- cultural growth of the population;
- the ability of the population to be informed,
- the opportunity to choose the doctor and service he wants, public or private healthcare system.

In the future, this trend will have the necessary ground to be furtherly improved.

Preeclampsia remains a disease which requires many actresses and factors to improve the results in its treatment. At the local level, it would also be beneficial for the treatment of the preeclampsia to organize a reference system.

In this way, they would find a more accurate and more relevant response to any obstetric service, in the following questions:

A. Which type of patients do we have to treat? (Every service should be determined based on the results and the possibilities it has, to what extent a patient should be treated by him. This assessment should be made for the mother, but mainly for the future of fetal maternity.
B. Each service should determine what level of complications a pregnant patient should be transferred.
C. The third step in the referral system is where to transfer a patient. Generally, transference is performed in a parallel or higher level service.
D. Finally, how should transportation be performed and whose responsibility is it. This transfer is related to the selection of the staff that will accompany the patient and the means of transport. In a preliminary agreement, transportation may be the responsibility of the hospital where the patient or the hospital he/she will go to. Here is not excluded the possibility of personal car transports.

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Coexistence of Psychiatric Symptoms and Chiari Type I Malformation - A Case Report

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Abstract

Introduction: *Chiari type I malformation has been described infrequently in association with defined psychiatric syndromes.* ***Method:*** *There is a limited literature about obsessions in comorbidity with Chiari malformation. It is described a case of an adolescent with obsessive compulsive disorder and Chiari I malformation and it is reviewed the literature regarding Chiari I malformation and psychiatric disorders. The child came to the attention of child psychiatrist at the age of 7 years old when he manifested developmental delay and various kinds of behavioral symptoms. He was followed up at the age of fifteen when he developed obsessions and upon MRI was identified a Chiari malformation type I.* ***Discussion:*** *This paper discusses the likely under recognized co-occurrence of Chiari malformation and psychiatric symptoms. Currently available data from case reports associate Chiari I malformation with a variability of psychiatric symptoms: autism spectrum disorder, bipolar disorder, seizures, developmental delay, generalized anxiety disorder, panic attacks, Tourette's syndrome, OCD, ADHD, cognitive disorder NOS and psychosis. It is concluded that mixed psychiatric symptoms and developmental delay might be a more common finding in comorbidity with Arnold Chiari type I malformation.*

Keywords*: case report; obsessions; Arnold Chiari type I malformation; coexistence*

Introduction

Various types of congenital lesion are associated with neuropsychological impairments and behavioral changes (Riva et al., 2011). Chiari I malformations are poorly understood and in need of much greater systematic investigation that has been the case up until now. The brain regions involved in this condition are highly sensitive and can easily contribute to deficits in the development of self-regulation or executive function (Koziol et al., 2013).

This paper discusses the co-occurrence of Chiari type I malformation and psychiatric symptoms. Arnold Chiari malformation is characterized by four subtypes with different degrees of herniation of the cerebellar structures through the foramen magnum. The Chiari I malformation is defined as herniation of the cerebellar tonsils through the foramen magnum (at least 3 to 5 mm). In the type I malformation there is a caudal descent of the cerebellar tonsils only, while in type II the cerebellar vermis and possibly the fourth ventricle and pons are involved (Chisholm et al., 1993). Chiari III and IV malformations are rare. Type III is characterized by an encephalocele, the descent of both cerebellum and brainstem into the spine and internal sac and type IV is associated with cerebellar atrophy. Although Chiari I malformation is considered to derive from a mesodermal disorder resulting in underdevelopment of the posterior fossa relative to its content, evidence for a possible heterogeneous etiology also has been reported (Grosso et al., 2001).

