

DANDY WALKER MALFORMATION IN AN ASYMPTOMATIC PATIENT

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ABSTRACT

Dandy-Walker malformation is a rare congenital malformation that involves the cerebellum and fourth ventricle. Symptoms, which often occur in early infancy include developmental delay, enlarged head circumference, or signs and symptoms of hydrocephalus. We present the case of a 70 year old female who has led a normal life with an incidental Dandy Walker malformation discovery.

Key words: Dandy Walker malformation, asymptomatic adult patient

BACKGROUND

Dandy-Walker malformation is characterized by agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle, and enlargement of the posterior fossa. It may be associated with atresia of the foramen of Magendie and, possibly, the foramen of Luschka (1).

Dandy and Blackfan described for the first time the Dandy-Walker malformation in 1914 (2). It was not until 1963 that studies by D'Agostino in 1963 and Hart et al in 1972 defined the characteristic triad of Dandy-Walker malformation as consisting of: complete or partial agenesis of the vermis, cystic dilatation of the fourth ventricle, and an enlarged posterior fossa with upward displacement of lateral sinuses, tentorium, and torcular herophili (3). Approximately 90% of patients have hydrocephalus, and a significant number of children have associated anomalies. Infants present with a rapid increase in head size and a prominent occiput. Most children have evidence of long tract signs, cerebellar ataxia, and delayed motor and cognitive milestones, probably due to the associated structural anomalies (4).

Classically, posterior fossa cystic malformations have been divided into Dandy-Walker malforma-

tion, Dandy-Walker variant, mega cisterna magna, and posterior fossa arachnoid cyst. Precisely differentiating the malformations may not be possible using imaging studies. Dandy-Walker malformation, variant, and mega cisterna magna are currently believed to represent a continuum of developmental anomalies on a spectrum that has been termed the Dandy-Walker complex (5).

CASE STUDY

We present the case of a 70 year old female admitted in the Neurology Department of Emergency University Hospital for complaints of cognitive impairment and depression.

Neuropsychological testing reveals major depressive disorder and mild cognitive impairment with severe short-term memory loss and important drop in long-term storage with a MMSE score of 29 out of 30 pts. A physical examination was performed as part of the general evaluation to determine whether conditions capable of causing MCI are present; the clinical examination shows no neurological deficits.

Because multiple etiologies can be incriminated in a complaint of cognitive impairment an organic brain disorder was suspected. We performed a non

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contrast CT scan which revealed large cystic lesion occupying most of the posterior fossa (measuring 60/112mm) extending into a dilated fourth ventricle, with vermian and cerebellar hypoplasia predominantly on the left side and supratentorial ventriculomegaly. Cerebelli tentorium is displaced upward. We completed the evaluation with cerebral and cervical MRI scan. It shows no encephalic or spinal cord signal abnormalities with minimal disc protrusions and confirmed the diagnosis of Dandy Walker malformation.

Incidental Dandy Walker malformation finding in adult is extremely rare with only a few cases reported (6) (7). This case is particular because of the oldest age of presentation reported so far in an asymptomatic patient.

