Psychogenic non-epileptic psychogenic seizure (PNES): a diagnostic wandering

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I. Introduction

The psychogenic non-epileptic seizures (PNES) are behavioral or motor manifestations not having an epileptic origin, defined as sudden and paroxysmal changes in motor, cognitive, emotional, sensation or consciousness behavior, for a limited period of time, which primarily evoke seizures, but which are related to a psychogenic origin and not to an excessive neuronal discharge (no concomitant critical electrophysiological activity) [4].

A clear predominance of women is reported (75%) in most studies [17]. The majority of these patients have a history of trauma or abuse [10].

The PNES is a frequent, expensive and serious pathology. This is a poorly understood pathology, diagnosed late with an average diagnostic delay of between 5 and 7 years [20]. This difficulty in recognizing them is partly explained by the semiological proximity between PNES and epileptic crises and their possible entanglement. The PNES are associated with non-psychiatric and psychiatric comorbidities [15].

The PNES require close collaboration between neurologists and psychiatrists for diagnosis, follow-up and therapeutic management. These are often disabled by seizures, but also by associated comorbidities, see exposed to the side effects of antiepileptic drugs which are ineffective on this pathology.

II. Définitions

We must distinguish the term non-epileptic seizure from the PNES. The non-epileptic seizure designates any paroxysmal manifestation which can be mistakenly confused with an epileptic seizure without prejudging its etiology (organic or psychogenic).

The PNES are defined as “repetitive paroxysmal manifestations that at first glance evoke epileptic seizures, but related to unconscious psychogenic processes and not to excessive neuronal discharge” [14]. The term "pseudo-crisis" should be avoided because it can mean a simulation by the patient.

The DSM-IV-TR hoards PNES at the level of somatoform disorders in the category of conversion disorders. Conversely, the ICD-10 refers to them within dissociative disorders.

III. Epidemiology

Its incidence in the general population is estimated by a recent study at 4.9 / 100,000 / year [8]. A quarter of the patients seen by an epileptologist suffer from NEB. The PNES concern 10 to 50% of adult patients consulting a specialized epilepsy center and are the cause of 20 to 30% of "seizure" resistant to anticonvulsant therapy [9].

The PNES are comorbid in authentic epileptic seizures in 20 to 40% of cases. The preponderance of women is mentioned by several authors in favor of a sex ratio of 1/7, with an onset of the disease which is generally in the second or third decade (between 20 and 30 years) [1]. They are rare before 10 years, but can be seen in the elderly.

IV. Etiopathogenic hypotheses

The etiopathogenesis is multifactorial, including predisposing factors, triggering factors and maintenance factors.

Two etiopathogenic mechanisms are discussed: the role of mental trauma as an inducer of dissociative disorder, and / or a neurobiological predisposition.

A coexistence of PNES with post-traumatic stress disorder, on the one hand [5], and dissociative disorders, on the other hand [5], is found. Psychological trauma is implicated, often resulting from child abuse and in particular from sexual violence. The traumatic report of a serious somatic pathology for oneself or those close to you can also trigger the onset of PNES.
Several studies suggest that neurobiological elements could influence the onset of PNES. Without undoubtedly arguing for organicity, the onset of PNES may be secondary to neurosurgical operations related to interventional treatment of epilepsy or unrelated to refractory epilepsy [21]. Neurobiological predisposition may play a role in the onset of PNES.

Recent models place emotional dysregulation at the center of the problem. Functional imagery has revealed functional anomalies between the brain areas involved in emotions and the motor areas.

V. Diagnostic Approach

1- The neurologist’s approach:

Neurologists are often confronted with this situation, since several of these cases are referred for drug-resistant epilepsy. The history is a good sign of orientation. The clinical semiology of seizures obtained by the witnesses and ideally recorded on video coupled with the analysis of the EEG per critical allows the diagnosis to be made with certainty.

In routine clinical practice, the diagnosis of PNES is most often evoked before an epileptic syndromic presentation with drug-resistant motor attacks. But the doctor rarely observes the crisis himself, the description of which is reported by those around him.

a- Patient specifics:

The PNES often affect young adults and women (75%). Epilepsy is frequently associated (5 to 30%), a history of minor head injuries (20 to 30%), and a history of learning disabilities (10%) are reported [4]. Non-neurological comorbidities have also been reported (asthma, hypertension, peptic ulcer, gastroesophageal reflux, obesity) [6].

Repetitive admissions to the emergency reception service are noted (an immediately high frequency of crises). The occurrence of PNES during additional examinations or a gesture (anesthesia or surgery) is possible.

b- Description of the patient:

These patients imprecisely describe the possible subjective symptoms, with less detail, most often an incomplete answer to the questions and formulated in a negative way “I don’t remember anything, I don’t know what happened” [19].

c- Critical semiology:

The semiological elements observed in epileptic seizures such as the opening of the eyes, the presence of an injury, the loss of urine and the bite of the tongue, may be present in the PNES.

Critical movements are often more extensive and less well systematized than in epileptic seizures, they rarely alternate a linear sequence of symptoms following a precise neurological somatotopia. Finally, there is most often the absence of post-critical confusion in the PNES.

