Case Report

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Acute Psychosis Associated with Septo-Optic Dysplasia (De Morsier Syndrome): A Case Report

Goyal DK^{1*}, Singal A², Goyal A³ and Sandhu G⁴

¹Department of Psychiatry, University of Nevada, Reno, USA

²University of San Francisco, San Francisco, USA

³Memorial Health System, Marietta, Ohio, USA

⁴University of South Dakota, Vermillion, USA

***Correspondence:** Dharmendra K Goyal, Department of Psychiatry, University of Nevada, 5190 Neil Road, Reno, NV 89502, USA, Tel: 347-654-7109; Fax: 775-789-4246; E-mail: doc.dharam83@gmail.com

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Abstract

Background: Septo-optic dysplasia (SOD), a variable combination of abnormalities of cerebral midline structures, is a clinically heterogeneous syndrome. Psychiatric symptoms in SOD were implicated in one case due to abnormal regulation of neuroendocrine structures.

Objective: Discussion of a case of SOD associated with psychosis and depression, exploring the role of anatomical and endocrine abnormalities in producing psychiatric symptoms.

Methods: Literature was reviewed on the PubMed, Medline, and Web of Science. One case of SOD with psychosis was reported in 2008.

Conclusions: Disruption of complex neural pathways, including the septum pellucidum and other limbic structures may have been involved in the psychiatric disturbances in this case. Further reporting of similar cases is needed for more understanding of these associations.

Keywords: Septo-optic dysplasia, Psychosis, Optic nerve hypoplasia, Pituitary insufficiency

Abbreviations: SOD: Septo-Optic Dysplasia; GH: Growth Hormone

Introduction

SOD is rare congenital disorder composed of a variable constellation of midline brain developmental abnormalities including optic nerve hypoplasia, agenesis of the septum pellucidum, abnormalities of the corpus callosum and pituitary dysfunction [1]. Reported incidence of SOD is 1/10,000 [2]. The clinical features of SOD can vary in different individuals including visual impairment, partial pituitary insufficiency, psychomotor retardation, seizures, altered thermoregulation, and conjugated hyperbilirubinemia. In some studies, 62% of patients show isolated pan hypopituitarism and 30%, optic nerve

hypoplasia, and agenesis of midline structures [3]. Growth hormone (GH) deficiency is the most common hormonal dysfunction reported, whereas vision and optic disc alterations may be absent. SOD has also been observed in autopsic series of unexpected deaths, at all ages [4,5]. In the past multiple studies have shown many different forms of abnormalities caused midline brain developmental disorders and schizophrenia [6,7]. Yet, very few have looked closely at particular cases, as to how these midline brain abnormalities were formed, diagnosed, and treated to produce SOD. This can be due to the limited studies and research found in publication. Our purpose is to provide a preliminary understanding

of SOD and to expand upon the limited knowledge of the disorder.

Case Report

A 22-year-old African American female, with no significant past psychiatric history or medical history of congenital visual impairment, was admitted to the medical ward. She had self-reported fevers up to 102°F, auditory and visual hallucinations, and bizarre behavior. Past history was obtained from the mother, patents was born following a prolonged pregnancy at 10 months of gestation. She had neonatal jaundice with Neonatal Intensive Care Unit (NICU) care. She developed a seizure disorder around the age of 4 and was treated with phenobarbital for one year. The patient had not yet experienced menses and was noted as per the family to be anhidrotic. She had short stature (157 cm) in comparison to parental height and episodes of intermittent temperature elevations (37.7°F).

Mental status examination

A young female with staged age and good hygiene. She had poor eye contact. Mood was anxious and depressed with constricted affect. Speech was limited and focused. Thought process with loss of association, included hyper-religious auditory and visual hallucinations. Insight and judgment were also impaired.

Labs and radiological tests

Serum chemical, microbiological and cerebrospinal fluid analysis for infectious, autoimmune, and paraneoplastic phenomenon were negative. Laboratory analysis revealed low normal growth hormone level (0.03 ng/mL), depressed insulin-like growth factor binding protein-3 (0.07 ng/mL), and depressed morning plasma corticotrophin (8.8 pg/mL). MRI of the brain in figure 1 shows ectopic neurohypophysis and hypoplasia of optic nerves, optic chiasma, and optic tracts that confirmed the diagnosed of septo-optic dysplasia.

Treatment

Patient's symptoms of depression and psychosis improved with a trial of fluoxetine (40 mg daily) and risperidone (1 mg at bedtime).

Discussion

Diagnostic criteria include the presence of ≥ 2 of the following: (i) optic nerve hypoplasia, (ii) agenesis/hypoplasia of septum pellucidum and/or corpus callosum and (iii) hypothalamic-pituitary dysfunction.

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Mutations in the homeobox genes HESX-1, OTX2, SOX2, and PAX6 are associated with SOD. SOD is also associated with recurring seizures, developmental delay, thermoregulatory dysfunction, and conjugated hyperbilirubinemia. In 2008, Bini et al. reported a case of SOD associated with depression and psychosis, treated with antipsychotics and mood stabilizers. To our knowledge, we report the second such association.

Figure 1: MRI Images above show ectopic neurohypophysis, and hypoplasia of optic nerves, optic chiasma, and optic tracts.

Conclusion

SOD induced fluctuations in neuroendocrine circuits may be variably associated with psychopathology of depression and acute psychosis. SOD should be included in the differential in young adults presenting with psychotic symptoms. Further literature reporting of this rare condition should be encouraged to aid in early diagnosis and proper management.

Declarations

All authors have no conflicts of interests. No funding needed for the data gathering. All authors give consent for the publication.

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