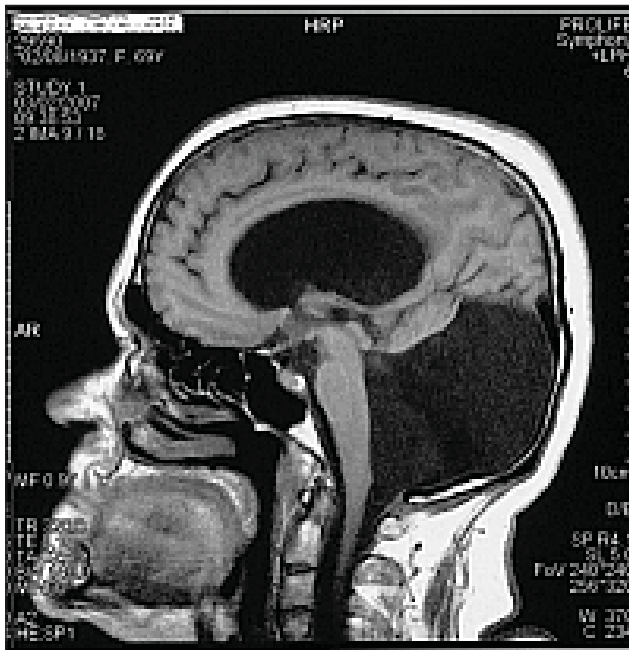


FIGURE 1

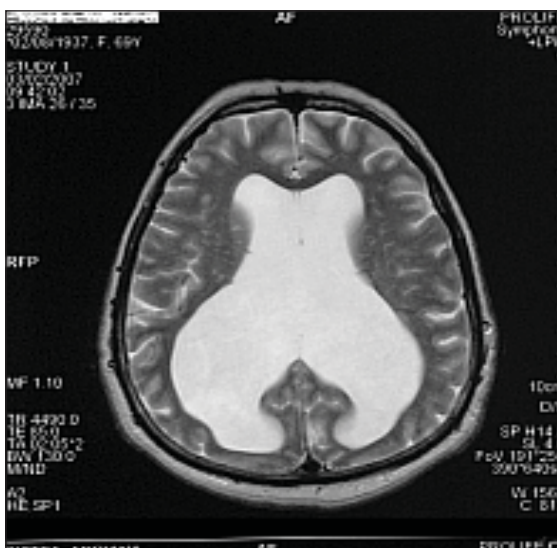


FIGURE 2

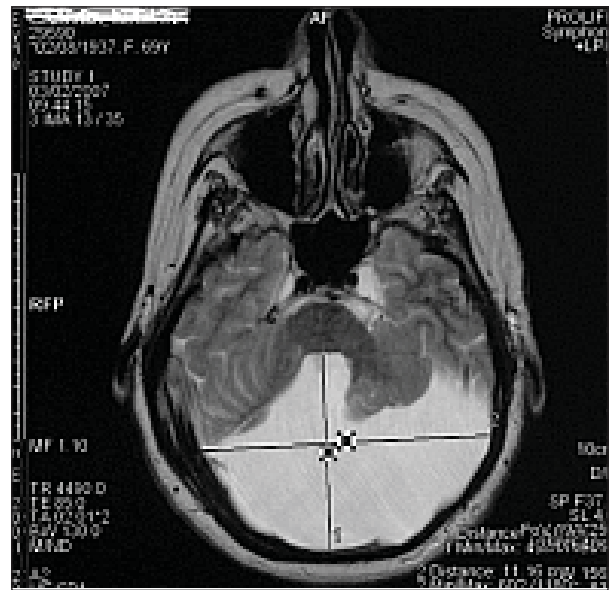


FIGURE 3

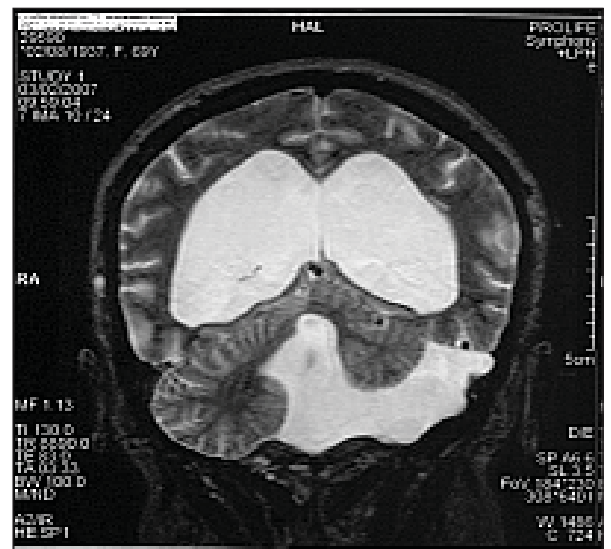


FIGURE 4

Sagittal T1-weighted MRI (Fig 1) shows cystic dilatation of the fourth ventricle and upward displacement of the lateral sinuses, tentorium cerebelli and torcular.

Transvers and coronal T2 weighted MRI (Fig 3 and Fig 4) show a large posterior fossa cyst measuring 60/112 mm with vermian and left cerebellar hemisphere hypoplasia and supratentorial ventriculomegaly (Fig 2).

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