Cognitive evaluation following the evolution of brain myofibroblastic tumor in the adolescence: a case study report

Avaliação cognitiva após a evolução do tumor miofibroblástico cerebral na adolescência: relato de estudo de caso

ABSTRACT

Objective: To report the case of a teenager (12 years old) diagnosed with a brain tumor in the right frontal-parietal region emphasizing the main characteristics observed in neuropsychological examinations. Methods: In the pre-surgical evaluation, the patient presented behavioral alterations, including deficits in verbal comprehension, perceptual organization, working memory, processing speed, and slight alterations regarding constructive praxis. Results: A reevaluation after two years surgery revealed significant improvement in verbal and perceptual comprehension and constructive praxis while remaining a slight change in processing speed. These results suggest that the tumor’s surgical resection produced significant improvements in the patient’s neurocognitive context, especially in executive functions. This study also indicates that Neuropsychological evaluation are useful for pre- and post-surgical evaluation of cognitive functioning and its evolution. Conclusion: Brain tumor causes cognitive and behavioral changes and its resection can result in improvements in the patient’s quality of life.

KEYWORDS
Cognition, brain tumor, adolescence, neuropsychological evaluation.

RESUMO

Objetivo: Relatar o caso de uma adolescente (12 anos de idade) diagnosticada com tumor cerebral na região frontoparietal direita, enfatizando as principais características observadas em exames neuropsicológicos. Métodos: Na avaliação pré-cirúrgica, a paciente apresentou alterações comportamentais, incluindo déficits na compreensão verbal, organização perceptual, memória de trabalho, velocidade de processamento e pequenas alterações na praxia construtiva. Resultados: Uma reavaliação dois anos após a cirurgia revelou melhora significativa na compreensão verbal e perceptiva e na praxia construtiva, permanecendo uma ligeira alteração na velocidade de processamento. Esses resultados sugerem que a ressecção cirúrgica do tumor produziu melhoras significativas no contexto neurocognitivo da paciente, sobretudo nas funções executivas. Este estudo também indica que a avaliação neuropsicológica é útil para avaliação pré e pós-cirúrgica do funcionamento cognitivo e sua evolução. Conclusão: O tumor cerebral causa alterações cognitivas e comportamentais e a sua ressecção pode resultar em melhorias na qualidade de vida do paciente.

PALAVRAS-CHAVE
Cognição, tumor cerebral, adolescência, avaliação neuropsicológica.
INTRODUCTION

Tumors of the central nervous system (CNS) in adolescence significantly impact morbidity and mortality and often follow cognitive and behavioral changes. Neuropsychological evaluation (NPE) is an instrument used to assess cognitive changes\(^1\) severity and plan rehabilitation. CNS tumors are more prevalent in individuals aged 0-14 years; mean annual incidence rate adjusted by age is 5.65 per 100,000 inhabitants\(^1,2\). The inflammatory myofibroblastic tumor (IMT) is also described as inflammatory pseudotumor or plasma cell granuloma, among the benign or low malignancy potential lesions. IMT is a rare neoplasm composed of myofibroblastic fusiform cells, usually accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils\(^3,4\) and occurs mainly in soft tissues and the viscer of children and young adults\(^3,5\). CNS tumors, in general, are capable of producing signs and symptoms through local invasion, compression of adjacent structures, and increased intracranial pressure. In children and adolescents, these manifestations can be unspecific\(^5\). In 7% of the cases of adolescents with IMT, deficits in behavioral and school performance may occur\(^6\). Also, children treated for low-grade CNS tumors present an increased risk for impairment of neurobehavioral function\(^6\).

Neuropsychological evaluation in CNS tumors can contribute to the early diagnosis of these conditions, decisively influencing the evolution and prognosis\(^7\). In the following case, we describe social behavior and cognition changes in a 12-year-old patient diagnosed with IMT. We sought to describe the main neuropsychological alterations in the preoperative and postoperative periods and the case’s evolutionary outcome.

