Incidental Presentation of Dandy Walker Variant in 66 Year Male Patient

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Abstract

Dandy-Walker variant consists of vermian hypoplasia and cystic dilatation of the fourth ventricle, without enlargement of the posterior fossa is a distinctive entity believed to represent a mild subtype of Dandy-Walker complex. We report a case of 66 year male presented with right sided hemiparesis due to ischemic stroke whose imaging showed incidental findings Dandy walker variant. This Incidental Dandy Walker malformation finding in adult is rare with only a few cases reported till date.

Introduction

Dandy-Walker syndrome (DWS) is a rare autosomal dominant condition involving posterior fossa malformation, characterized by aplasia or hypoplasia of the cerebellar vermis, presence of a membranous cyst of the fourth ventricle; and commonly hydrocephalus. It is usually observed during the prenatal period or early infancy and very rarely in adults. The aim of our case report is to reveal the asymptomatic nature of Dandy Walker syndrome in 66 year male patient who came with ischemic stroke and had not shown any cerebellar and hydrocephalus signs and led normal life till ischemic stroke occurred which is unique and rare.

Case Report

A 66 Year male patient presented in emergency department in December 2014 with complains of sudden onset weakness of right side of body and slurring of speech.Patient was a known case of systemic hypertension since 1 year taking regular medication and chronic alcoholic since 30 year but he had stopped alcohol consumption since last 4 years. He had normal developmental history.

On general examination pallor was present. Blood pressure was 168/84 mm of Hg.

Neurologically patient was conscious well oriented and alert. Deviation of angle of mouth to left side, slurring of speech was present. Motor examination revealed weakness on right side of face (UMN Type), upper limb and lower limb (Grade 2/5). Increased tone and hyperreflexia present in right upper limb and lower limb. Babinski sign (Plantar reflex) was positive on right side. Sensory system and other cranial nerves examination was unremarkable. Cerebellar and meningeal signs were absent. Frontal bossing or macrocephaly was absent. Carotid bruit was absent. Other system examination was normal. Patient had no psychiatric complains and hearing defect.

On investigation, complete blood count showed Hb-12.3 (Normocytic normochromic). Lipid profile –Serum cholesterol 204 mg/dl, HDL- 65 mg/dl, LDL- 83 mg/dl. Blood chemistry, liver function test, urine examination was normal. Fundoscopy did not reveal any significant abnormality. CSF examination was also normal.

CT SCAN suggested:

1. Diffuse white matter hypodensity in bilateral fronto-occipital projections, periventricular deep white matter, corona radiata, centrum semiovale, suggestive of acute infarct and mild age related cerebral and cerebellar atrophy.

2. Mildly dilated both lateral ventricle, 3rd and 4th ventricle suggestive of communicating hydrocephalus.

3. Giant cistern magna appears to be communicating with IV ventricle giving “Key hole appearance.”

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Inferior cerebellar vermis appears hypoplastic and Mild communicating hydrocephalus, above finding suggestive of possibility of Dandy Walker variant.

Immediately MRI BRAIN was done to confirm above incidental finding and Dandy Walker variant was confirmed.

Patient was treated with antiplatelet and antihypertensive agents and supportive management including physiotherapy and patient improved and subsequently discharged after 12 days of hospitalisation. Patient came for regular follow up and still did not reveal cerebellar or hydrocephalus signs till date.

Discussion

The Dandy-Walker syndrome (DWS) is a rare posterior fossa malformation, characterized by aplasia or hypoplasia of the cerebellar vermis, involving the cortex and deep cerebellar nuclei; the presence of a membranous cyst of the fourth ventricle; and, commonly, hydrocephalus.1,2 This malformation was first described by Dandy and Blackfan (1914). Taggart and Walker (1942) revised this malformation characterizing the main clinical and pathologic features. Benda (1954) finally labelled the disease as “Dandy-Walker syndrome”.

The DWS is a very rare autosomal dominant congenital malformation of the posterior fossa, with an incidence ranging between 1/25000 and 1/35000 births, usually observed during the prenatal period or early infancy and very rarely in adults.1,2 DWM is thought to develop between the 7th and the 10th week of gestation; early descriptions of the syndrome propose that the main pathogenic mechanism was the failure of the foramen of Magendie and Luschka to open through embryonic life, specifically at the end of the 4th foetal month.

The DWS has been frequently associated with trisomies 3q, 6p, 9p, 11 or 22, and more rarely with chromosome translocations. More recently, a first critical region associated with DWS, encompassing two adjacent Zinc finger in cerebellum genes, ZIC1 and ZIC4 has been identified.

Three main types of Dandy Walker Complex are
1. Dandy walker malformation
2. Dandy walker variant
3. Mega cisterna magna

It is usually diagnosed at birth or in early childhood. Adult presentation of the Dandy-Walker syndrome is extremely rare.3,4 The extensive search of literature revealed only few case reports where patient never developed evidence of cerebellar involvement till adulthood and Dandy-Walker syndrome was an incidental finding when being investigated for other disability.5,6 Syndromes associated with Dandy walker malformation include “PHACE syndrome” (posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of aorta and cardiac defects and eye abnormalities) and Ellis-van Creveld syndrome.

The aim of our case report is to reveal the asymptomatic nature of Dandy Walker syndrome in 66 year male patient who came with ischemic stroke. He had not shown any cerebellar and hydrocephalus signs and led normal life till ischemic stroke occurred which is unique and rare.

References