CASE REPORT

Case Report: Atypical psychotic onset of type I Arnold-Chiari malformation [version 1; referees: awaiting peer review]

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v1 **First published:** 28 Sep 2015, **4**:915 (doi: 10.12688/f1000research.6975.1)
Latest published: 28 Sep 2015, **4**:915 (doi: 10.12688/f1000research.6975.1)

Abstract

Introduction: We report a case of type I Arnold-Chiari malformation that is very peculiar because of its particular onset especially characterized by psychiatric symptoms. These symptoms were so prevailing that, for fifteen years, they masked the neurological aspects and the patient was treated with high doses of psychotropic drugs without any benefit. If the Arnold-Chiari malformation had been diagnosed before the development of severe hydro-syringomyelia, the patient could have underwent decompressive neurosurgery which may have improved her quality of life.
It is worthwhile to highlight that psychotic symptoms may be caused by this congenital malformation, that typically has an aspecific onset. Therefore it's important to consider an eventual organic etiology while challenging a resistant clinical picture with unusual presentation.
Case description: A 51-year-old woman reported neurological symptoms consisting of headaches, blurred vision, diplopia, tinnitus, vertigo and psychiatric symptoms including obsessive ideas about the fear of killing her son, auditory and visual pseudo-hallucinations. The symptoms had developed suddenly at the age of 35 years and persisted thereafter. She underwent multiple hospitalizations in psychiatric units and was treated with a variety of psychopharmacological approaches without substantial improvement. We performed a brain MRI that identified a type I Arnold-Chiari malformation. We assessed psychiatric symptoms using the Structured Clinical Interview for DSM IV Axis I Disorders, the Brief Psychiatric Rating Scale, and the Yale-Brown Obsessive-Compulsive Scale. A tailored psychopharmacological therapy led to a partial improvement in mood and anxiety but not in hallucinations.
Discussion: We want to highlight how important is, in everyday psychiatric clinical practice, not to focus only on psychiatric aspects but consider the patient globally, because in this case psychiatric problems were the onset presentation of a rare neurological syndrome.

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How to cite this article: Di Genova C, Charitos S, Ba G and Viganò CA. **Case Report: Atypical psychotic onset of type I Arnold-Chiari malformation [version 1; referees: awaiting peer review]** *F1000Research* 2015, **4**:915 (doi: 10.12688/f1000research.6975.1)

Grant information: The author(s) declared that no grants were involved in supporting this work.

Competing interests: No competing interests were disclosed.

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Introduction

Arnold-Chiari malformation (ACM) is a congenital brain anomaly characterized by herniation of the cerebellar structures through the foramen magnum. Its prevalence ranges from 0.2 to 1%[1]. It is characterized by four subtypes: type I consists of cerebellar tonsil herniation, which may be associated with syringohydromyelia, skull base alterations and hydrocephalus; in type II the herniation also involves the contents of the posterior fossa and it's frequently associated with myelomeningocele and hydrocephalus[2]; ACM type III and IV are sporadic and characterized by the descent of both the cerebellum and brainstem into the spine and internal sac; and type IV is associated with cerebellar atrophy[3]. In Arnold-Chiari malformation type I and II, symptom onset is delayed until about the third decade[4]. General symptoms and signs include headaches, dizziness, tinnitus, visual or oculomotor symptoms, dysphagia, trunk or extremity dysesthesias, ataxia and drop attack[4]. In addition, psychiatric complications like anxiety and mood disorders are frequent and affect the quality of life and the global functioning[5]. There is a paucity of literature pertaining to psychiatric illnesses associated with ACM and, to date, only three cases have been reported of an anxiety disorder in association with this specific malformation[6–8] and just two cases of ACM associated with psychotic features[9,10].

Case description

We report the case of a 51-year-old woman, who was married and had a 23-year-old son.

She was hospitalized in December 2014 in the psychiatric department of our hospital for persistent auditory hallucinations and visual pseudo-hallucinations together with headache, diplopia, tinnitus and vertigo. She was severely obese (BMI 34) and she was taking Olanzapine (30 mg/day), Duloxetine (90 mg/day), and Alprazolam (3 mg/day).

Since the age of 35, the patient had experienced headaches, oculomotor symptoms like blurred vision and diplopia, tinnitus and vertigo. These symptoms were so debilitating that she spent long periods of time lying in bed in a room with half-light and reduced noise. During the same period she started to experience psychiatric symptoms, at the beginning characterized by excessive anxiety, epigastric discomfort, restlessness and insomnia. Some months later she began to develop obsessive ideas about the fear of killing her son, who was 12-years-old at the time, without any compulsive behaviours. She also experienced auditory hallucinations with imperative voices and visual pseudo-hallucinations. In this period her mood was slightly depressed without suicidal intent.

