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Case Report

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Ntenga Syndrome: About an Observation

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Abstract

Ntenga syndrome, is one of the highly epileptogenic, non-metabolic craniopathy whose aetiology is not yet known. This syndrome makes a differential diagnosis with that of Morgagni-Stewart-Morel which is rare and / or rarely mentioned in current clinical practice (entity made of frontal hyperostosis, neuropsychiatric and endocrine disorders). We report here a 58 years old female patient from Lubumbashi/ Democratic republic of Congo, followed for several years for multiform seizures, in whom the explorations of a status epilepticus, made possible to set up a new syndromic entity, called Ntenga syndrome made of a symptomatic triad (persistent multiform epileptic seizures, absence of endocrine disorder, hyperostosis frontalis interna). To date, a therapeutic protocol made of valproic acid and levetiracetam has significantly reduced to one seizure per month or even every 2 months. I think it is not without interest to report a very rare and / or new entity in the clinic.

Keywords: Ntenga Syndrome, Persistent Multiform Epileptic Seizures, Absence of Endocrine Disorders, Hyperostosis Frontalis Interna.

Introduction

Science in its diversity, with its dynamic aspect, several pathologies and / or syndromes are not necessarily the same as those described centuries ago by eminent scientists. There are more patients than pathologies that means here, according to the North South gradients, particularities are numerous. Clinicians must seek, on a case-by-case basis, the specificity of the picture so as not to fall into routine and repetition, which repetition could slow down the evolution of science. Here we describe a syndrome called Ntenga Syndrome which has both similarities and differences from Morgani morel syndrome.

Case Presentation

We report a 58 years old patient, divorcee (due to repetitive seizures), who had been followed up since 2010 at the Joseph Guislain Neuropsychiatric Centre (CNPJG) Brothers of charity of Lubumbashi in the Democratic Republic of Congo, the main presenting complaints were headache, a psychiatric picture summarized in a behavioural disorder (agitation, aggression, logorrhoea), multiform convulsive seizures (partial motor, generalized tonic-clonic, with no cognitive disorders). This picture led the physician to put her on levetiracetam, urbanil and then haloperidol tablet. This treatment was subject to modifications with other antiepileptic drugs following the persistence and variability of seizures. No significant past history was noted in this patient. As of 24.12.2018, she was readmitted in the picture of status epilepticus for which she was treated until complete recovery and laboratory investigations were unremarkable. This led us to request a CT scan brain which was not done because of lack of funds until one year later in 2019 and which will highlight an enlarged inner table frontal bone sparing the outer table and the diploe. From the sum of all these features, Ntenga Syndrome was evoked as a new syndrome which makes differential diagnosis with the disease of Morgani Morel. Up to now she is on a treatment based on valproic acid (2 x 500 mg / day) and levetiracetam (1000 mg / day), with this

protocol, we have reduced to a frequency of one episode per month or even every 2 months, with a post-critical EEG which does not present paroxysmal graphic elements.

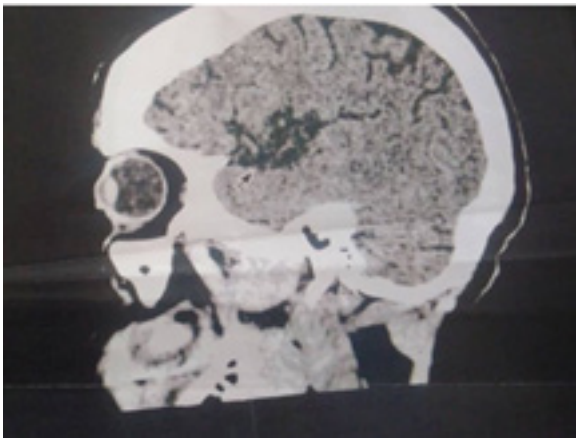

Figure 1 : CT scan brain showing an enlarged frontal bone

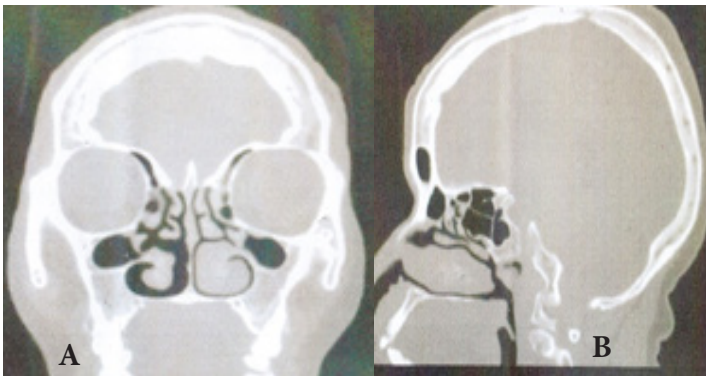

Figure 2 : Bone Windows A (coronal) B (sagittal) showing hyperostosis of the inner table of frontal bone

