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Clinical Manifestations and Neuroanatomical Correlates of Reversible Impairment of Cognitive Functions in Patients with Normotensive Hydrocephalus: Case Report

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ABSTRACT

From a neuropsychological perspective, normotensive hydrocephalus NPH is characterized by cognitive impairment associated with the compression exerted by ventricular dilatation on the cingulate cortex and fronto-striatal circuits. The Rey-Osterrieth Complex Figure Test (ROCFT) is a neuropsychological assessment measure widely used by clinicians and researchers because of the wide variety of cognitive information it allows to estimate.

We present the case of young male patient with refractory structural epilepsy secondary to a childhood brain injury resulting from a metastasis of a renal tumor. Referred to Neuropsychology for behavioral changes and cognitive impairment; presenting atypical performance in the copying phase of the ROCFT.

Failures in visual integration and coding of the ROCFT copy show that, in cases of NPH, brain structures such as the cerebellum and posterior cingulum can also be compromised by the compression exerted by the cerebral hemispheres as they move due to ventricular dilatation. Failures in integration, planning, rotation and displacement of ROCFT elements have been reported in patients with secondary psychotic conditions and posterior cingulate tumors.

Failures in visual integration associated with NPH may be related to the involvement of more postero-caudal brain structures such as the cerebellum and posterior cingulum or to secondary psychosis.

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INTRODUCTION

Hydrocephalus (HC) is a neurological condition characterized by an increase in cerebrospinal fluid volume that causes compression of brain tissue against the skull, resulting in widespread damage to neural structures1. HC can affect patients of any age, usually presenting at birth or shortly thereafter, but can also appear suddenly in adulthood in the form of normal pressure hydrocephalus (NPH)2.

NPH is usually secondary to infection, tumor or trauma[3]. HPN is a progressive condition characterized mainly by medial frontal syndrome (apathy, gait apraxia and sphincter dyscontrol)[1]; has distinctive radiological features in brain imaging, such as an Evans index ≥ 0,3[4].

NPH usually presents with a gradual worsening of cognitive symptoms resulting from the degradation of paraventricular and subcortical structures5. However, few studies have investigated and described the cognitive functioning in NPH. The Rey-Osterrieth Complex Figure Test (ROCF) is a neuropsychological assessment measure widely used by clinicians and researchers because it generates a wide variety of information that allows us to measure the status of skills such as: integration of visual information, visual memory, planning and motor execution to create or reproduce patterns, shapes or designs[6]. As such, this tool is valuable for understanding an individual's brain functioning and for identifying possible areas of cognitive impairment.

Atypical performance on the ROCF can be observed in several neuropsychiatric and neuropsychological conditions. Some of the pathologies in which atypical performance on this test has been described include: Alzheimer's disease,

Parkinson's disease, Lewy body, vascular and frontotemporal dementia, cerebellar ataxia, amyotrophic lateral sclerosis, acquired brain injury, brain tumors, eating disorders, and schizophrenia[7], in addition to autism spectrum disorders[8]. No information related to ROCF performance in NPH populations has been found.

CLINICAL CASE PRESENTATION

We present the case of a 20 year old male patient with refractory structural epilepsy secondary to a childhood brain lesion resulting from a metastasis of a renal tumor. He was managed with chemotherapy and radiotherapy, and at the age of 9 years presented a first episode of hydrocephalus that was managed with ventriculoperitoneal shunt. He maintained low average cognitive functioning until a few months prior to the onset of clinical care. He was referred to Neuropsychology (NPS) for significant behavioral changes and cognitive impairment.

In attention with NPS, the performance in the copying phase of the TFCR is striking (Figure 1). The copy, completely disintegrated, evidences primarily executive flaws, in addition, it is an unconventional figure, loaded with atypical errors.

