PAMJ Clinical Medicine
Pan African Medical Journals

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Dandy-Walker variant with precocious puberty: a rare association

Hassan Baallal[1,&], Ali Akhaddar[1]

[1]Department of Neurosurgery, Avicenne Military Teaching Hospital, University Caddi ayyad, Marrakech, Morocco

[&]Corresponding author: Hassan Baallal, Department of Neurosurgery, Avicenne Military Teaching Hospital, University Caddi ayyad, Marrakech, Morocco

Received: 28 Apr 2020 - Accepted: 18 May 2020 - Published: 18 May 2020

Domain: Neurology (general), Neuroradiology

Key words: Dandy-Walker, puberty

Images in clinical medicine | Volume 3, Article 21, 18 May 2020 | 10.11604/pamj-cm.2020.3.21.23083

Available online at: https://www.clinical-medicine.panafrican-med-journal.com/content/article/3/21/full

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The Dandy-Walker malformation (DWM) is a rare congenital malformation involving the posterior fossa. It was first described in 1914 by Dandy and Blackfan. It is diagnosed when the following 3 main signs are identified: agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle, and an enlargement of the posterior fossa. However, the co-existence of the DWM with precocious puberty is rare. Precocious puberty is characterised by premature appearance of secondary sexual characteristics before the age of 7 years in girls and 9 years in boys. There are rare reports of the co-existence of the DWM and precocious puberty. A 6-year-old male child presented with enlargement of penis and appearance of axillary and pubic hair. His parents noticed a recent change in his voice, he was admitted to the hospital with complaints of headache for 3 months. He was free from dizziness, nausea, vomiting, or limb weakness. He had normal vision, muscular strength and muscular ton. Magnetic resonance imaging MRI axial T1, and sagittal T2 flair showed an atrophy of the cerebellar hemispheres, hypoplasia of the vermis, and an enlargement of the fourth ventricle, which communicated with the occipital cistern forming a posterior cerebellar cyst compatible with variant DWM.

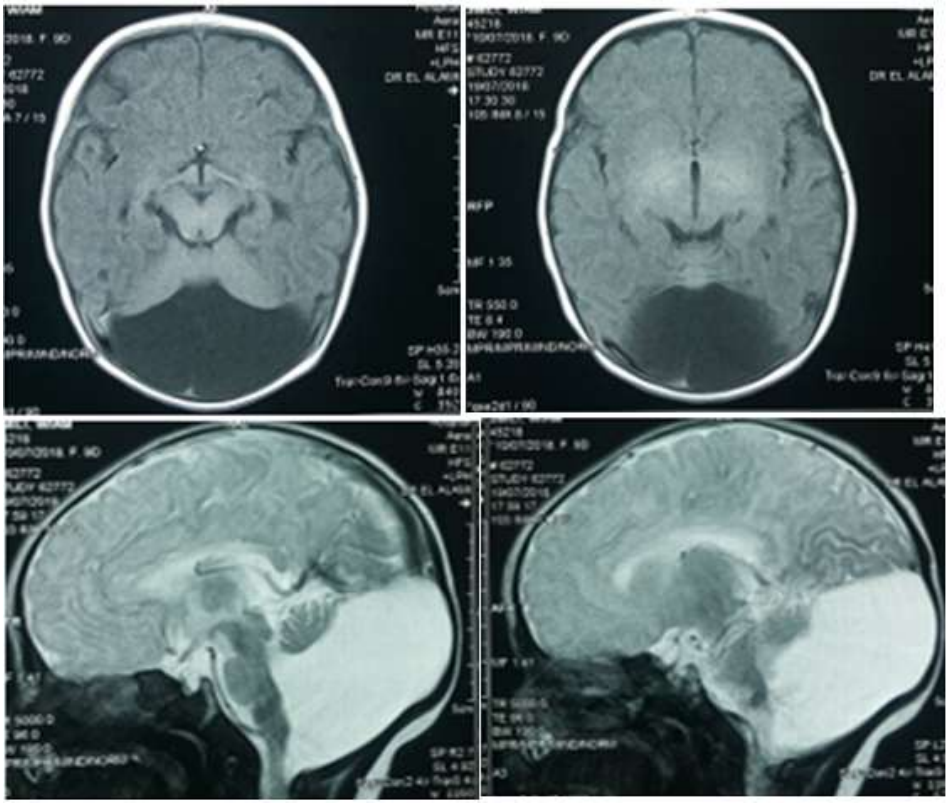

Figure 1: magnetic resonance imaging MRI axial T1, and sagittal T2 flair showed an atrophy of the cerebellar hemispheres, hypoplasia of the vermis, and an enlargement of the fourth ventricle