The cerebellum was once believed to be almost exclusively involved in coordinated voluntary movement. Currently the role of cerebellum in the modulation of the higher cognitive functions is becoming increasingly clear. Numerous studies on adults have confirmed that the cerebellum has a role in processing higher brain functions as intelligence; language; higher social functions; perceptual, language-related; visual-spatial and cognitive & affective functions, procedural and declarative memory and evidence of this role has emerged more recently in developmental age as well. In Chiari malformation, the cerebellar structures are squeezed and crowded inside the posterior fossa and along the time this could generate various kinds of cognitive and behavioral disorders (Riva et al. 2011). Therefore it is hypothesized that the compression of the structure in the posterior fossa could negatively affect how these cerebellar structures function. Chiari malformation can also cause cortical deficit. Chiari malformations cause obstruction to cerebrospinal fluid flow in the posterior fossa and foramen magnum that elevates cranial pressure and this can cause hydrocephaly. It can damage neural tissue by ischemic and mechanical forces. Gonzales and Campa-Santamarina (2018) provided evidence of possible deficits or anomalies in the cognitive executive functions of patients with Chiari type I. They were affected in the processes of inhibition and self-control as well as in attention

capacity and maintaining a course of thought and action. According to Koziol and Barker (2013) patients with Chiari I have behavioral disorders with impairments in executive functions, verbal fluency, abstract thinking and working memory.

Method

There is a limited literature about psychiatric symptoms with Chiari malformation, and to my knowledge two cases have been reported of obsessions in association with this brain anomaly (Tubbs et al., 2003; Zayman and Erbay, 2016). This paper discusses the likely under recognized co-occurrence of Chiari I malformation and psychiatric symptoms. It is described the case of a child who has obsessions and Chiari I malformation and the review of the literature regarding Chiari I malformation & other psychiatric disorders.

Case Description

M. is a fifteen years old male who lives with both parents. He is the only child in the family. Pregnancy and birth were unremarkable, except a subcutaneous head hematoma.

Mother recalled that during infancy M. had sleep disturbances (late falling asleep, 1-2 a.m. during the first three years of life). When M. started walking he was a hyperactive toddler, running without direction. As a toddler he was inpatient and sometimes aggressive in kindergarten. He also had language delay and started to talk at about 5 years old.

The child came to the attention of child psychiatrist at the age of seven years old, when he manifested developmental delay and various kinds of behavioral symptoms. The teacher at primary school noted that M. was easily distracted by noises and showed academic difficulties. He spoked clearly, except some pronunciation difficulties related to specific consonants. He could read, but had difficulty to retell, which might suggest memory problems. He showed poor math abilities. M. was consulted by the school psychologist. It was administered Son-R test which reported mental retardation (chronological age of 7 years and 8 months, mental age 6 years old). M. displayed some autistic features. In respect to social interaction he interacted with peers, but became easily nervous and sometimes was aggressive toward them. M. didn't obey to the rules of play and insisted that other children should met his rules, so he couldn't enjoy the play and as the consequence it was easily interrupted. M. worn shoes on the opposite foot until he was 11 years old. He had short eye contact span and was often inpatient. M. was referred to the child psychiatrist who suspected hyperactivity. At that time M.

was also showing some unusual interest which could have been autistic traits or precursor of obsessive symptoms. He liked very much the topic war and soldiers, played with soldier figures and while having a conversation with children liked to switch inappropriately the topic of the conversation to war and soldiers. Parents attributed this unusual interest to the TV films with war themes he saw, despite they noticed it was incoherent.

Teenager years marked new and more severe symptoms. Obsessive compulsive disorder symptoms started when he was 12 years old and persisted six years later. The main type of obsessive thoughts were aggressive, followed by sexual ones which were hardly explored. M. at the age of fifteen had thoughts of harming with a knife the peers who bully him. He still was verbally aggressive to peers who bullied him. M. also had thoughts about harming his parents: with a knife, burning them or causing accidents (for example he had the urge to move the step while the father was climbing in the terrace, so he could fall). He felt bad about these ideas for his loved parents and tried to resist his thoughts – kept himself busy, when he was close to a knife he moved backwards or asked his mother to take it away. The mother described her son as childish, immature and had a feeling like she was talking with a child despite him being an adolescent.

The child was referred to several doctors following a care path from the school social worker and school psychologist at the age of 7 years old to the child psychiatrist. At the age of 13 years old was performed a brain Magnetic resonance imaging that identified a type I Chiari malformation (cerebellar tonsils were 7 mm under the foramen magnum).

