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RESEARCH ARTICLE

INTERMITTENT ACUTE PORPHYRY AND SCHIZOPHRENIA: ABOUT A CASE.

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Abstract

Several of heterogeneous psychiatric symptoms have been associated with acute intermittent porphyria such as anxiety, affective distortions, behavioral changes, and personality and psychotic symptoms.

These symptoms can be difficult to identify as being related to porphyria, as symptoms can arise at any time during the disease process. Therefore, some patients may present psychiatric disorder following acute intermittent porphyria.

As well, an early diagnosis and an appropriate treatment of psychiatric manifestations impact positively the course of the disease.

Through this clinical case of patient presenting comorbidity "acute intermittent porphyria + schizophrenia", we will highlight the psychiatric manifestations and the psychopharmacological difficulties encountered.

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Introduction:-

Porphyries are hereditary diseases characterized by an enzymatic deficiency in the heme synthesis chain. Intermittent Acute Porphyria (IAP) is considered the most common variety, it evolves by attacks activated by porphyrinogenic factors, whose the most incriminated are barbiturates.

Acute hepatic porphyria has in common the risk of acute porphyria attacks. In fact, three major syndromes characterize them initially : abdominal pain, neurological disorders and mental disorders.

The confirmation of diagnosis is made by the urinary dosage of porphyrins: Uroporphyrins, coproporphyrins and their precursors. The definitive diagnosis of IAP is based on the discovery of a 50% deficit in the Urosynthetase's activity.

It has been observed in various studies that individuals with a manifest Inflammatory Bowel Disease (IBD) are fourfold risked to have schizophrenia or bipolar disorder. Otherwise, first-degree relatives of people with (IBD) are dual risked to develop schizophrenia or bipolar disorder.

In this regard, we will illustrate this study with a clinical case of a patient who presents an intermittent acute porphyria with schizophrenia.

Clinical case:

Mr. Z is born in 1999. He is the third of a sibship of 03. Its psychomotor development was without peculiarity. He has for antecedents a psychotic maternal uncle with IAP.

At the age of 8, he had abdominal pain, seizure, respiratory distress, and quadriplegia. He was hospitalized in intensive care, where the suspicion of porphyria was confirmed by a deficit of urosynthetase activity. Two years later, he would have presented the same symptoms requiring a second hospitalization in intensive care, but this time he came out with motor sequelae.

He was able to resume his studies until the ninth grade, then he integrated a vocational training that he would have stopped because of the appearance of a symptomatology made of isolation, insomnia, verbalization of delusional idea of persecution, and denial of filiation with hetero-aggressive behavior.

He was taken care in psychiatric ambulatory for one year. An internal medicine opinion was requested, which did not reveal any peculiarities. In addition, a biological assessment and a cerebral Computed tomography (CT) were revealed without particularity, and his evolution was favorable at the beginning.

However, the clinical evolution has made it possible to make the diagnosis of schizophrenia because of the persistence of a delusional syndrome and the absence of socio-occupational reintegration. He was first put on 10 mg/day olanzapine, stopped following neutropenia revealed during a control of complete blood count. Thereafter, a bad response to risperidone and then Aripiprazol 30 mg/day had shown treatment with chlorpromazine 700 mg/day with a good evolution.

Discussion:-

The prevalence of IAP is 1/75 000. In general, the disease manifests after puberty and affects women (80% of cases) preferentially.

They are presented by a very intense abdominal pain (>95% of patients), neurological disorders and / or psychological disorders. Seizures are most often triggered by exogenous factors (porphyrinogenic drugs, alcohol, infections, hypocaloric diet, stress) and / or endogenous (hormonal, related to the menstrual cycle). (1)

IAP is due to a deficiency of porphobilinogen deaminase (PBG-D). The enzyme deficiency follows the mutations of the HMBS gene (11q23.3) coding for PBG-D. The transmission is autosomal and dominant. (2)

In a recent survey based on Swedish data, 16% of individuals with manifest IAP and 7% of latent mutation carriers had a "psychiatric illness". These results strongly support the idea that IAP is linked to schizophrenia and bipolar disorder. (3)

Actually, it supports the idea of a shared genetic etiology. IAP can be considered as a natural model of psychotic and affective illness because the signs and symptoms are similar to schizophrenia and bipolar disorder. In IAP, there is a decrease in the function of tryptophan 2, 3-dioxygenase and indoleamine 2,3-dioxygenase which constitutes the first enzymatic step limiting the rate of production of kynurenine.

Curiously, the kynurenine pathway has been implicated in schizophrenia and bipolar disorder. The modified metabolism of kynurenine, although associated with many diseases, could therefore be a mechanism behind the associations observed. (4)

A first study that explored the association between IAP and bipolar disorder revealed that compared to individuals without obvious IAP, people with (IBD) were four times more risked to have schizophrenia or bipolar disorder. First-degree relatives of people with (IBD) had a dual risk of developing schizophrenia or bipolar disorder. (5)

IAP can begins with polymorphic psychiatric disorder ranging from frequent mood disorders with anxiety involvement to delusions, hallucinations and confusional syndrome. (6)

The triggering factor is most often a medicinal in take. So, it is interesting to have an idea about some prohibited drugs in the field of psychiatry in case of IAP including: sertraline, miancerin, amisulpride, alprazolam, zolpidem, clorazepam, nitrazepam MAOI, lamotrigine, sodium valproate, carbamazepine, and hydroxyzine. (7)

In chronic psychiatric symptomatology, schizophrenia should be considered, in patients with IAP, discomfort because of the high risk compared to the general population.

Conclusion:-

In sum, there is a solid association between IAP, and schizophrenia that deserve increased attention in research and clinical practice. The interest of this clinical case is how important to be careful during prescription with patients who present IAP, and to firstly think of the schizophrenia during appearance of psychiatric symptoms.

Conflicts of interest

The authors do not declare any conflict of interest for this work.

Authors' collaboration

All authors were actively involved in the development of this work.

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