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Acute Respiratory Distress Revealing Dandy Walker Syndrome: One Case Report and Recent Review of Literature

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ABSTRACT

Dandy-walker syndrome is a rare disorder characterizsed by complete or partial agenesis of the vermis, cystic dilatation of the fourth ventricule and an enlarged posterior fossa. The precise aetiology is unknown, although there have been reports of associations with risk factors like maternal virus infections (rubella, toxoplasma and cytomegalovirus) and alcohol consumtion [1]. It describe a rare case of five months old boy with acute respiratory distress with the exploration objective a Dandy-walker syndrome.A five months old boy with no medical history was admitted to the intensive unity care fot an acute rispiratory distress, the exploration of the respiratory distress lead as to realising a cerebral CT scan which objictived dilatation of the ventricular system and enlargement of the fourth ventricular and vermin hypoplasia related to Dandy-walker syndrome.A dandy walker is a rare condition and the image found may not be the cause of the acute respiratory distress. Clinical presentation and prognosis depends on the prenatal and early diagnostic dering the first year of live and the treatement is surgical

Introduction

Dandy-walker syndrome is a rare condition, which consists of hypopkasia of the cerebellar vermis, dilatation of the fourth ventricule and an elarged posterior fossa. The syndrome is considered to be the commonest cerebellar malformation. It is poorly understood, and the incidence is unknown. Although this malformation can be diagnosed prenatally by neuroradiology, identifying patients is difficult as there is no specific symptom or sign. The outcome is highly variable and ranges drom normal or nearly normal development disability or even early death.

Clinical case

A five months old boy, with no medical history, was admitted to the intensive care unity for acute respiratory distress.

Evaluation of vital signs revealed pyrexia (T :37 C), an hypotonic, non reactif patient, tachypnea of 50 breaths/minute, pulse oxymetry was 75% at ambient air and 92% with high concentration mask, and signs of respiratory struggles. Hemodinamacally, blood pressure was high (100/50 mmHg), tachycardia with a pulse of 170 beats/min.

The initial arterial blood gas revealed an acidemy with respiratory acidos and severe hypoxémia (PaO2/FiO2 ratio at 230)

At this point, he was on a mechanical ventilator

The initial exploration : thoracic radiography, pleural ultrasound and transthorasic echocardiography were negative

A cerebral CT scan was performed to search a central cause of the respiratory distress which objictived tha dilatation of the ventricular system and enlargement of the V4 and vermin hypoplasia related to Dandy Walker syndrome [figure 1].

The evolution was unfavorable with the installation of acute distress respiratory syndrom

The patient passed away three days after his admission

Discussion :

The incidence of dandy walker syndrom was reported to vary between 1 to 25 in 35000 live births, with predominance of male sexe (ratio 3 :1) with a mortality rate independent drom the symptomatology or from the âge of diagnosis between 5% and 20% [2-3].

The syndrome is detected prenatally, by ultrasound or magnetic resonance imaging [4}, or increased head circumference (prenatally or postnatally), the diagnosis is challenging because of lack of specific symptomatology [5].

The aetiology is unclear, however it was reported some risk factors like maternal viral infection (rubella, toxoplasma, cytomegalovirus) and alcohol ingestion [6]. genetic and chromosomal abnormalities have also been found like chromose 13 trisomy, 18 y 21, 3q, 6p, 9p trisomy, 13q deletion [7-8].

The clinical manifestation of Dandy-walker syndrome are so variable, and occur during the first year, it associat psycho-motor delay, visual and hearing difficulties, hypotony, respiratory difficulties, spasticity and convulsions, while others may not present clinical manifestations [9].

The treatment of dandy-Walker syndrome consists of dealing with hydrocephalus by various approaches, whoever this is still controversial [10].

Conclusion

Dandy-walker syndrome is a rare yet serious disorder, characterized by the disgenesia or agenesia of cerebellar vermis, dilatation of V4 and an enlarged posterior fossa.