Certain clinical signs of seizures are suggestive of PNES: a long duration, a fluctuating evolution of signs during the same crisis, the asynchronous nature of the movements when they are bilateral, the movements of flexion and extension of the pelvis, the movements denial of the head, closing of the eyes, critical crying, taking posture in opisthotonos. However, none of these signs taken in isolation can confirm with certainty the diagnosis of PNES.

Contrary to popular belief, the semiology of PNES is not completely anarchic. There are five clinical subtypes [13]:

- **Brief hyperkinetic with automatic gestural activity of an emotional nature**: lasting less than 5 minutes, dystonic postures are frequent, with motor behavior with a strong emotional tone (anger, fear).

- **Prolonged hyperkinetic with axial implication**: progressive onset with an extended duration (> 5 min), there are flexion / extension movements of the trunk or even a posture in opisthotonos, toned postures of the limbs. An aura and hyperventilation are possible. The evolution is fluctuating.

- **Prolonged hyperkinetic without axial manifestation with hyperventilation**: progressive onset with an extended duration (> 5 min). Absence of axial manifestations, the motor signs of the limbs are varied and fluctuating (dystonic, tremors, clones, etc.), auras are possible, and hyperventilation is frequent. The evolution is fluctuating.

- **Paucikineti**c **with preserved contact**: progressive onset with variable duration, contact is preserved for most of the crisis, the motor signs are often focal, rare or discreet (example: fine distal tremor), immobility of the axis, and an aura is possible.

- "pseudo-syncope" or "dialeptic": sudden onset with a duration of less than 5 minutes, there is an impairment of contact (often eyes closed), motor signs such as clonies, tremors, myoclonus, movements possible denial, and hyperventilation is possible.
d- The suggestion:
The PNES can be evoked by provocative maneuvers during video-EEG such as hyperpnea, intermittent light stimulation, reinforced by patient information that these maneuvers can cause epileptic seizures.

2- the psychiatrist's approach:
The psychiatrist has his place in the diagnostic process to support or not the diagnosis of PNES mentioned by the neurologist, by looking for the different factors of predisposing, precipitating and perpetuating.

a- Predisposing factors (vulnerability):
In patients with PNES, exposure to a traumatic event (physical violence, sexual assault, etc.) is often found. Neurocognitive factors are also found (learning difficulties, low intellectual level, head trauma, epilepsy). Psychiatric comorbidities are frequent (70–95%) [4], such as personality disorders, emotional disturbances which may consist of an alexithymia (difficulty in identifying and verbalizing one's emotions), a strong dissociative tendency, even a disturbance of the autonomic nervous system. The Patients with PNES most often have dissociative or somatoform disorder, but also have a thymic or anxiety disorder [5].

A history of head trauma is observed in almost 30% of subjects [24]. The presence of comorbid epilepsy also represents a vulnerability factor in 20 to 30% of subjects. A study has shown an association between PNES and a number of somatoform syndromes (fibromyalgia, irritable bowel syndrome), chronic pain (tension headache, chronic pelvic pain) and chronic disease with intermittent symptoms (migraine, asthma and gastroesophageal reflux) [7].

b- The precipitating factors (triggering):
The precipitating factors fall into two categories:
- The factors that preceded the onset of symptoms during the past year: stressful situations (death or serious illness of a loved one), conflict situations, psychosocial stress, acute psychiatric disorders, injury or non-psychiatric illness [18].
- The factors present regularly just before the onset of symptoms: distressing emotions (anxiety, sadness or anger), or even positive emotions (joy, surprise), situations of conflict, frustration or requiring patience, consultancies or medical examinations [22].

c- Perpetuating (maintenance) factors:
It is a question of the multiplication of the opinions and medical examinations, a strong attention of the entourage, financial or social benefits, the refusal of the psychogenic etiology, the depression and the anxiety. They cause an exacerbation and a perpetuation of symptoms. Family difficulties and anxiety of loved ones are also possible maintenance factors.

VI. Diagnosis of PNES:
Two main distinct psychopathological profiles have been identified [12]:
- A traumatized group: majority, characterized by a clear predominance of women, numerous psychiatric comorbidities, a strong dissociative tendency and alexithymia. The most common triggers for PNES in this group are feelings of helplessness and anxiety. The strong dissociative tendency appears to be the main underlying mechanism.
- A non-traumatized group: minority, characterized by a male over-representation, a low intellectual level, a tendency to learning difficulties, a history of head injuries, little psychiatric co-morbidities, a low propensity to dissociation and a weaker alexithymia. The most common triggers in this group are annoyance and frustration. Neurodevelopmental or neurobiological factors are in the foreground.

The video-EEG recording of the PNES and their interpretation by a neurologist experienced in epilepsy constitute the reference method for diagnosis. When the crises could not be observed by a clinician, the diagnosis based on the anamnesis of the patient and eyewitnesses and the normality of the inter-critical EEG is only possible. When the crises have been seen or observed by a clinician, the degree of certainty depends on his experience and whether or not there is a normal critical EEG.

