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RESEARCH ARTICLE

PRES REVEALING A TAKAYASU ARTERITIS IN THE EMERGENCY DEPARTMENT : A CASE REPORT

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Abstract

Posterior reversible encephalopathy syndrome is a rare clinical and radiological entity which can lead to seizures. Some of its risk factors are hypertension and vasculitis, as in the case of Takayasu Arteritis, a large vessel arteritis which damages the abdominal vasculature. We report the case of a patient who presented in a generalized convulsive status epilepticus secondary to a PRES, which was induced by a renovascular hypertension secondary to Takayasu Arteritis. The aim of this study is to report the case of a rare association of Takayasu Arteritis and PRES, and discuss the timeline of the assessment of our patient's condition, which allowed us to start the treatment as quickly as possible.

Introduction:-

Generalized convulsive status epilepticus have a lot of known etiologies, which has a major influence on vital prognosis and functional outcome [1]. Therefore, it is vital to identify the etiology, and treat it to avoid recurrence of seizures.

We report the case of a 19 years old female patient who presented with a generalized convulsive status epilepticus, which revealed a Takayasu Disease after a rigorous exploration, including the assessment of a secondary hypertension which led to a Posterior reversible encephalopathy syndrome.

Case Report

Patient information :

We report the case of a 19 years old female patient, with no past medical history of seizures, who was brought to the hospital after her dad noticed a rhythmic involuntary movement of the patient's limbs for at least 10 minutes, followed by the onset of unconsciousness. Prior to the onset of seizures, the patient's main complaint was a headache and decreased visual acuity.

Clinical Findings :

In the emergency room, the patient was unconscious with a Glasgow Coma Scale at 3, with blood in her mouth, a normal capillary blood glucose, hypertension at 222/106 with a mean arterial pressure of 144, and a bruit on auscultation of the abdominal aorta. Two additional convulsions were observed, the last stopping at the administration of two doses of midazolam 5 minutes apart. The patient progressively regained consciousness after 45 minutes.

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Diagnostic assessment :
After ruling out the presence of a bladder globe and hypercapnia, we performed a head CT-Scan without injection of contrast product, which came back normal.

Complete blood count, lumbar puncture and metabolic blood tests came back normal, except for hypoalbuminemia (21 g/L) and hypoproteinemia (50.97 g/L) with no disturbance of GFR (123.7 ml/mn). B-HCG was negative. Erythrocyte sedimentation rate was high (105 mm at the first hour) with raised C reactive protein (87.9 mg/L). An MRI is then performed with results in favor of a posterior reversible encephalopathy syndrome.

The cause of the hypertension is then sought. A doppler ultrasound of the renal arteries is performed, and finds a stenosis of the left renal artery with a small left kidney. An abdominal CT angiography is then carried out and finds a stenosis of the upper abdominal aorta and confirms the stenosis of the left renal artery (Image 1).

Given the presence of three criterias of the 1990 American College of Rheumatology Classification criteria of Takayasu arteritis, we suspected this disease to be the origin of our patient's hypertension leading to her posterior reversible encephalopathy syndrome.

Therapeutic intervention :
Hypertension was managed with a titration of nicardipine which allowed us to reduce blood pressure to 178/91 with a mean arterial pressure of 126 after an hour. We continued anticonvulsant treatment with valproic acid and phenobarbital. We started oral corticosteroids at the dose of 1 mg/kg.

Follow up and outcomes :
We transferred the patient to the internal medicine service, once her neurological state was stable. There, gradual tapering of oral glucocorticoids is started, without seizure recurrence. A pre therapeutic assessment is started to assess the possibility of introducing maintenance therapy. An electroencephalogram realized one month later shows no anomaly.

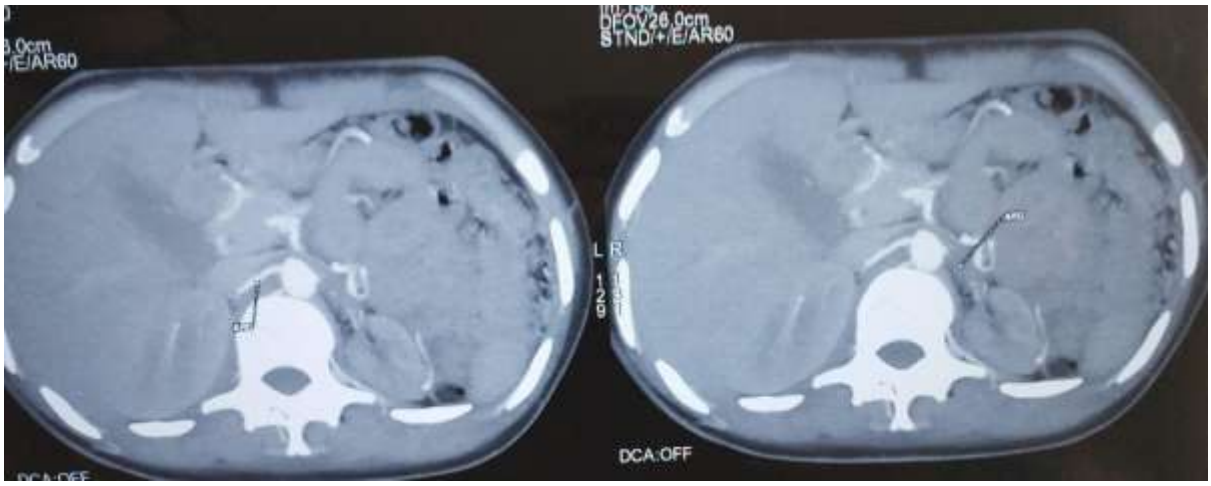

Figure 1:- Abdominal CT angiography showing the stenosis of the left renal artery.