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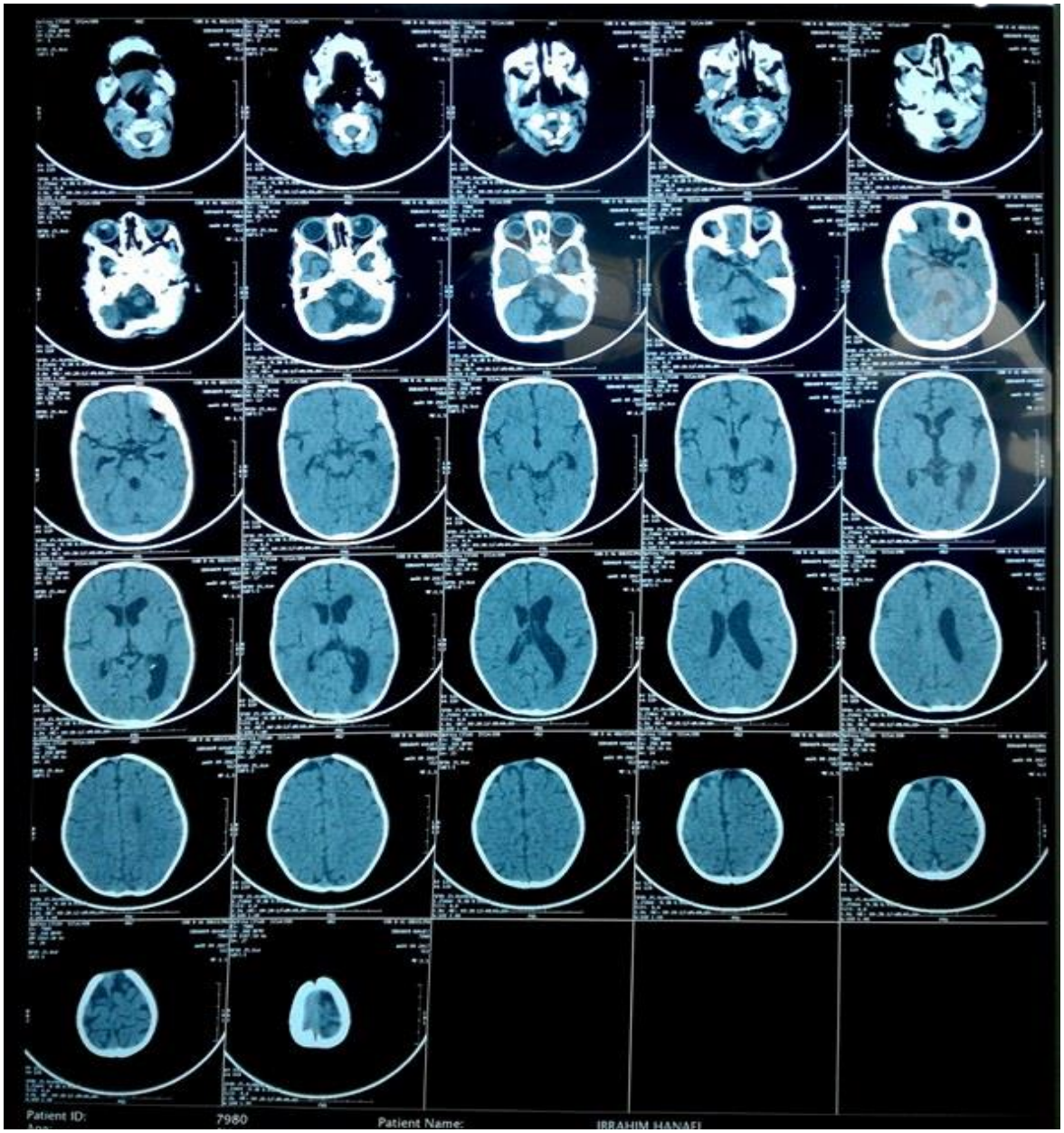

Clinical presentation and prognosis depends on the prenatal and early diagnostic dering the first year of live and the treatement is surgical

References

1. H.K Al-Turkistani. King fahd hospital of the university Alkhobar, Journal of Taibah university Medical sciences 2014
2. Figueroa-Angel V, Reyes-Moreno I, García-López R, Rodriguez-Aceves C, Martínez-Muniz I.Síndrome de Dandy Walker. Arch Neurocien (Mex). 2013;18:92---8.
3. Guzman-Lepe A, Baltazar-Alba I, Padilla-Soto S.Síndrome de Dandy-Walker: presentación de un caso. Hipoc Rev Med.2011,3 :9_10
4. Donkelaar HJ, Lammens M, Wesseling P, Thijssen HOM, Renier WO. Development and developmental disorders of the human cerebellum. J Neurol 2003; 250: 1025-1036.
5. Maria BL, Zinreich SJ, Carson BCM, Rosenbaum AE, Freeman JM. Dandy-Walker syndrome revisited. Pediatr Neurosci 1987; 13: 45e51.
6. Altman NR, Naidich TP, Braffman BH. Posterior fossa malformations.AJNR Am J Neuroradiol 1992; 13: 691e724.
7. Congenital malformations of the brain and skull. En: Barkovich J, editor. Pediatric Neuroimaging. 4th ed. Philadelphia, USA: Lippincott Williams & Wilkins; 2005. p. 292-439.
8. Mytilinaios DG, Tsamis KI, Njau SN, Polyzoides K, Baloyanis SJ. Neuropathological findings in Dandy Walker variant. Developmental Neurorehabilitation. 2010;13:64---7
9.Ryan M, Grenier E, Castro A, Nemeroff CB. New-onset psychosis associates with Dandy Walker variant in a adolescent female patient. J Neuropsychiatry Clin Neurosci. 2012;24:241---6.