CASE REPORT

# Asymptomatic Dandy Walker malformation in an elderly male with acute haemorrhagic stroke: A case report

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## Abstract

Dandy-Walker Malformation (DWM) is a rare congenital anomaly of the posterior cranial fossa. Features of DWM include hypoplasia of the cerebellar vermis, enlargement of the posterior fossa, and cystic dilatation of the fourth ventricle. MRI is the modality to confirm the diagnosis. Treatment is usually symptomatic and required when signs of hydrocephalus develop. Rare cases of asymptomatic DWM diagnosed incidentally are reported in literature. We report a case of DWM in a 60-year-old male who presented with haemorrhagic stroke and was later found to have DWM on brain imaging.

**Keywords:** Dandy-Walker Malformation, Haemorrhagic Stroke.

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## Introduction

Dandy-Walker Malformation (DMW) is a rare congenital anomaly of the posterior fossa with a frequency of 1:25000 to 1:35000 live births in the United States. It is characterised by hypoplasia of the cerebellar vermis, enlarged posterior fossa, and cystic dilatation of the fourth ventricle, displacing the torcula and tentorium upwards.[1] It usually presents in children before the age of 1 year. The most common manifestation is macrocephaly. Other features are vomiting, nystagmus, ataxia, features of hydrocephalus, and bulging occiput.[2,3]

Rare cases of asymptomatic DWM diagnosed incidentally in adults are reported in the literature.[4] Stroke is an extremely rare presentation of DWM, with only one case of haemorrhagic stroke reported in the literature.[1] Here, we have a case of a 60-year-old male presenting with a haemorrhagic stroke who was incidentally diagnosed with DWM based on imaging. To the best of our knowledge, this is the second reported case of an elderly male with DWM in the literature presenting as haemorrhagic stroke who had no symptoms of the disease. This is the first reported case of incidental DWM in an elderly male from Pakistan.

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## Case Report

A 60-year-old male was brought to the emergency department of Civil Hospital Karachi on October 2021 with complaints of sudden onset of weakness and numbness of the right side of the body and slurring of speech for one day. He was right-handed and a tailor by profession. His past medical history was remarkable for asthma and hypertension. His asthma was well controlled on medications but hypertension was poorly controlled due to non-compliance. The patient denied any history of frequent falls, fits, gait changes, tremors, and speaking difficulty in the past.

On general physical examination, he was oriented to time, place, and person, and his GCS was 15/15. His blood pressure was 190/110 mmHg, his pulse rate was 99 beats per minute, his respiratory rate was 22 breaths per minute, and his temperature was 98°F.

On neurological examination, cranial nerves were intact. The bulk was decreased on all four limbs, and the tone was normal. Power was 4/5 of the right upper and lower limbs and 5/5 of the left upper and lower extremities. Sensory examination showed decreased pinprick sensation and fine touch on the right side. The reflexes were normal, and plantars were down going bilaterally. Cerebellar signs could not be assessed because of motor and sensory weakness. The patient was admitted to the ward and was given intravenous labetalol to control blood pressure. The head end of the bed was elevated to reduce intracranial pressure. His blood pressure was being monitored continuously. He did not develop asthma exacerbation during the course of his hospital stay and remained vitally stable.

Computed tomography (CT) scan (Figure 1) was done that showed a hyperdense lesion in the left thalamic area, consistent with haemorrhagic stroke. Other findings included asymmetrical enlargement of the lateral ventricles but temporal horns and periventricular leukodensities were not visible and cortical sulci were not effaced, suggesting hydrocephalus ex vacuo secondary to age-related cortical

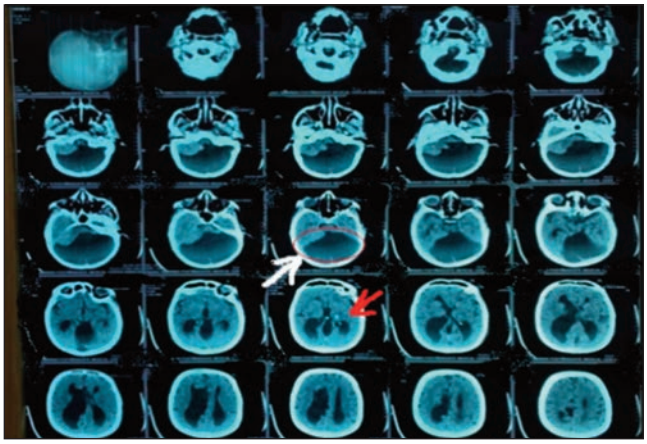

**Figure-1:** CT scan plain axial view showing intracranial bleed in the left thalamic region (red arrow). The posterior fossa shows cystic dilatation (white arrow and red ccircle).

