Case Report

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JOURNAL OF NEUROLOGY AND NEUROSCIENCE
ISSN 2171-6625

2017
Vol. 8 No. 3: 197

DOI: 10.21767/2171-6625.1000197

A Case of Empty Sella Syndrome Presenting with Neuropsychiatric Symptoms

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Abstract

Empty Sella Syndrome (ESS) is a condition where the pituitary gland is partially or totally absent from the pituitary fossa. It can be asymptomatic in most cases and found accidentally in imaging studies of brain. In symptomatic cases it presents with endocrine abnormalities due to pituitary damage visual defect from compression of optic chiasma neuropsychiatric symptoms benign intracranial hypertension CSF leakage from the nose and headache. We present a case of a 24-year-old male with psychotic symptoms, soft neurological signs with learning disabilities, empty sella syndrome, visual symptoms and cardiac valvular defect. This case helps us to understand the neuroscience behind the psychiatric illnesses having access to advanced medical technologies to look inside the brain.

Keywords: Empty sella syndrome; Psychosis; Learning disabilities; Neurological symptoms

Received: May 02, 2017; **Accepted:** June 03, 2017; **Published:** June 06, 2017

Citation: Bardoloi PS, Abba-Aji A. A Case of Empty Sella Syndrome Presenting with Neuropsychiatric Symptoms. J Neurol Neurosci. 2017, 8:3.

Introduction

Empty sella syndrome (ESS) is a condition where the pituitary gland is partially or totally absent from the pituitary fossa [1,2]. It can be asymptomatic in most cases and found accidentally in imaging studies of brain. In symptomatic cases it presents with endocrine abnormalities due to pituitary damage [3], visual defect from compression of optic chiasma [4], neuropsychiatric symptoms [5], benign intracranial hypertension [6], CSF leakage from the nose and headache [1,7]. We present a case of a 24-year-old male with psychotic symptoms, soft neurological signs with learning disabilities, empty sella syndrome, visual symptoms and cardiac valvular defect. The presence of various systemic involvements in this male, who was admitted to a general psychiatric ward for psychosis, evoked the question in mind if these are associated due to developmental defect or are present incidentally. As we are heading towards an era where we are trying to understand neuroscience of psychiatric disorders [8], this seems to be an intriguing case.

Although ESS can be found commonly as incidental finding during brain studies [7], symptomatic case of ESS is rare. ESS can be primary or secondary. Primary ESS is due to defective development of diaphragma sellae, with arachnoid herniation to the pituitary fossa due to increased intra cranial pressure [9], which is found mostly in young people [10]. Secondary ESS is due to infarction or atrophy of pituitary gland and usually found in obese and middle aged women [11,12]. Incidence of primary ESS is 8-35%. Male to female ratio is 1:5 [12].

Associated conditions include ophthalmologic, endocrinal, neurologic and psychiatric conditions [11]. Along with the midline structural abnormalities [13], it was reported in cases of 22q 11 deletion syndrome [14] and cases of psychosis and schizophrenia [5].

Case Presentation

We present a 24-year-old single man with ESS, who was admitted to adult psychiatry unit. He presented with visual hallucinations of images of giant spiders and distorted human figures, tactile hallucination of something crawling on his skin, associated with tingling and feeling of numbness in one side of his body. He also reported some visual difficulties, distorted images with increasing sensitivity to bright light and sound. These symptoms were present for about a year, worsened for about 2 months prior coming to the hospital. This case helps us understand the neuroscience behind the psychiatric illnesses, having access to advanced medical technologies to look inside the brain Patient denied using any alcohol, drugs or nicotine.

When he was 8-year-old he was diagnosed with borderline learning disability and he completed high school with average grades. At age of 11 he was diagnosed oppositional defiant

disorder (ODD) and possible attention deficit hyperactivity disorder (ADHD). He was not on any medication and did not have any psychiatric follow up for years after that, until his recent presentation with psychotic symptoms.

He was diagnosed with congenital valvular heart disease, is being followed up regularly and is waiting for surgical repair. His mother had fibromyalgia, maternal grandmother and great grandfather had Schizophrenia and bipolar disorder respectively. His paternal grandfather was diagnosed with multiple sclerosis. He struggled in school and was unable to keep a steady job. He preferred to stay by himself as he felt socially awkward. He underwent ophthalmological examination that ruled out major ocular problems and was diagnosed with possible ocular migraine.

Neurological examination excluded major neurological disorders, but neurological aetiology of visual hallucinations could not be ruled out. He was investigated for pituitary hormones but no major abnormalities were noted.

Investigations

Blood work

- Mostly normal except low free testosterone and slightly low luteinizing hormone.

CT scan of the head

- It showed a small region of white matter low density in the right frontal lobe. The sella turcica appeared nearly empty.

MRI head with/without contrast

- Showed increased FLAIR/ T2 signal near anterior horn of right lateral ventricle (upto 1.2 cm), small pericallosal and periventricular lesions as well as few subcortical lesions, mostly in anterior region and sparing occipital lobes. No mass effect was evident.

EEG

- Showed dysrhythmia with occasional slowing in right temporal region. High amplitude sharp slow complexes were prominent in frontal region. The EEG was interpreted by a neurologist as nonspecific and not suggestive of epilepsy.

Echocardiogram

- Showed bicuspid aortic valves with mild aortic stenosis and mild aortic regurgitation.

Psychological tests

- Had Full-Scale IQ in the average range of intellectual functioning (42nd percentile) on the Wechsler Abbreviated Scale of Intelligence (WASI-II). His Executive function indicated mildly impaired task planning and moderately impaired verbal working memory. His Perceptual-motor skills did not show deficits in visual or motor functions.

Differential diagnosis

- Psychosis secondary to another medical condition (DSM-5) [15].
- Schizophrenia Spectrum Disorder (DSM-5) [15].

Treatment

Patient received olanzapine 10 mg at night for his psychotic symptoms with resulting improvement of the symptoms.

Outcome and follow-up

Patient was discharged home with follow up appointments to see psychiatrist, cardiologist and neurologist.

Discussion

As we progress through the 21st century, we are increasingly more eager to understand the neuroscience behind the psychiatric illnesses [8], having access to advanced medical technologies to look inside the brain. Although historically not much evidence were found to support any correlation between Schizophrenia and ESS, case report of a set of monozygotic triplets, with schizophrenia with ESS was found, which discuss possible correlation between the two [5,16]. The triplets with similar psychotic symptoms were found to have empty sella turcica in MRI findings, along with other identical abnormalities, suggesting a common genetic origin of ESS and schizophrenia. Midline anomaly with the presence of cardiac abnormalities in our case is yet to be studied for genetic link [17].

Conclusion/Learning points/Take Home Messages

Schizophrenia is a polygenic, multifactorial disorder that can have associated other genetic anomalies. Empty sella turcica is a radiologic finding and mostly asymptomatic. However in symptomatic cases, symptoms may be due to developmental abnormalities or due to physiological effect of displaced or absent pituitary or compression effect of pituitary. Uncommon cases like this would help us gain more insight into the neurobiology of schizophrenia.

Disclosures

Our patient was not involved in any clinical trial.

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