CASE REPORT

Identification and history of the disease

ALS, 12 years old, female, an elementary school in progress. The first consultation occurred in 2018 with the family’s main complaint of challenging behavior and the presence of signs of disinhibition, motor stereotypes, and saccadic eye movements.

The patient had a history of motor, cognitive, and socio-affective development within the expected standards until seven years old when she started to decrease school performance and psychomotor retardation. The condition evolved with progressive irritability, difficulty concentrating, and the onset of obsessive-compulsive symptoms.

After three years, she began to present episodes of frequent and brief changes in the level of consciousness, associated with paroxysmal vertigo and paresthesia in the left upper limb. Magnetic resonance imaging demonstrated brain tumor (BT) in the right frontal-parietal region. Three months later surgical resection of the lesion was performed (Figure 1) and the subsequent anatomopathological study revealed an IMT-CNS.

Preoperative evolution

According to the relative’s report, the patient developed mood dysphoria, decreased will and general disinterest, evidenced mainly in the academic context, and aggravated the behavior opposing the norms. In the following months, she presented a substantial reduction of appetite and weight loss, about 6.0 kg in 2 months. There was also alternation of the saccadic ocular movements, body rocking stereotypes, and frequent difficulties in performing daily life activities. Subsequently, there was an increase of the oppositional behavior and adoption of a defiant attitude, evidenced by the non-acceptance of rules, intentional speech with words of low slang and disinhibition, manifested in various contexts, in addition to a sharp drop in school performance and frequent school occurrences.

Initial neuropsychological examination

During the pre-surgical evaluation (December 2018), the patient presented a non-collaborative attitude (she needed encouragement to perform the requested activities) and was uninhibited; the speech focused on sexual issues, irritable mood, unstable affection, hypobulia and absence of morbidity judgment.

The Wechsler Intelligence Scale for Children (4th Edition – WISC-IV) was applied and reading in the first and second evaluation was medium (see Table 1). There were changes in the factorial indices: verbal comprehension with difficulties in interpreting complex sentences and texts; perceptual organization with scores below the predicted for the age; working memory with scores lower than expected for immediate, short-term and working memory; processing speed with a score below the predicted for her age. In the Rey Auditory-Verbal Learning Test (RAVLT), a lower than average score was observed in most test components (see Table 1).

Rey Complex Figure Test was also applied, with an average result in the copy of a complex geometric figure = 36 (PC = 90) and above the average in visual memory = 9,5 (PC = 50), reading in copy was upper-middle and reading in memory was medium. Similar results were seen in the Benton Visual Retention Test (BVRT). However, there were slight difficulties in carrying out activities involving constructive praxis, visual-spatial orientation and fine motor skills (see Table 1).

Cognitive reassessment (post-surgery)

Eighteen months after the first neuropsychological evaluation, the patient, now 14 years old, was reevaluated, and there were significant improvements in behavior and cognitive performance. The evaluation took place in five sessions of approximately 90 minutes each.
### Table 1. Results of the neuropsychological evaluation in 2018 and 2020

<table>
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<tr>
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<td>Intelligence</td>
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#### Rey Complex x Figure Test
- **COPY** = 36
- **MEMORY** = 9,5
- PC = 90  
- PC = 50

#### Benton Visual Retention Test
- **MEMORY** = 25
- PC = 40  
- PC = 20

#### Rey Auditory Verbal Learning Test
- **MEMORY** = 25
- PC = 40

#### Wechsler Intelligene Scale for Children

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### Interpretation
- **MEMORY** = 25
- PC = 40
- PC = 20

- **COPY** = 36
- PC = 90
- PC = 50

Medium

#### Wechsler Intelligene Scale for Children

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#### Medium

- **COPY** = 36
- PC = 90
- PC = 50

- **MEMORY** = 9,5
- PC = 50

Upper Middle

Medium
Figure 1. Axial Tomography the level where the cystic lesion is first visible (red arrow/rectangle), particularly the parietal (A) and right frontal lobe (B). The coronal slice (C) depicts the area after surgical removal.

