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CT Scan Contribution in the Diagnosis of Schizencephaly and Dandy-Walker Malformation: A Rare Association Case

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Abstract | **Case Report**

Schizencephaly and Dandy-Walker syndrome are two rare congenital malformations of the central nervous system, distinguished by their location and characteristics. Complex developmental processes involving both the cerebral cortex and the cerebellum are thought to contribute to this anomaly. We report the case of a 3-year-old girl, who underwent surgery two years earlier for hydrocephalus with the placement of a ventriculoperitoneal shunt, and has a history of epilepsy. In this context, she underwent a brain CT scan for etiological purposes, which revealed schizencephaly associated with Dandy-Walker malformation.

Keywords: Schizencephaly, Dandy-Walker Malformation, Brain CT scan.

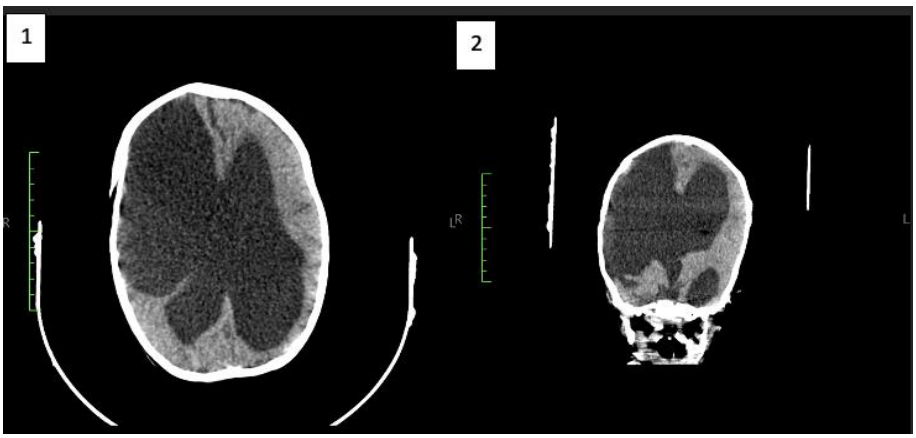
INTRODUCTION

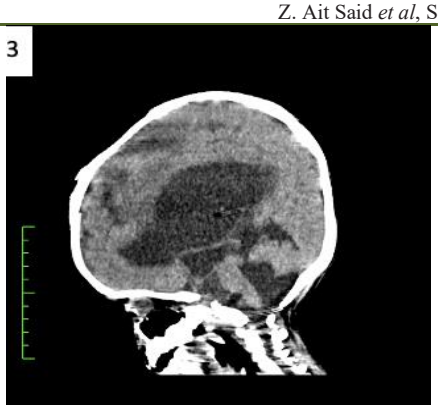
Schizencephaly and Dandy-Walker malformation are two severe cerebral malformations that may coexist, although this association is exceedingly rare. Complex developmental processes involving both the cerebral cortex and the cerebellum are thought to contribute to this anomaly [1]. Although the underlying mechanisms are still poorly understood, brain CT scan is a key tool in identifying these malformations, allowing visualization of cortical clefts, hypoplasia of the cerebellar vermis, and ventricular abnormalities, thereby providing a detailed assessment of the extent and location of the lesions.

PATIENT AND OBSERVATION

We report the case of a 3-year-old girl, who underwent surgery two years earlier for hydrocephalus with the placement of a ventriculoperitoneal shunt, and has a history of epilepsy. In this context, she underwent a brain CT scan for etiological purposes.

The brain CT scan revealed a large right fronto-temporal cleft, which appears to communicate with the ipsilateral lateral ventricle, along with cystic dilation of the posterior cranial fossa that communicates with the fourth ventricle. These abnormalities are consistent with schizencephaly associated with Dandy-Walker malformation.





Figures 1, 2, 3: Brain CT scan in axial (1), coronal (2), and sagittal (3) cuts show significant dilation of the ventricular system, hypoplasia of the cerebellar vermis with communication between the fourth ventricle and a large retro-cerebellar cystic cavity, consistent with Dandy-Walker malformation. An expanded cortical cleft, suggestive of schizencephaly, is also observed

DISCUSSION

Schizencephaly and Dandy-Walker syndrome are two rare congenital malformations of the central nervous system, distinct in their location and characteristics. Schizencephaly is characterized by a cerebral cleft lined with gray matter, connecting the ventricular cavity to the cortical surface, while Dandy-Walker syndrome is associated with agenesis or hypoplasia of the cerebellar vermis, dilation of the fourth ventricle, and a posterior fossa cyst [2].

Clinically, both conditions share common neurological signs such as psychomotor delay, axial hypotonia, balance disorders, as well as seizures or epilepsy. Macrocephaly may be observed, especially in cases of associated hydrocephalus [3].

On CT scan, schizencephaly is characterized by the presence of cortical clefts in the cerebral cortex, which appear as hypodense linear or cystic areas. These clefts can be unilateral or bilateral and are often in communication with the lateral ventricles or subarachnoid spaces. Lateral ventricle dilation is frequently observed, particularly at the level of the lateral ventricles, associated with abnormalities of the corpus callosum such as hypoplasia or agenesis. CT imaging in Dandy-Walker malformation shows cystic dilation of the posterior fossa, particularly affecting the fourth ventricle, which is visible as a distinct hypodense cyst. The cyst often communicates with the fourth ventricle, causing its expansion, as well as hypoplasia of the cerebellar vermis and enlargement of the lateral ventricles. The expansion of the cyst also causes displacement of the brainstem, typically downward [4,5].

While CT scans provide good visualization of large brain structures and are commonly used for diagnosing Dandy-Walker malformation and schizencephaly, MRI remains the gold standard for detailed evaluation of these abnormalities. MRI offers superior resolution and allows for precise analysis of cerebellar malformations, cortical clefts, and subcortical structures. Therefore, it is an essential complement to CT scanning for a comprehensive assessment of complex brain malformations [1].

The management of schizencephaly and Dandy-Walker syndrome is multidisciplinary. This includes neurodevelopmental monitoring to track the progression of motor and cognitive deficits, early intervention in the case of seizures, as well as specific treatments such as ventricular shunting for hydrocephalus [6].

CONCLUSION

The association of schizencephaly with Dandy-Walker malformation represents a rare and complex combination of brain anomalies. Cerebral computed tomography is an essential tool for visualizing these anomalies, providing a detailed assessment of cortical and ventricular lesions. With its ability to deliver clear images, CT not only facilitates diagnosis but also helps evaluate the extent of the abnormalities, guiding therapeutic decisions.

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