Discussion:-

Status epilepticus is an emergency, defined by its duration. Its prognosis is linked to the underlying etiology and the quickness and effectiveness of management [2] which relies on multiple drugs, such as midazolam in the first line, and phenobarbital and valproic acid in the second line [3]. Management also should include the active search for etiology.

In non epileptic patients, cerebrovascular diseases are a major etiology. Status epilepticus complicates only 5 to 15% of Posterior reversible encephalopathy syndrome (PRES) cases [4].

PRES, also called reversible posterior leukoencephalopathy syndrome, hyperperfusion encephalopathy or brain

capillary leak syndrome [5], is a clinico radiological entity with acute neurological symptoms [6]. These include Headache, visual changes, seizures and altered mental status [7]. Its risk factors include hypertension, chronic kidney disease and or/failure, exposure to immunosuppressive medications, vasculitis and rheumatoid arthritis [7] [8].

Its diagnosis relies on MRI, but CT findings are described for this disease ([7]. Typical MRI findings are bilateral lesions, with parieto-occipital or posterior frontal cortical-subcortical FLAIR hyperintensities. Also high intensity signals are seen on the apparent diffusion coefficient maps.
Management of PRES relies on the treatment of the underlying cause. In patients with acute hypertension, blood pressure should be reduced progressively in order to avoid cerebral coronary and renal ischemia [8].

Onset of hypertension at an age younger than 30 years old and abrupt onset of hypertension are indications for screening for secondary hypertension. Moreover, hypertension of abrupt onset with abdominal systolic diastolic bruit on auscultation are indications for screening for a renovascular disease [9]. This is performed with a renal doppler ultrasound, with confirmation of ambiguous results with CT scan or MRI [10]. Renovascular hypertension secondary to arterial narrowing can develop in Takayasu arteritis [11] [12].

Takayasu arteritis is a large vessel arteritis, with a predominance of the disease in the abdominal vasculature, bilateral disease of the subclavian and carotid arteries, or focal disease limited to the left subclavian artery [13]. Takayasu arteritis affects women predominantly, with a mean age of diagnosis of 25-30 years old [11] [14]. Diagnostic criteria relies on a combination of clinical, biological and radiological findings. We can cite The 1990 American college of rheumatology classification criteria for takayasu arteritis (table 1 [15]) for diagnosis.

The American College of rheumatology/vasculitis foundation issued new guidelines in 2021. Patients with active takayasu arteritis should receive high dose oral glucocorticoids, in association with a nonglucocorticoid immunosuppressive agent [16].

However, we remind our lectors that immunosuppressive agents are a risk factor for the PRES [8].

Criteria	**Definition**
Age at disease onset < 40 years	Development of symptoms or findings related to TA age < 40 years.
Claudication of extremities	Development and worsening of fatigue and discomfort while in use especially the upper extremities.
Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries.
Blood pressure difference > 10 mmHg	Difference of > 10 mmHg in systolic blood pressure between arms.
Bruit over subclavian arteries or aorta	Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta.
Arteriogram abnormality	Arteriographic narrowing or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia or similar causes; changes usually focal or segmental.

Table 1:- The 1990 American college of rheumatology classification criteria for takayasu arteritis.

In this case, our patient presented with a generalized convulsive status epilepticus and hypertension. We ruled out the metabolic and infectious cause, and performed an MRI which allowed us to find a PRES.

Considering the age, the presence of bruit on auscultation of the abdominal aorta, and the brutal onset of hypertension, we suspected a renovascular hypertension, confirmed by a renal doppler ultrasound.

Given the presence of three criterias out of six of the ACR 1990 criteria for the classification for Takayasu arteritis, we started oral high dose glucocorticoids after specialist consultation with internists.

Conclusion:-

In our case report, a PRES revealed a Takayasu Arteritis. Following the guidelines for the management of PRES helped us to identify hypertension as the first clear cause of this syndrome.

Controlling our blood pressure and identifying the underlying disease behind the secondary hypertension allowed us to start the adequate treatment.

PRES syndrome and takayasu arteritis is a possible association which can lead to the worst complications such as general convulsive status epilepticus.

Abbreviations

ACR : American college of rheumatology.
B-HCG : Human chorionic gonadotropin.
CT-Scan : Computed tomography scan.
GFR : Glomerular filtration rate
MRI : Magnetic resonance imagery
PRES : Posterior reversible encephalopathy syndrome.
TA : Takayasu arteritis.

Data availability:
Not applicable

Author contributions
HEH : Study concept, data collection, data analysis, writing the paper. IC : Writing the study, data collection , data analysis. MEA : Data collection. AB : Data collection. ZEH : Data collection, critical review. MA : Supervision and data validation.. All authors read and approved the final manuscript.

List of tables and figures :
Table 1 : The 1990 American college of rheumatology classification criteria for takayasu arteritis
Figure 1 : Abdominal CT angiography showing the stenosis of the left renal artery.

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Consent for publication
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editor in chief of this journal on request.

Competing interests
Nothing to declare.

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