In the evaluation with WISC-IV, a change in the intelligence quotient (IQ) was obtained (Table 1). In the factorial rates: presented good verbal comprehension; perceptual organization with scores slightly below the predicted for the age; working memory with scores lower than expected for immediate, short-term and working memory; processing speed with a score below the predicted for her age. In the RAVLT, scores within the average were observed in most test components (Table 1).

The Rey Complex Figure Test, with below-average results in copying a complex geometric figure = 30 (PC = 20) and visual memory = 25 (PC = 40), reading in copy was lower and reading in memory was medium. Results within the average and superior to those of the first evaluation were observed in the BVRT (Table 1). Better dexterity was observed in carrying out activities that involved constructive praxis, visual-spatial orientation and fine motor skills, compared to the previous assessment.

In the WISC-IV and RAVLT tests, there was a compromise in activities related to interpreting complex phrases and texts, perceptual organization, immediate memory, short-term memory, working memory and processing speed. The Rey Complex Figure and BVRT tests were also applied, in which there was some compromise in the activities related to constructive praxis, visual-spatial orientation, perceptual organization and fine motor skills.

**DISCUSSION**

Individuals with CNS tumors often live with impairments resulting from the injury itself or its treatment, with cognitive dysfunction being the most frequent complication as a long term outcome. Cognitive changes resulting from brain injuries can be assessed through ANP, helping to understand the impact of the injury and associated treatment on performing activities of daily living and managing possible changes in mood or personality.

Changes in cognitive functions and neuropsychiatric impairment, in the context of intracranial neoplasms, may affect from 51% to 78% of cases. Specifically, in the context of low-grade brain tumors, it is already documented that the presence of the tumor itself contributes as a primary factor for cognitive dysfunction. In patients with frontal or temporal tumors, before treatment, 90% of the patients presented impairments in at least one area of cognitive functioning. Brain tumors in the pediatric population, cognitive impairment is observed in functions such as language and non-linguistic skills, attention, working memory and processing speed. Memory is significantly impaired in tumors located in the third ventricle and thalamus region’s temporal and frontal lobes. Executive dysfunctions are frequently observed in primary tumors located in the frontal lobe or regions with frontal connections as well as changes in the speed of information processing and working memory.

A meta-analysis assessed the association between psychiatric symptoms and tumor location. It was observed that specific locations, e.g., frontal and temporal areas, were more likely to be affected, leading to personality changes. Such associations have long been described, as well as changes in mood/affection. What is described in the literature is true in this case, with the patient’s significant behavioral changes and the presence of frontal involvement.

From the neurobiological point of view, the changes presented are associated to the disconnection of limbic-frontal pathways, related to social cognition, affecting different modalities, such as decision making, inhibitory control, the intensity of the experience of immediate...
Cognitive evaluation in brain myofibroblastic tumor

The brain tumor caused changes in the patient’s cognitive and behavioral sphere, being these closely related to the location of the lesion and the findings in the NPE. Tumor resection resulted in improvements in various cognitive and behavioral domains. Besides, the performance of a multidisciplinary follow-up with a psycho-educationalist, a psychologist of Behavioral Cognitive Therapy approach (TCC), the neurologist and the neurosurgeon physician certainly contributed to the significant improvement of the picture initially presented.

INDIVIDUAL CONTRIBUTIONS

Cândida Helena Lopes Alves – Cognitive assessment, manuscript writing.
Gilberto Sousa Alves – Study design, manuscript writing and final revision.
Anna Alzira Macau Furtado Ferreira – Data revision, manuscript writing.
Eliza Maria da Costa Brito Lacerda – Data revision, manuscript writing.
Carlos Tomaz – Study design, manuscript writing and final revision.

CONFLICTS OF INTEREST

The authors report no conflicts of interest.

ACKNOWLEDGMENTS

We thank all those who allowed this study to be carried out successfully.

REFERENCES


