



Dandy walker syndrome with unusual presentation and unusual radiologic sign on Ct-Scan

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Abstract

Background: Dandy-Walker syndrome is a rare heterogeneous congenital disorder characterized by a triad of cystic dilatation of the fourth ventricle, complete or partial agenesis of the cerebellar vermis and an enlarged posterior fossa. Most patients develop hydrocephalus with increased head size and variable degree of mental retardation, and my develop spasticity and seizures. The syndrome has not been well-documented in Iraq. The aim of this paper is to report the unusual presentation of the syndrome with a presentation like ataxic cerebral palsy with mental retardation without increase in head size because of hydrocephalus. The patient also had bilateral pachygyria more on the left side on brain CT-scan which is also unusual radiologic sign in this condition.

Results: The boy was hypotonic since early infancy and experienced delayed development, unsteadiness and uncoordinated movements, and poor feeding. At the age of five years, he had ataxic movements, and was unable to sit unsupported, unable to feed self, and had no bowel control. He was not saying any word. He had poor cognitive abilities and did not understand simple commands. He was hypotonic and had normal tendon reflexes. Family history was negative for simillar illness. Brain CT-scan showed bilateral Pachygyria more on the left side in addition to the radiologic signs of Dandy-Walker syndrome.

Patients and methods: A five year boy was referred with a clinical diagnosis of ataxic cerebral palsy and mental retardation was studied at the pediatric neuropsychiatry clinic of the Children Teaching Hospital of Baghdad Medical City during the year 2019.

Conclusion: The of Dandy-Walker syndrome in Iraqi boy with unusual presentation, a presentation like ataxic cerebral palsy with mental retardation without increase in head size because of hydrocephalus is reported. In addition, the patient also had bilateral pachygyria more on the left side on brain CT-scan which is also unusual radiologic sign in this condition.

Keywords: Dandy-Walker syndrome, unusual presentation, pachygyria, brain CT-scan

Introduction

The brain ventricular system consists of four communicated fluid-filled cavities including left and right lateral ventricles which represent the cavity of the cerebral hemisphere, the third ventricle, and the fourth ventricle. The fourth ventricle is located within the pons or in the upper part of the medulla oblongata. CSF entering the fourth ventricle through the cerebral aqueduct can exit to the subarachnoid space of the spinal cord through two lateral apertures and a single, midline median aperture. Dandy-Walker syndrome is a rare heterogeneous congenital disorder characterized by a triad of cystic dilatation of the fourth ventricle, complete or partial agenesis of the cerebellar vermis and an enlarged posterior fossa. The disorder presents with variable of neurological features. The disorder is typically associated with lifelong disability of variable severity. Most patients develop hydrocephalus with increased head size and variable degree of mental retardation, and my develop spasticity and seizures [1-11]. The syndrome has not been well-documented in Iraq. The aim of this paper is to report the unusual presentation of the syndrome with a presentation like ataxic cerebral palsy with mental retardation without increase in head size because of hydrocephalus. The patient also had bilateral pachygyria more on the left side on brain CT-scan which is also unusual radiologic sign in this condition.

Patients and methods

A five year boy was referred with a clinical diagnosis of ataxic cerebral palsy and mental retardation was studied at the pediatric neuropsychiatry clinic of the Children Teaching Hospital of Baghdad Medical City during the year 2019.

Results

The boy was hypotonic since early infancy and experienced delayed development, unsteadiness and uncoordinated movements, and poor feeding. At the age of five years (Figure-1), he had ataxic movements, and was unable to sit unsupported, unable to feed self, and had no bowel control. He was not saying any word. He had poor cognitive abilities and did not understand simple commands. He was hypotonic and had normal tendon reflexes. Family history was negative for simillar illness.

Brain CT-scan showed

Posterior fossa filled with cerebrospinal fluid and fourth ventricle dilated and causing compression of the cerebellum. Absent inferior vermis.

Cerebellar atrophy

Bilateral pachygyria more on the left side. Normal supratentorial ventricular system.



Fig 1: At the age of five years, the boy was unable to sit unsupported

Discussion

The condition was recognized during the 19th century by English surgeon John Bland-Sutton in 1887 who described the condition as hypoplasia of the cerebellar vermis, an enlarged posterior fossa and hydrocephalus. Thereafter, it was also reported by Fusari [Fusari R. *Mem Accad Sci Inst Bologna Ser* 1891-92; 5 (2): 643] and Rossi who reported two cases [Rossi U. *Sperimentale (Mem.)* 1891; 45, 518] ^[3, 12].

Early during the 20th century the condition was described in 1911 by Woskressenski [Woskressenski S. *Z ges Neurol Psychiat (Orig.)* 1911; 6, 38] ^[3]. Shortly later, the condition was described by American neurosurgeon Walter Dandy and American pediatrician Kenneth Blackfan in 1914 ^[1]. Thereafter, in 1942, the condition was also described by American physician John K Taggart and Canadian-American neurosurgeon Arthur Earl Walker attributed the disorder to underdevelopment of the foramina of Luschka and Magendie ^[13]. A German psychiatrist Clemens Ernst Benda called the condition Dandy-Walker malformation in 1954 ^[2].

Macrocephaly caused by hydrocephalus is the most common clinical feature, affecting 90 to 100% of patients during their first months of life, and most patients have evidence of raised intracranial pressure during the first year of life. Kumar, Jain, and Chhabra (2001) studied 42 patients with Dandy-Walker syndrome and found hydrocephalus in all patients at the time of diagnosis, vermian hypoplasia in 88%, and cerebellar hypoplasia in 59% ^[14, 15, 16].

Conclusion

The of Dandy-Walker syndrome in Iraqi boy with unusual presentation, a presentation like ataxic cerebral palsy with mental retardation without increase in head size because of hydrocephalus is reported. In addition, the patient also had bilateral pachygyria more on the left side on brain CT-scan which is also unusual radiologic sign in this condition.

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