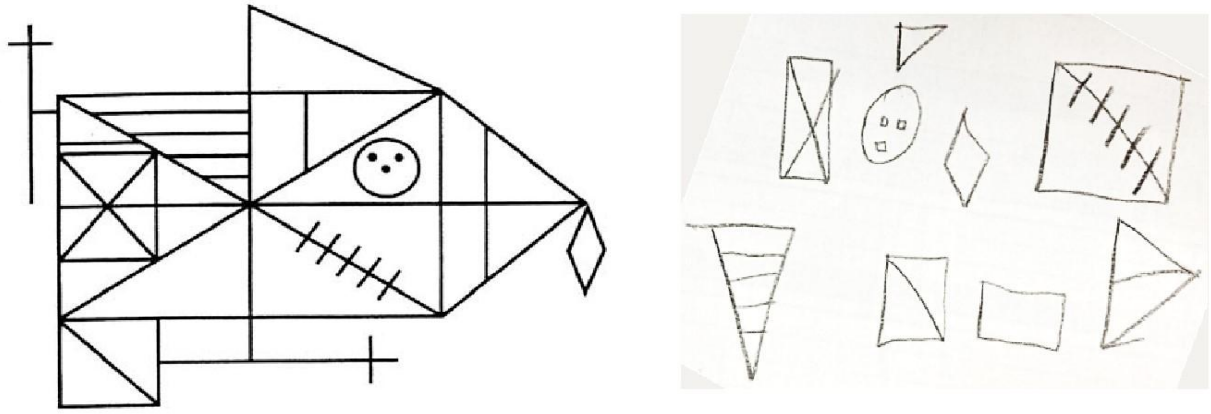

Figure 1.On the left side the ROCF model is shown. On the right side the patient's execution.

Shortly after NPS appointment, the patient is admitted to the emergency for vomiting and severe gait disturbance. Specialists in neurology and neurosurgery determine NPH (Figure 2). After surgical management, and although the Patient presented important complications due to infection of the surgical site by wound dehiscence, cognitive and behavioral improvement was evidenced in the patient, returning to his previous functionality.

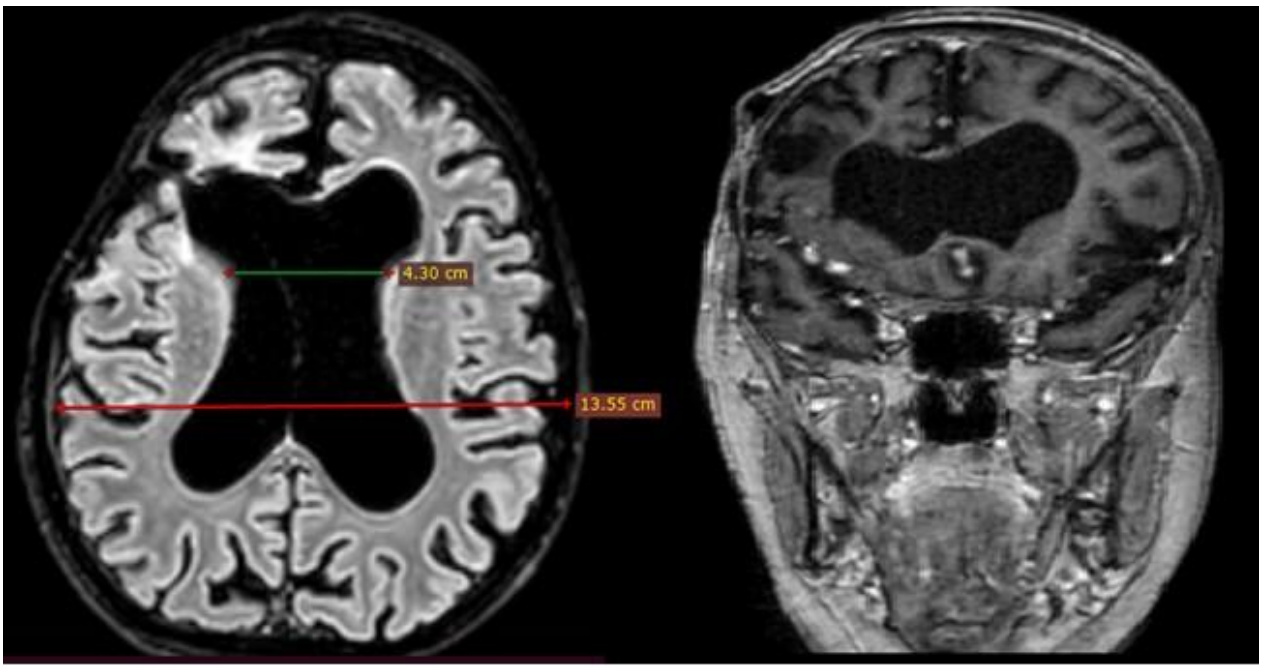

Figure 2. Magnetic resonance image, right frontal gliosis and hydrocephalus are identified by Evans index by 0.31.

The patient's performance at this second time point, through the integrated copy of the ROCF (Figure 3), maintains errors typical of his epilepsy, but evidences radical differences in terms of his pre-surgical functioning.

