ORBITAL CELLULITIS REVEALING STURGE-WEBER SYNDROME: CASE REPORT

Abstract:

Sturge-Weber syndrome (SWS), also known as encephalofacial angiomatosis, is a rare congenital neurocutaneous and ocular condition. It is characterized by two types of malformations: a congenital facial port-wine stain and a capillary-venous leptomeningeal angioma, typically homolaterally located, often in the parieto-occipital region. The diagnosis of SWS largely relies on neuroimaging, particularly magnetic resonance imaging (MRI), which is crucial for identifying anomalies before the onset of neuro-ocular complications. We present the case of a child in whom SWS is suspected due to the presence of a facial and leptomeningeal angioma.

INTRODUCTION :

Sturge-Weber syndrome, or encephalo-trijeminal angiomatosis, is a rare disease characterized by facial, leptomeningeal, and choroidal angiomas. Mainly diagnosed in patients with epileptic seizures, it affects both sexes with a slight male predominance. This condition is extremely rare globally and uncommon in Morocco, with only a few cases reported. MRI is the examination of choice for early detection and monitoring of this disease [1].

OBSERVATION :

We report the case of an 11-year-old female child from healthy non-consanguineous parents. Having a history of a congenital facial plane angioma of purplish color, good psychomotor development, notion of convulsive seizure at the age of 3 years placed on valproic acid stopped after 1 year by the parents and intellectual delay. The history of the illness dates back 4 days with the onset of unquantified fever associated with right eyelid swelling and purulent secretions complicated on the day of his admission by a partial convulsive attack of the right upper hemibody, giving way after injection of valium intrarectally, without coma or post-ictal deficit. A brain CT was performed showing orbital cellulitis with subcortical cortical atrophy and gyriform calcifications suggestive of Sturge Weber syndrome and a CRP of 195.3. The patient was put on IV antibiotic therapy with good clinical and biological improvement. The EEG showed the presence of a trace of global cerebral distress with the presence of bursts of slow waves and diffuse slow spikes predominant in bilateral temporo-occipital. A brain MR angiography was done secondarily and showed widening of the subarachnoid spaces and cortical sulci more marked on the left, Signal gyriform anomaly in SWI signal in right frontoparietal and left occipital regions, hypertrophy of the choroid plexuses, bilateral leptomeningeal enhancement and dilatation of the bilateral fronto-parietal cortical veins and more marked trans-parenchymal veins. at the right frontal level,Infiltration and thickening of the right eyelid soft tissues with

individualization of contiguous collections under periosteal fluid, with enhanced wall after injection of gadolinium, measuring 19x6mm and: 17x8mm extended over 22mm without bone lysis on the side, associated with infiltration of extra and intrafat conical with a swollen and strongly enhanced appearance after injection of Gadolinium of the superior rectus muscle, the whole is responsible for stage II exophthalmos,and bilateral pan sinusitis and mastoiditis. Furthermore, the ophthalmological examination was in favor of glaucoma, the abdominal ultrasound showed homogeneous hepatomegaly and ETT was normal. The diagnosis of Sturge Weber Syndrome was made, the patient was placed on antiepileptic medication based on Carbamazepine and beta blocking eye drops with good clinical progress and regular ophthalmological monitoring.