Only the clinical semiology of crises obtained by reliable eyewitnesses and ideally recorded on video coupled with the analysis of the per-critical EEG allow to base the diagnosis with certainty.

The diagnosis of PNES is a diagnosis of elimination of organic pathologies, then a diagnosis of elimination of other psychiatric disorders. VII- Differential diagnosis:
There are many organic differential diagnoses of PNES:
First, epilepsy: the differential diagnosis between these two entities is essential, but the situation is sometimes complicated, epilepsy can be a risk factor for the development of PNES. Several clinical situations must therefore be underlined, potentially responsible for diagnostic difficulties, especially during certain partial epilepsy, parietal (sensitivomotor symptoms without modification of the surface EEG), temporo-insular (experiential and / or dysautonomic phenomena see ictal syncopes), or partial seizures with complex behavioral changes.
Lipothymia, syncope, abnormal movements, and parasomnias (somnambulism, narcolepsy, and sleep apnea).
In another study carried out in neurological resuscitation on eighteen patients initially treated for a state of refractory tonic-clonic epilepticus, eight subjects actually presented a "psychogenic epileptic condition" [2]. The clinical diagnosis of PNES is difficult. The difficulty in establishing a sensitive and specific clinical diagnosis necessitates carrying out certain additional examinations (brain imaging, cardiac exploration, etc.).

VII. Paraclinical explorations:
Video and electroencephalographic recording in monitoring remains the examination of choice to differentiate CPNE and epileptic seizures. However, if EEG criteria are necessary to make the diagnosis of epilepsy, the absence of these is not sufficient to confirm or rule out the presence of PNES. In a healthy subject, there may actually be non-specific paroxysmal activities outside of any clinical epileptic seizures or PNES. In addition, the inter-critical EEG does not make it possible to discriminate CPNE and epileptic seizures because anomalies or even epileptiform discharges may be present. If the frequency of seizures is low, an EEG-video recording may not be enough to explore the disorder during a 24-hour hospital stay. However, techniques for raising awareness of epilepsy by hyperpnea or intermittent light stimulation can suggest an PNES, even in an epileptic patient. Adding ECG to video-EEG is important for monitoring the heart rate during the critical phase.

VIII. Diagnostic announcement:
Announcing the diagnosis of PNES is a very difficult but crucial step. The quality of the diagnostic announcement of PNES has a strong impact on adherence to diagnosis and treatment and therefore on the prognosis of PNES patients. The diagnosis of PNES often comes to replace or supplement a preliminary and incorrect or incomplete diagnosis of epilepsy, sometimes established for several years. In addition, the clinician may be uncomfortable himself. With the patient's agreement, the presence of a family member during the announcement is desirable. The doctor must be available to explain the stages of the clinical process that documents the diagnosis and explain why it sometimes took several years to ensure the absence of comorbid epilepsy. The message must be passed on to the patient "that he is considered to be a real patient, that he is believed and that he is not a simulator". You have to explain to him that he has a real illness, and that his illness has been clearly identified. The most important points are [11]:
- Reassure the patient about his illness and its consequences;
- Name the disease;
- The affection is real and recognized. It is not a simulation;
- Answering questions by avoiding any stigmatization or trivialization of disorders;
- Explain the existence of this disease and the difficulty of its diagnosis (PNES are frequent);
- Discuss the possible causes and factors;
- Offer therapeutic treatment;
- The more certain the clinician is of the diagnosis and the more comfortable with the explanations, the more the chances of acceptance of the diagnosis by the patient and his family.

IX. Treatment And Prognosis
Faced with the comorbidity PNES/ epileptic seizures and the diagnostic delay, the majority of the authors agree on the interest of setting up multidisciplinary consultation centers jointly engaging neurologists and psychiatrists. Baker et al. there are no studies with a sufficient level of scientific evidence to conclude on unanimous therapeutic recommendations [3].

The therapeutic objective does not consist simply in the disappearance of the symptom because the remission of the crises is not here a criterion of medical cure or better social adaptation. Several authors emphasize the importance of treating associated psychiatric comorbidities. Psychotropic treatment is indicated in more than half of the cases [16]. If the PNES are isolated, without associated epilepsy, the anticonvulsant treatment will be stopped. Psychotherapy should be offered. In all cases, the neurologist retains a central role even after the diagnosis of PNES is announced to the patient. Despite all these therapeutic orientations, the medium-term prognosis is reserved. In a cohort study, Reuber et al. find that four years after diagnosis, more than 70% of patients still have PNES and more than 50% are dependent on social assistance with significant secondary benefits [23].
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X. Conclusions

The PNES are often confused with drug-resistant epilepsy. Their diagnosis is often difficult and delayed. A misdiagnosis which can be harmful, responsible for iatrogenism (anticonvulsant treatment is often wrongly prescribed) and increased morbidity. Video-electroencephalographic recording is the para-clinical examination of choice.

The difficulty of diagnosing PNES requires a joint approach between neurologists and psychiatrists. The announcement of the pathology is essential and conditions the treatment. The earliest diagnosis and the quality of the announcement are the most important parameters of therapeutic management.

Bibliography:


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