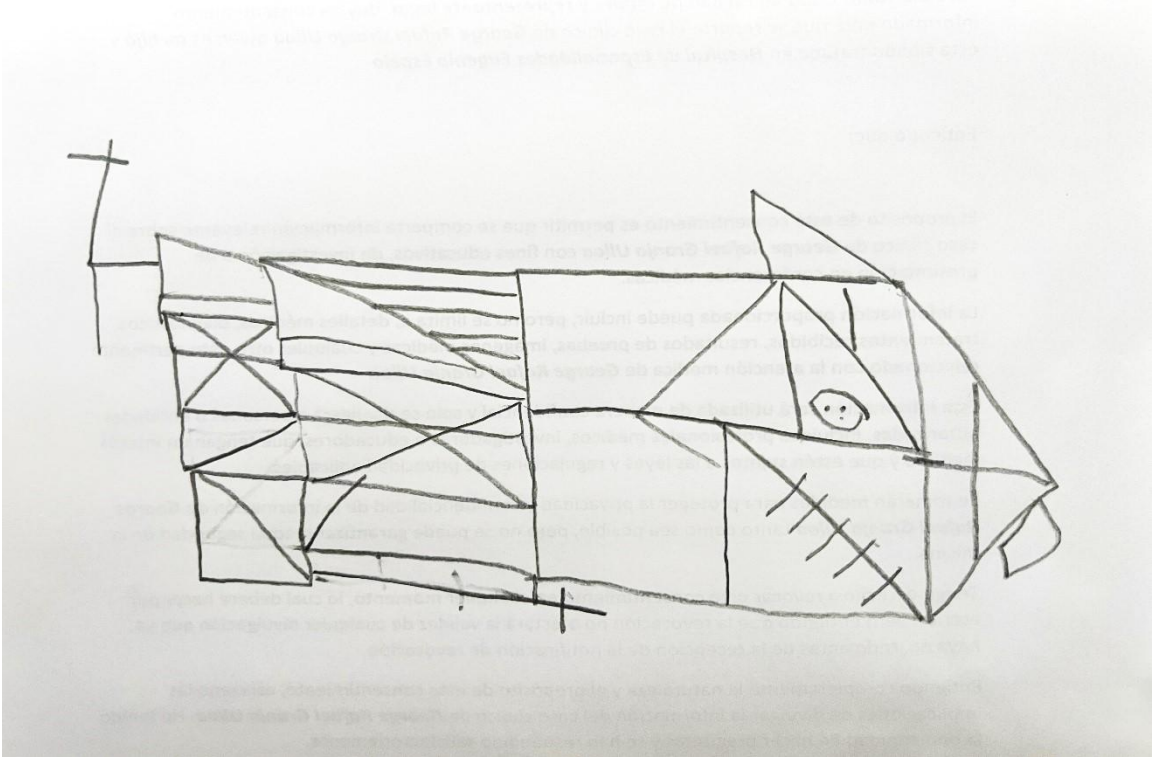

Figure 3. ROCF copying phase after surgical resolution

DISCUSSION

We report the case of a young adult with NPH presenting atypical performance on the ROCF, primarily denoting failures in the integration of visual information and executive function**.** There have been reports of patients who undergo ventriculoperitoneal shunt surgery and significantly improve their cognitive functioning. This work represents one of the few reported cases, to the authors' knowledge, of failure to integrate visual information in a patient with NPH[9]. And the only one that has used the ROCF as an evaluation tool.

NPH is expected to affect executive functions in patients, given the compression exerted by ventricular dilatation on the cingulate cortex, the frontostriatal circuits and, in cases of intracranial hypertension, on the dorsolateral prefrontal cortex[7,10] . However, the phenomenon of disintegration denotes that, in addition, there is a degree of compression in other specific brain structures.

The cerebellum plays an important role in the integration of visual information and its encoding[7, 11]; two glaring failures in patient throughput. In the case of hydrocephalus and intracranial hypertension, the cerebellum may be involved by the compression exerted by the cerebral hemispheres as they are displaced by ventricular dilatation.

There are several reports and case series of integration, planning, rotation and displacement failures in the ROCF in patients with psychosis secondary to anti-NMDA encephalitis[12]; and in cases of posterior cingulum tumors[13].

The latter, in contrast to several reports and case series of NPH in patients with schizophrenia[14,15] supports the hypothesis that disintegrative elements in ROCF may occur in secondary psychotic disorders; and by the involvement of more postero-caudal brain structures.

CONCLUSION

The failures in the integration of visual information evidenced in this case of TNH may be the result of the involvement of more postero-caudal brain structures such as the cerebellum and posterior cingulum, or appear in association with secondary psychosis.

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CONFLICT OF INTEREST

The authors of this manuscript have no conflicts of interest to declare.

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