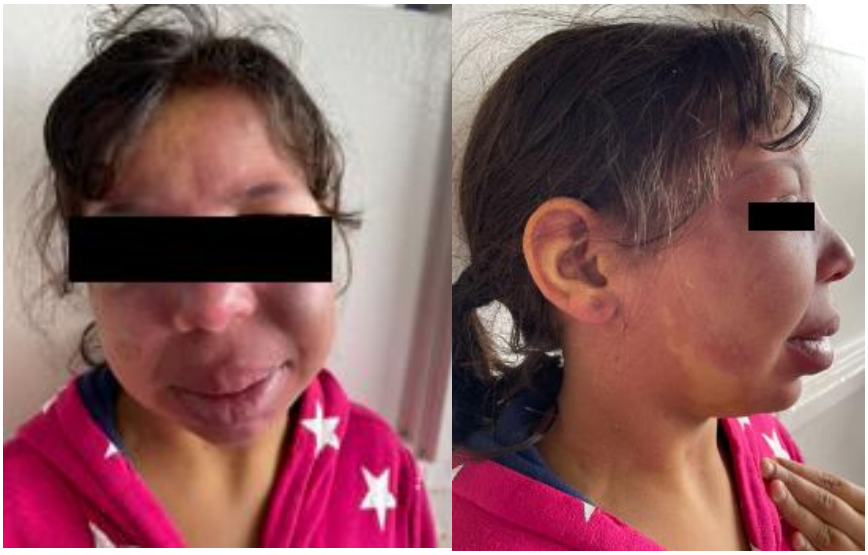

Figure1: plane angioma of the face

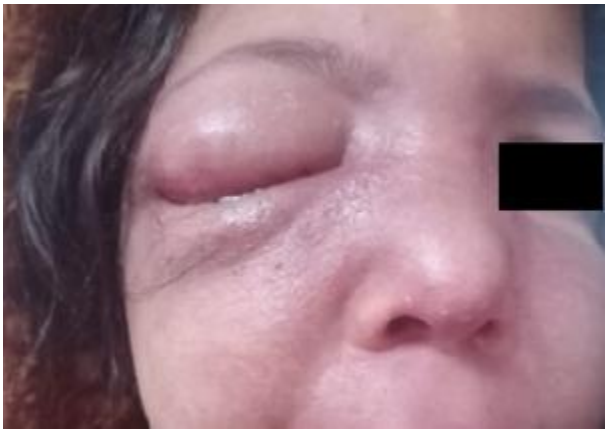

Figure2: right pre-septal cellulite

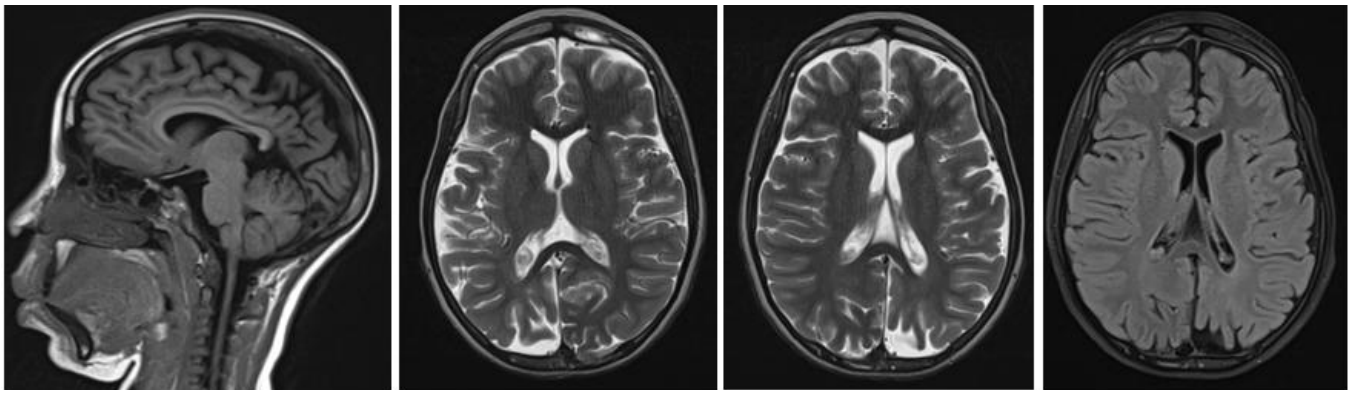

Figure 3: The cerebral MRI angiogram revealed widening of the subarachnoid spaces and cortical sulci, as well as bilateral leptomeningeal enhancement.

DISCUSSION :

Sturge-Weber syndrome takes its name from two doctors who contributed to its initial description: William Allen Sturge and Frederick Parkes Weber. In 1879, Sturge first described the syndrome in a patient with facial angioma, glaucoma, and seizures. Later, in 1922, Weber reported brain calcifications in a patient with similar symptoms, which helped to better define the clinical features of the syndrome [2].Heis a serious neurocutaneous condition characterized by a facial plane angioma affecting the first branch of the trigeminus (V1), neurological abnormalities such as a leptomeningeal angioma ipsilateral to the cutaneous angioma, and variable ocular abnormalities [3]. Its incidence is estimated at approximately 1 in 20,000 to 50,000 births.[4] Encephalofacial angiomatosis has been divided into three types based on the variable distribution of the angioma, including isolated facial involvement versus associated leptomeningeal involvement. The Roach scale was used to classify encephalofacial angiomatosis into 3 types: [5]

- Type I: Presence of facial and leptomeningeal angiomas; may be associated with glaucoma (classic Sturge-Weber syndrome).
- Type II: Presence of facial angioma only (no CNS involvement); may be associated with glaucoma.
- Type III: Presence of isolated leptomeningeal angioma in the brain; usually without glaucoma.

