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Mini Review

Association between schizophreniform symptoms and Dandy-Walker Syndrome: Mini-Review

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Abstract

The Dandy-Walker Syndrome (DWS) is a rare congenital disease, characterized by anatomical alterations affecting structures in the posterior fossa, as cerebellum and fourth ventricle. There are 19 case reports and two case series associating DWS and schizophreniform symptoms. Probably both conditions are related, as well as a role of the cortico-thalamic cerebellar circuits in the onset of psychosis leading to a Cerebellar Cognitive-Affective Syndrome (CCAS). Secondary cases of psychosis deserve adequate investigation and personalized treatment. They may contribute to a better understanding of the mechanisms of the primary psychosis.

Keywords: Dandy-Walker syndrome, Schizophrenia, Psychosis

Abbreviations: Dandy-Walker syndrome, CCAS - cerebellar cognitive-affective syndrome

Introduction

Psychotic disorders are characterized by the presence of delusions, hallucinations, and disorganization of thought and behavior, classified into primary and secondary categories. Primary disorders include the spectrum of schizophrenia and mood disorders. Secondary disorders are associated with the use of medications, psychoactive substances, or medical conditions [1]. It is estimated that the prevalence of schizophrenia is between 0.3-0.7%, with schizophreniform disorders having a similar prevalence [2].

In recent years, correlations of neuroanatomical changes in subjects with psychotic disorders have been investigated. In schizophrenia, significant reductions in the gray matter have been identified globally and regionally in the frontal and temporal lobes. Evidence from recent studies points to a hub involved with psychotic symptoms formed by basal ganglia, corpus callosum, insula, and cerebellum [2].

The cerebellum is associated to balance, fine motor coordination, cognitive functions (as attention and executive functions), emotional and behavioral processes. Cerebellum automatically maintains behavioral homeostasis, based on implicit learning and according to the context [3]. Many aspects still need to be clarified on the potential role of the cerebellum in psychotic disorders. Cases of Dandy-Walker Syndrome (DWS), which have schizophreniform symptoms and cerebellar burden can provide important information about this complex relationship [4]. This mini narrative review aims to explore scientific evidence of the relation of cerebellum and schizophreniform symptoms by studying cases of DWS.

Materials and Methods

Keywords "schizofrenia", "psychosis" and "Dandy Walker Syndrome" were inserted in two databases: Google Scholar and PubMed, in November 2023. Articles must be written in english or spanish, address the proposed theme and be published no longer than 15 years ago. The articles that were not case reports, case series, those in which DWS was not the main topic, and those that explored other aspects of the syndrome besides psychosis were excluded.

Results and Discussion

750 articles were found, 21 met the inclusion criteria, divided into 19 case reports, shown in (Table 1), and two case series are from articles by Stamboliu, 2017 and Leslie, 2023.

Table 1: Case reports and letters to editors found in the literature review.

Chen, 2022		Porras Segovia, 2021	Martinez, 2020	Gama Marques, 2019
Trébout, 2017	Dawra, 2017	Sinha, 2017	Isidro-Garcia, 2017	Rochanchandra, 2016
Williams, 2016	Orsolini, 2016	Blaettner, 2015	Aune, 2014	Zincir, 2014
Mauritz, 2014	Buonaguro, 2014	Sidana, 2013	Ryan, 2012	Gan, 2012

Influence of the Cerebellum on the Psychopathology of Schizophrenia

Studies have shown that volumetric reduction of the cerebellar vermis, alterations of the cortico- thalamo-cerebellar-cortical pathway (CTCCP) [3], including the reduction of Purkinje cells and their excitation by granule cells 5, may underlie the emergence of cognitive and clinical symptoms of Schizophrenia [3,5]. Cerebellum is linked with the frontal orbital cortex, limbic system, and reticular structures. A lesion in cerebellum can trigger a cerebellar cognitive affective syndrome (CCAS), leading to problems in almost all cognitive domains, psychosis and mood disorders. A striking feature is thought dismetria, which can be related to psychotic symptoms [6]. Many causes can impact in cerebellar structure, as Niemann-Pick type C or other genetic anomalies, early developmental disorders as DWS or toxic injuries. This could explain similar cerebellar dysfunction between developmental schizophrenia and other disorders [7].

Dandy-Walker Syndrome and Schizophreniform Disorders

DWS is a rare congenital brain malformation, first described in 1914 by the neurosurgeon Walker Dandy. The diagnosis is based on neuroimaging exam containing the classical triad of complete or partial agenesis of the cerebellar vermis, cystic dilation of the fourth ventricle, and enlargement of the posterior fossa, Anatomical variations of this triad may occur [8,9]. The prevalence estimated is 1 in 25,000 to 30,000 live births [9]. Most cases are sporadic and first-degree relatives have increased risk [8]. Symptoms of DWS emerge in early childhood, as macrocephaly, increased intracranial pressure, hypotonia, motor deficits and problems in intellectual development 8. When the clinical symptoms emerge in late adolescence, hallucinations, delusions, and mood alterations are more frequent [10].

Although there are no studies of prevalence or clinical trials, the comorbidity psychosis/Dandy- Walker Syndrome (DWS) is frequently reported in the literature, a expressive number considering the rarity of the syndrome. This suggests a causal relationship. The hypoplasia of the cerebellar vermis of DWS may precipitate psychosis or increase the risk to a brain or person already vulnerable to it [5,7].

Interestingly, Leslie and colleagues [11] reviewed 18 cases of psychosis associated with predominant hypoplasia of the cerebellar vermis and none of the individuals had the Dandy-Walker malformation. Instead, they had symmetrical predominant hypoplasia of the cerebellar vermis with variable enlargements of the posterior fossa. They suggested a genetic basis for such alterations [12-25].

Conclusion

The early detection of the congenital alteration called DWS becomes important for understanding psychotic manifestations and personalized treatment. However, not all patients with DWS will present with schizophreniform symptoms, and not all patients diagnosed with schizophrenia will have cerebellar alterations. The relationship between DWS and psychosis is complex and requires further studies to better understand the connection between these conditions. MRI is as interesting tool for screening causes of secondary psychosis.

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Conflict of Interest

Authors declare that they have no conflicts of interest.

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