Family history was positive for his maternal ancestry, with an anxiety disorder in his mother, a mood disorder in his maternal grandmother, his uncle has been hospitalized for schizophrenia and two cousins (uncle's daughter and aunt's son) also suffered mental problems.

M. received over more than 10 years several prescriptions of Concerta at the age of 7 years and a half, Atomoxetine and Risperidal with no improvement. At the age of 15 years old was prescribed sertraline and M. showed moderate improvement. His Obsessive compulsive disorder continued so far at the age of 18 years old. He did not underwent surgery for Chiari I malformation.

Literature review

Association of Chiari I malformation with psychiatric syndromes

Chiari type I malformation has been described infrequently in association with defined psychiatric syndromes. There are some case reports that highlight the

association of Arnold–Chiari malformation (ACM) with psychiatric symptoms. Bakim et al. (2013) assessed the association between Arnold Chiari malformation and psychiatric symptoms and risk factors in terms of psychiatric morbidity. They found out that about 43.8% of the patients had a psychiatric disorder. There are various reports of mental symptoms in children and adolescents associated with Chiari Malformation type I ranging from developmental delay to a more specific disorder, during the last 20 years (Figure 1).

FIGURE 1: Association of Arnold Chiari I malformation & psychiatric syndromes

Brill et al. (1997) reported on 11 children with Chiari I malformation who presented with seizures and developmental delay in motor or language function with or without autistic features. An association between Chiari I malformation and seizures or neurodevelopmental deficits or both had not been previously reported to their knowledge up to that time. They believed that Chiari I malformation should not be considered an incidental finding in these patients, but may be a marker for subtle cerebral dysgenesis. Chiari I and II malformations may constitute a complex but continuous spectrum, related to the timing and severity of a shared underlying embryologic mechanism. Autism cases were also bought into attention by Jayarao et al. (2015), who reported on 9 children with Autism spectrum disorder and Chiari

I malformation in a sample of 125 children. They stated that Chiari I malformation and Autism Spectrum Disorder may coexist and be under recognized.

Grosso et al. (2001) aimed to elucidate the relationship between Chiari I malformation and mental retardation, speech delay, and epilepsy to consider a possible specific pathogenetic background. They had a sample of 35 patients with Chiari type I malformation. Out of them nine patients (four boys and five girls) were affected by mental retardation, speech delay and epilepsy. They argued that the association of Chiari I malformation with epilepsy, speech delay, and mental retardation may not be a mere incidental finding but may be a marker for a different pathogenetic background.

Zeegers et al. (2006) evaluated the prevalence of brain abnormalities (MR study) in a group of 45 young children (mean age 43 months, SD=12, four females) with developmental disorders, specifically including children that came to the attention of a child psychiatrist before the age of 3 years. They found out about 50% intracranial abnormalities in this sample (22 children). One female was diagnosed with a Chiari I malformation. They argued that radiological findings do not contribute to the diagnosis of developmental disorders. However, young children with developmental disorders may not be able to express discomfort associated with brain abnormalities, such as a Chiari I malformation.

Riva et al. (2011) studied 35 children (mean age 6 and half years) of whom 20 children had associated conditions and symptoms and 15 children represented the control group. They had two cases with Chiari malformation and interestingly reported their behavioral changes after surgery. One case was that of a 5 year old boy with a history of language delay and behavioral issues. The child underwent surgery at the age of 3 and a 3 months. After surgery the child language improved, he showed increase in the verbal IQ measured by the Test of language assessment. On the other hand his behavioral issues which included behavioral difficulties, attentional and motor instability became worse than before surgery. The second case was that of a 15 year old girl who underwent surgery at 11 years. The girl's pattern of recovery took an opposite course. Before surgery her behavior had been characterized by mild hyperactivity and she was easily distracted, but after decompression she behaved normally. While her attentional instability improved considerably, her language skills became worse after surgery.