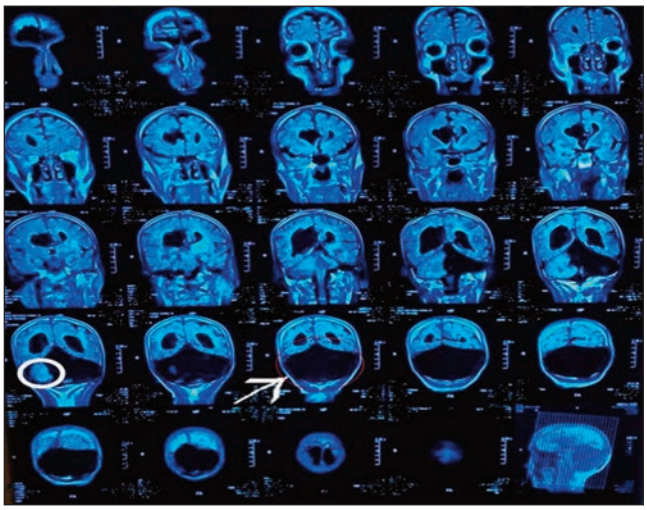

**Figure-2:** MRI showing cystic dilatation in the posterior fossa communicating with the 4th ventricle (white arrow), and cerebellar hypoplasia (white circle) consistent with Dandy Walker Malformation (DWM).

atrophy. There was an incidental finding of cystic enlargement of the posterior fossa suggestive of Dandy-Walker Malformation. Differential diagnoses included Blake's cyst, mega cisterna magna, enlarged 4th ventricle cyst, and retrocerebellar arachnoid cyst. Magnetic resonance imaging (MRI) (Figure 2) was done that confirmed the diagnosis of Dandy-Walker Malformation.

The patient's sensory system abnormalities were due to thalamic haemorrhage. The weakness in limbs could be due to oedema around the haemorrhage affecting the internal capsule.

The patient was managed in the hospital with antihypertensives and physiotherapy. He recovered completely and was discharged. On follow-up after 3 weeks, his power in all four limbs was 5/5, and there was no sensory deficit. Neurological examination showed the absence of cerebellar signs such as dysdiadochokinesia, dysmetria, limb ataxia, gait abnormalities, nystagmus, rebound phenomenon, scanning speech, and pendular knee jerk. Follow-up after 2-weeks was unremarkable for any neurological deficit, and the patient was compliant with medications. Verbal consent was taken from the patient to publish this report.

## Discussion

Dandy-Walker Malformation was first described by Dandy and Black fan in 1914. In 1954, Benda used the term Dandy-Walker Malformation for the first time.[5] Dandy-Walker complex is a broad term that includes Dandy-Walker Malformation, DW variant, and mega cisterna magna. Dandy-Walker Malformation comprises enlarged posterior fossa, cystic dilation of the fourth ventricle, and hypoplasia of the cerebellum. Mega cisterna magna describes the enlargement of cisterna magna leading to an enlarged posterior fossa. The DW variant is characterised by hypoplasia of the cerebellar vermis and cystic dilation of the fourth ventricle, but the posterior fossa is not enlarged.[5] DWM is more commonly reported in females. The aetiology of DWM is multifactorial and includes genetic causes, congenital infections, trauma, and syndromic associations.[6] The association between haemorrhagic stroke and DWM is not clear. One plausible explanation is that the hydrocephalus that develops in DWM due to 4th ventricle distention along with hypertension, stresses the cerebral vessels making them prone to rupture.[1]

We describe the case of a 60-year-old male with haemorrhagic stroke who was incidentally found to have DWM on imaging. The patient had no symptoms of the disease and led a regular life. The MRI showed cerebellar vermis hypoplasia with cystic enlargement of the posterior fossa communicating with the fourth ventricle. Our case is unique because it is the first case of DWM presenting with a haemorrhagic stroke in an old-age male. Contrary to a similar case described by Hallem,[1] our patient recovered completely and developed no complications during the hospital stay. This patient did not develop hydrocephalus and thus did not require any treatment for the anomaly.

Hallem and Joseph reported a case of an undiagnosed asymptomatic Dandy-Walker Malformation in a 34-year-old male presenting with haemorrhagic stroke. The patient had obstructive hydrocephalus and was treated with right frontal ventriculostomy and external ventricular drain. Unfortunately, the patient died due to acute respiratory distress syndrome.[1]

Rakesh reported a case of incidental finding of the Dandy-Walker variant in a 66-year-old male with an ischaemic

stroke. The patient had no symptoms of hydrocephalus and cerebellar involvement.[7]

C Tanya and Bernardo described a case of a 56-year-old woman who presented with diplopia, headache and vomiting. Her examination and history were consistent with an acute brainstem infarct. In addition, MRI showed the incidental finding of underlying DWM.[8]

MRI is crucial for the diagnosis of Dandy-Walker Malformation. The differential diagnosis of DWM includes persistent Blake's cyst, mega cisterna magna, and retro-cerebellar arachnoid cysts. These disorders can be differentiated on the basis of imaging. The treatment of DWM is aimed at controlling hydrocephalus.[6] The treatment options include ventriculoperitoneal or cyst-peritoneal shunts and endoscopic third ventriculostomy.[9] Prognosis depends on the associated anomalies and degree of hydrocephalus. The patient in our case had no associated hydrocephalus and was discharged on antihypertensive medications, with recommended follow-up.

## Conclusion

Although commonly diagnosed before the age of 1 year, DWM can be asymptomatic in an otherwise healthy patient. We report a case of an elderly male with incidentally diagnosed DWM on imaging. Our patient did not require any treatment because of asymptomatic nature of the anomaly. Symptoms of hydrocephalus were evaluated on routine follow-up.

**Disclaimer:** The ethical approval for publishing this case report was taken from the head of department who is also an author of this report.

**Conflict of interest:** The ethical approval was signed by the author of this study.

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