Consequently, in 2001 at the age of 36, she was hospitalized in a psychiatric unit with the diagnosis of Schizophrenia. In the previous years she had never been hospitalized, there was no personal history of previous psychiatric disorders or substance misuse and also no family history of mental illness. The patient has a high school degree and since the age of 28 she worked as a secretary. During hospitalization no abnormalities were found in blood parameters, electrocardiography and urine analyses. She was prescribed Amisulpride tablets (300 mg/day) and Paroxetine tablets (40 mg/day), but after 5 days the patient developed extrapiramidal symptoms and akathisia. At this stage Amisulpride was discontinued and Haloperidol, orally administered at the dose of 6 mg/day was introduced. She experienced only a partial benefit: hallucinations and obsessive ideas persisted although the patient acknowledged an improvement in mood and anxiety. After 4 weeks she was discharged from the hospital and began follow-up outpatient care with a psychiatrist. Although she was treated sequentially with various antipsychotic drugs and antidepressants (Promazine, Aripiprazole, Olanzapine, Citalopram, Venlafaxine, Sertraline) she had only relative clinical improvements in mood and anxiety but not in hallucinations. She also continued to suffer from headaches, dizziness and tinnitus.

During the hospitalization in our unit, in consideration of the neurological symptoms, a brain MRI was performed. The results revealed a 7 mm cerebellar tonsils herniation into the foramen magnum and the presence of an irregular cavity in the spinal cord, particularly between C4 and D1. An electroencephalograph (EEG) was inconclusive. A diagnosis of ACM-I with syringohydromyelia was established, the neurological assessment was within normal limits. We also had a consultation with the department of neurosurgery which ruled out the possibility of a decompression surgery because of the presence of a consolidated hydromyelia not likely to remit with surgery. We assessed the psychiatric symptoms using the Structured Clinical Interview for DSM IV Axis I Disorders (SCID I) that, in consideration of the MRI result, was diagnostic for a Psychotic Disorder Due to Another Medical Condition[11]. We also administered the Brief Psychiatric Rating Scale (BPRS, score: 30)[12] and the Yale-Brown Obsessive-Compulsive Scale (YBOCS, score: 9)[13]. The decided treatment was: Risperidone tablets (3 mg/day; Olanzapine was withdrawn in consideration of the body weight), Sertraline tablets (150 mg/day) and Pregabalin tablets (100 mg/day). We observed that the patient underwent a substantial improvement in mood, insomnia, epigastric discomfort but she still did not experience a concrete improvement in obsessive ideas and hallucinations. The patient was discharged after three months of hospitalization in a low intensity psychiatric unit and, afterwards, began outpatient psychiatric and neurological follow-up.

Discussion and conclusions

In this report we aimed to present the case of a patient with a peculiar presentation of ACM in which there is a paucity of neurological signs and symptoms and a prevalence of psychiatric aspects. As a result the patient has always been treated as a pure psychiatric patient despite the lack of impact of various psychotropic drugs on her symptoms; the neuroimaging examination, in fact, came only recently. Although the presentation is characteristic of a psychiatric disorder, the presence of nonspecific neurological symptoms should suggest a general medical condition causing psychiatric symptoms, especially in the absence of some of the most important "psychiatric clues" (such as family history, substance abuse or previous psychiatric subthreshold symptoms). To our knowledge, there is a lack of literature pertaining to psychiatric illnesses associated with ACM. In fact only three cases of an anxiety disorder in association with ACM have been reported to date[6–8], and just two reports of ACM associated with psychotic features (ACM association with epileptiform events[10] and with a major neurocognitive disorder[11]). Moreover, although anecdotal, some cases of ACM with psychotic features have already

been described[14]. We decided to present this case because we consider mandatory not to underestimate the psychiatric symptomatology and to perform global investigations, particularly of neuroradiological nature. We believe, indeed, that the patient's psychotic symptoms, especially auditory hallucinations and visual pseudo-hallucinations, are due to the organic pathology. It's clearly understandable that this type of symptomatology, especially if unrecognized, may have led to a worsening of the anxious and depressive aspects.

Consent

Written informed consent for publication of their clinical details was obtained from the patient.

Author contributions

Dr CDG and Dr SC followed the case and wrote the case report. Dr CV and Professor GB revised the paper. All authors agreed the final version of the manuscript for publication.

Competing interests

No competing interests were disclosed.

Grant information

The author(s) declared that no grants were involved in supporting this work.

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