Discussion

In terms of differential diagnosis, Morgagni-Stewart-Morel syndrome is a pathology formerly described around the middle of 19th century in which the author described a triad made of internal frontal hyperostosis, neuropsychiatric disorder and adiposis for mature patients who suffered from delusions of persecution with a genetic connotation without any genetic study to attest, of pituitary-diencephalic disorders, having been the subject of a proposal for hormonal treatment [1].

Very recently, Salma Louzi et al, 2019 in their study reported an observation of a 62-year-old patient who had seizures for several months, behavioural problems and progressive cognitive deterioration; with following risk factors: obesity, total hysterectomy, metabolic disorders (dyslipidaemia and hyperuricemia) and hypothyroidism, imaging done (CT and MRI) showed bilateral frontal hyperostosis of the internal table, sparing diploe and the external table [2].

Particularity and similarity of Ntenga Syndrome with Morgagni-Stewart-Morel syndrome.

Our patient was 58 years old, with persistent multiform seizures (in her evolution we noted partial motor crises, generalized tonic clonic crises, a status epilepticus, the later was not reported by previous authors), neuropsychiatric disorders and an enlarged frontal bone,

no endocrine disorders noted. These features spanned several years.

Reading these observations shows us master and constant ideas of the pathology including primo, an evolution which spreads over long period of time and therefore possibility of not necessarily knowing the diagnosis with a possibility of diagnostic wandering, secondly an hyperostosis (hypertrophy) of the frontal plate which can be bilateral or unilateral according to the patient, neurological disorders and / or psychiatric of any kind because this could be related to the individual mental constitution.

The question remains as to the origin of these epileptic crises, is it linked to the modification of the neuronal environment (frontal hyperostosis?) But if this is the case, in my opinion there would be on the frontal type of seizures. Otherwise, we should also know why they present with multiform seizures.

Our patient is in middle age (58 years) as the other authors point out. If the frontal bony hypertrophy is progressive, that means middle age is a negative risk factor, because in my opinion, the onset of the disease is not known. The insidious onset characterizes this syndrome.

The elements initially described in the triad of Ntenga syndrome are 1. Hyperostosis frontalis interna, 2. Persistent Multiform Epileptic Seizures in the foreground, 3. Absence of endocrine disorders.

We point out here that it is by investigating the aetiology of status epilepticus in our patient that we have managed to enrich our triad. And so, it is a question that an extended investigation is systematically made in patients with multiform crises without endocrine disorder objectified so as not to miss the diagnosis, and this regardless of the age of the patient.

In addition, If the clinical and radiological correlation helps the diagnosis for many of certain pathologies in neurology and other disciplines of medicine, it is also true to affirm on one hand, the rarity or the virtual absence of imaging (CT scan, MRI), specialists (neurologists, neurosurgeons, psychiatrists), on the other hand the cost of these explorations even if available, in many regions, particularly in Africa. Therefore, it seems logical to us to question this assertion which crosses the centuries on the rarity of certain pathologies long qualified as rare, because we can only think about it in our opinion the person who is informed about it. This hypothesis is supported by the number of years passed (from 2010 to 2020) to make the diagnosis in our patient.

The headaches reported in our patient, were also reported by C. Paris et al, 2017, in a 59-year-old woman consulted because of chronic headaches [3].

Conclusion

For persistent multiform epileptic seizures (variants) without proven endocrine disorder, with or without associated psychiatric pathology, a brain scan must be done systematically in any patient regardless of age to look for a bony enlargement of the frontal bone, and

therefore make the diagnosis of Ntenga Syndrome. Dual therapy with anticonvulsants must be started immediately aiming to reduce the number of attacks that turn the patient into a status of anxiety.

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