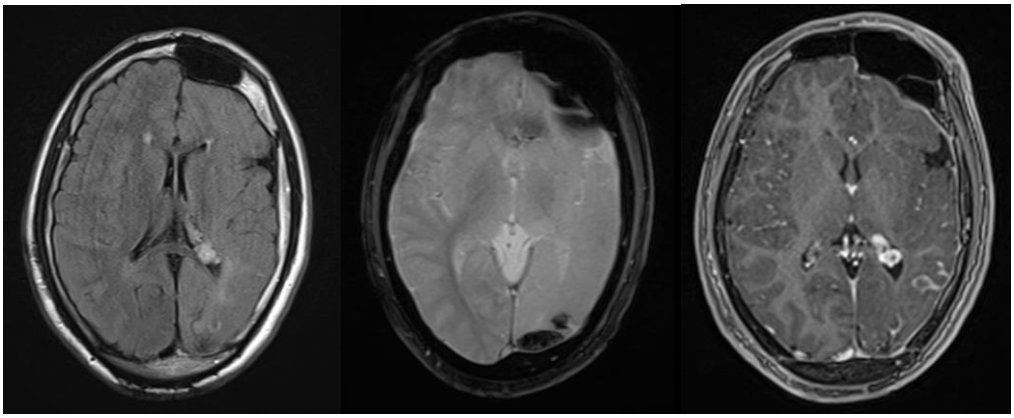

Figure 2: T2 FLAIR axial sequence: left parieto-occipital atrophy associated with a FLAIR hypersignal of a few left parieto-occipital cortical furrows axial sequence T2*: cortical-subcortical linear signal voids in relation to calcium deposits; spin echo T1 sequence after Gadolinium injection: thickened aspect of the left choroid plexus, and lepto-meningeal enhancement related to a pial angioma

DISCUSSION

Sturge-Weber syndrome (SWS) is a neurocutaneous and ocular phacomatosis, rare and sporadic, which results from a malformation of the fetal vasculature resulting in anoxia cortex [1]. In its complete form this syndrome features: a facial capillary malformation congenital type of flat angioma, an angioma capillaro-venous lepto-meningeal location the more often parieto-occipital, with atrophy cerebral and subcortical calcifications, and ocular abnormalities (glaucoma and disease choroidal vascular) [2, 3]. The anomaly fundamental physiopathology at the base of leptomeningeal angiomatosis is the failure of the normal development of the veins at the level of the cortex and the resulting persistence of the complex fetal primitive vascular disease [4]. During many years, this syndrome was defined by a distribution according to the territory of the nerve trigeminal. Recently, in 2014, Waelchli *et al*, proposed that the distribution of this syndrome follows the distribution of blood vessels of the face rather than that of the nerve trigeminal [5]. The presence of these vessels abnormal leads to impaired perfusion cerebral and progressive ischemia of the underlying brain parenchyma. It will still be aggravated in cases of seizures not controlled.

In 80% of cases, there is a violation unilateral cerebral hemispherical, which explains the high frequency of hemiatrophy brain in patients with SWS [6]. The clinical manifestations of the syndrome include refractory epilepsy to treatment, even hemiplegia, hemianopia and mental retardation.

Neuroimaging plays a crucial role in establishment of the diagnosis, ideally before the appearance of neuroophthalmological complications. MRI helps establish the diagnosis and to assess intracranial involvement, but there is no consensus on the people to screen, at the optimal time of imaging, the sensitivity of MRI or the advantage comprehensive identification of vascular abnormalities at risk. The presence of angiomatosis leptomeningeal has been considered as a direct characteristic (usually involving the ipsilateral posterior parietal or occipital cortex facial signs) [7]. MRI is the modality ideal imagery to search for this entity. The entire extent of the pial angioma is visualized under form of contrast enhancement in the subarachnoid spaces and facing gyrus. The atrophy of

the areas concerned characterizes the advanced cases. The echo-weighted sequence of gradient (T2*) is useful for visualizing deposits cortical-subcortical calcium. We can find an enlargement of the deep veins due to dysgenesis of the venous system superficial, causing a diversion of the blood. A thickening of the choroid plexus may also be found attributed to this bypass in the deep veins [8]. During of the first years of life, on the sequence of perfusion, this vascular malformation tends to becoming hypoperfused, by impaired drainage venous [9]. CT scans show calcifications in the subcortical white matter and in the adjacent gyriform cortex (tram track sign). The cause of these calcifications is attributed to chronic ischemia due to impaired venous drainage. A broad spectrum of malformations of cortical developments is observed in patients with SWS, ranging from polymicrogyria and schizencephaly to focal cortical dysplasia [10].

CONCLUSION

The diagnosis of Sturge-Weber syndrome is suspected in patients with angioma from the forehead. Faced with this suspicion, examination ophthalmology and an MRI with injection of gadolinium should be made to allow early diagnosis and reduce complications ophthalmological and cerebral.

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