There are also several single case reports. For example Tubbs et al. (2003) described a 13 year old adolescent with Chiari I malformation and cutis marmorata telangiectatica congenita, who also showed Tourette's syndrome, obsessive-compulsive disorder, and seizures. Ciprero et al. (2005) reported three children with Kabuki syndrome who had Chiari I malformation. Kabuki (Niikawa-Kuroki) syndrome is associated with a characteristic facial appearance, cleft palate, congenital heart defects, and developmental delay. A more detailed

review of studies and cases with Chiari I malformation and psychiatric disorders is summarized in Table 1 as follows:

TABLE 1: Literature cases with Chiari I malformation and psychiatric disorders

Psychiatric disorders	Author	Year	Sample	Cases with co-occurrence of Chiari I and psychiatric disorders
Obsessive-compulsive disorder	Zayman and Erbay	2016		case report
Schizophrenia, obsessive ideas	Di Genova et al.	2015		case report
Autism spectrum disorder	Jayarao M. et al.	2015	n=125	n=9
Psychotic disorder, depersonalization/ derealization, major neurocognitive disorder	Hoederath L. et al.	2014		case report
Psychiatric disorder (various)	Bakim B. et al.	2013	n=16	n=7
Psychosis and panic attack disorder	Casale A. et al.	2012		case report
Language delay, mild hyperactivity and attentional instability	Riva D. et al.	2011	n=35	two case reports
Panic disorder and agoraphobia	Kuloglu M. et al.	2009		case report
ADHD, OCD and bipolar disorder	Koziol L. F. & Budding D. E.	2009		case report
Generalized anxiety disorder	Caykoylu A. et al.	2008		case report
Cognitive Disorder NOS	Pearce M. et al.	2006		case report
Developmental disorders	Zeegers M. et al.	2006	n=45	n=1
Psychotic events and epilepsy	Ilanković N. N. et al.	2006		case report
Kabuki syndrome (developmental delay)	Ciprero KL. et al.	2005		n=3
Tourette's syndrome, OCD and seizures	Tubbs RS et al.	2003		case report
Mental retardation, speech delay, and epilepsy	Grosso S. et al.	2001	n=35	n=9
Seizures and developmental delay in motor or language function with or without autistic features	Brill C. B. et al.	1997		n=11
Panic disorder with agarophobia	Chisholm et al.	1993		case report

Discussion And Conclusions

The literature review on co-occurrence of Chiari malformation and psychiatric symptoms shows that some studies report this association, but currently data

remain inconclusive. This case illustrates and support comorbidity of psychiatric symptoms and Chiari I malformation. However, prospective longitudinal studies on sizable series will be needed to ascertain whether and to what degree Chiari malformations may negatively affect mental functioning in developmental age.

The literature data shows a variability of psychiatric symptoms associated with Chiari I malformation, not merely a specific clinical picture. This case represent several psychiatric symptoms in a patient who has Arnold type I malformation, more prominent ones are obsessions, autistic features and developmental delay. The majority of cases show that Chiari malformation and anxiety disorder can exist together. To date, two case reports described the association with obsessions, but there were more cases reports which described the association with developmental delay and mixed psychiatric symptoms. It is concluded that mixed psychiatric symptoms and developmental delay as a common one might be a more common finding in comorbidity with Arnold Chiari type I malformation.

In the case described here it is reported the association, not the cause. It is possible that in this patient both conditions are separate unrelated pathological events. On the other hand, Chiari type I malformation could have made the patient susceptible to psychiatric symptoms. One might speculate that the neuroanatomical anomaly led to symptoms in a patient who was predisposed. A family history of psychiatric disorders suggests a genetic predisposition in this case. However none of the family members was investigated for brain anomalies. Increasing evidence suggests that in some families, there are strong genetic contributors to the development of Chiari malformation type I (Szewka et al., 2006). This case highlight the importance of not focusing only on psychiatric aspects, but considering a neuro-radiological investigation which can detect a lesion that might otherwise go undetected. It is important to consider an eventual organic etiology in a child while challenging a clinical picture with mixed symptomatology and developmental delay.

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