The main neurological manifestations are dominated by epilepsy, affecting between 75 and 90% of patients, generally early and severe. These attacks are most often partial to the contralateral hemibody, observed in approximately 70% of cases. Around 50% of patients also have a motor deficit. This deficit can develop gradually or be caused by repetitive cerebrovascular accidents (strokes) (hemiparesis, hemianopia and hemiatrophy) associated with convulsions, headaches or migraine. Although psychiatric disorders and mental retardation have been reported, they remain rare. [1,6] Our case belongs to Sturge Weber syndrome type 1 according to Roach. The

predominant symptom observed in our patient is the presence of convulsive seizures, associated with mild mental retardation.
The facial plane angioma is usually located in the V1 trigeminal region and sometimes extends to the V2 and V3 territories. It is usually observed unilaterally, but can also be bilateral. It frequently affects the parieto-occipital region, although it can also be present hemispherically. [6]
For ophthalmological abnormalities associated with Sturge Weber syndrome include glaucoma, the frequency of which varies from 30% to 70%, the age of onset presents three peaks: during the first year (40%), between 5 and 9 years (23%) and after the age of 20 (20%). In addition, choroidal hemangioma is observed in 40 to 50% of affected patients. [4.7]
Computed tomography and magnetic resonance imaging are essential for the diagnosis of this syndrome. Brain CT scan aims to identify several characteristics including signs of brain atrophy, either focally or hemispherically, often ipsilateral to the angioma. It also examines the presence of "S"-shaped, gyriform or train-rail intracranial calcifications, which are generally located subcortically at the level of the meningeal arteries and cortical veins. In addition, she looks for hypertrophies and calcifications of the choroid plexus ipsilateral to the angioma, gyriform cortical enhancement.Magnetic resonance imaging (MRI) offers greater sensitivity than CT scanning. It makes it possible to detect early signs, sometimes even before the appearance of clinical symptoms. In addition to the elements looked for by CT scanning, MRI can highlight additional features. This includes visualization of pial angioma, for which specific magnetic resonance angiography (MRA) sequences may be particularly useful. In addition, MRI makes it possible to detect choroid plexus angioma, venous development anomalies, signs of cerebral atrophy, cerebral calcifications which appear as hypo signal on all sequences, as well as cortical malformations such as polymicrogyria, lissencephaly or localized pachygyria. [1,6,8,11,12]

The electroencephalogram (EEG) provides a non-invasive method for assessing brain function that can be repeated safely and as often as necessary. The most common electroencephalographic abnormality is an asymmetry, characterized by a decrease in amplitude and slowing of background activity in one or both hemispheres related to cerebral distress, as confirmed in our case. This asymmetry can be observed from the first months of life, but it becomes more marked with the progression of hemisphere atrophy. Focal discharges occur primarily in the affected cerebral hemisphere, but may also be present in the contralateral hemisphere in many cases. EEG is also extremely useful in differentiating migraines and stroke-like events from seizures as causes of acute paroxysmal events.[14,15]
Treatment of Sturge-Weber syndrome is symptomatic, with the use of antiepileptic drugs to control seizures. Low doses of aspirin may be prescribed to prevent strokes and reduce brain atrophy. In cases of refractory seizures, hemispherectomy is considered. Congenital glaucoma is managed by surgical and medical approaches, requiring regular ophthalmological monitoring. Physiotherapy may be recommended for motor deficits.[4,7,9,10,13,16]

CONCLUSION :

Sturge Weber syndrome remains a very rare disease which must be considered in front of a facial angioma in order to start antiepileptic treatment early and look for ophthalmological complications The coexistence of bilateral facial port-wine stains and cerebral developmental venous anomalies heightens the epilepsy risk in patients with Sturge-Weber syndrome.[1,2,7,17]

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