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Case presentation

Features of complex therapeutical rehabilitation management with favorable evolution in a patient with right hemiplegia, mixed aphasia and optic atrophy post surgical removal of be-nign intraventricular tumor (central neurocytoma)

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ABSTRACT: Central neurocytoma (CN) is a benign brain tumor located intraventricularly and classified as grade II by the World Health Organization in 2000 (classification of tumours of the central nervous system). CN is frequently found in young adults and may increase intracranial pressure causing obstructive hydrocephalus, manifested by neurological symptoms such as headaches and vision problems. CN has a relatively good prognosis, provided a complete surgical resection is performed. **Materials and Methods**: In this presentation, we describe the case of a 21-year-old patient with a personal history of headache, vomiting and decreased visual acuity in both eyes, who was admitted to the Neurosurgery Clinic III of THEBA. Following clinical and paraclinical assesements, the patient was diagnosed with a left lateral ventricular tumor with extension in the right lateral ventricle and the third ventricle. The neurosurgery team decided a total resection of the tumor would be the best approach in this case. The biopsy report revealed that it was a central neurocytoma. After the surgery the patient's neurological status improved and she was admitted in the Neuromuscular Rehabilitation CinicalDivision with flaccid right hemiplegia and mixed aphasia. The patient was functionally assessed using the following scales: Functional Independence Measure (FIM), Montreal Cognitive Assessment (MOCA), modified Ashworth, Penn Spasm Frequency Scale (Penn), Life Quality Assessment (QOL), FAC International Scale, Glasgow Outcome Scale-Extended (GOS-E), modified Rankin scale (mRS), Aphasia Screening Test (AST-Whurr). **Results**: The patient showed a favorable evolution with remitted aphasia and walking traing with self-support in tertrapod walking stick. At the same time, the patient can use the plegic upper limb in performing feeding activities. **Conclusion:** It should be noted that neurosurgical intervention and pharmacological treatment, associated with an individual rehabilitation program consisting of: physical, occupational and speech therapies and also rehabilitation nursing interventions in a patient diagnosed with flaccid right hemiplegia and mixed aphasia after total surgical resection of the intraventricular central neurocytoma has improved the control of symptoms and the patient's quality of life.

Keywords: *benign brain tumor, central neurocytoma, hemiplegia, rehabilitation*

1. INTRODUCTION

Central neurocytoma (CN) comprises 0,1%-0,5% of all brain tumors. It was firstly described by Haussoun et al. in 1982 (1) and was classified as grade II by the World Health Organization in 2000 (tumors are relatively slow-growing but sometimes recur as higher grade tumors).(3)

Epidemiology: CNs are most prevalent among young adults, and nearly 25% of all cases involve individuals in their thirties. About 70% of affected individuals are between the age group of 20 and 40 years, both sexes are almost equally affected, with a male-to-female ratio of 1,02:1.(2)

Some studies have indicated higher incidences of CNs in Korea, India, and Japan, which is possibly attributed to genetic differences among racial groups that make certain individuals more prone to CNs than others . The higher incidence in these Asian countries, make this tumor an important consideration when dealing with intraventricular tumors in these populations.(2)

Tumor location: CN is classically located in the lateral ventricle and/or the third ventricle with the septum pellucidum attachement to be one of the features of the tumor. However, the most common site is the anterior portion of one of the lateral ventricles followed by combined extension into the lateral and third ventricles. However, recently, they were also found in an intra-axial location (cerebral, cerebellar, brainstem, or spinal parenchyma) and termed as "extraventricular neurocytoma" (EVN). "Cerebral neurocytoma" is the term usually used to designate both CN and EVN.(4)

Clinical manifestations: CN may increase the intracranial pressure by obstructing the interventricular foramen, which can lead to hydrocephalus. Patients may also experience nausea, vomiting, headache, seizures, decreased consciousness, weakness, and memory or vision problems (2). In rare cases, intraventricular hemorrhage may also occur. Patients with EVN present with similar symptoms, in addition to weakness and numbness in the limbs (5). These symptoms are typically present for approximately 3–6 months, although the duration of symptoms can vary from a few days to many years. The duration seems to be mostly related to tumor location, and does not seem to be correlated to the aggressiveness of the tumor (2).

Hystopathological analysis: The characteristics of light microscopic examinations include predominantly benign lesions with morphological features similar to the so-called ependymoma of the foramen of Monro, of the oligodendrogliomas and of the neuroblastomas (2). Although oligodendroglioma presents very similar features on light microscopy, on electron microscopy central neurocytoma cells are found to contain numerous synapses and to exhibit neuronal differentiation. Jerdan et al.(6) noted neuronal differentiation in similar adult intraventricular tumors but, because of the absence of synapses, designated them "differentiated cerebral neuroblastomas"(8). Immunohistochemistry and electron microscopy confirmed the diagnosis in each case, showing expression of neuron specific enolase and synaptophysin and containing microtubuli, neurosecretory granules, and presynaptic vesicles. In agreement with the literature, the authors stress the benign behaviour of most of these tumors and the need for systematic immunohistochemical and ultrastructural study. (6)

Radiological features: CN can appear as a dense mass in computerized tomography (CT) scans indicating calcifications, which occur in up to 50% of all cases, and present a patchy and coarse appearances (Fig. 1) (2). The tumor can also be heterogeneous because of the hypodense areas related to cystic degeneration . In contrast-enhanced CT scans, CNs have mild to moderate enhancements. Unfortunately, there is no estab-lished criterion to distinguish between CN and other tumors such as oligodendrogliomas on CT scans and MRI (2).

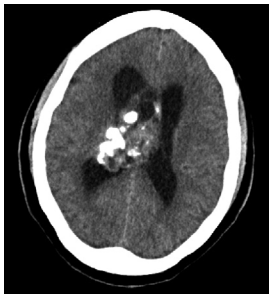


Fig. 1. Axial CT demonstrating a large hypodense central neurocytoma. Moderate, heterogenous hyperdensities are consistent with calcifications. (2)

Surgery: Surgical management with a gross-total resection is currently the gold standard treatment for CNs, which often has excellent prognosis and minimizes the chances of CN recurrence. Gross-total resection is achieved in nearly 30–50% of all CN

patients. In comparison, individuals who had surgery with only subtotal resection had an 86% five-year survival rate. Subtotal resection of CN increases the rate of recurrence and decreases the rate of survival (2).

Radiotherapy: Radiotherapy and radiosurgery are non-invasive adjuvant treatments, but the toxicities from radiation are still being weighed against the benefits of tumor control (9). Because CNs usually have excellent prognosis when gross-total resection is achieved, radiation is not always indicated (2). Radiotherapy and radiosurgery have been adopted as an adjuvant treatment when gross-total resection cannot be achieved, the patient is inoperable, or the tumor is aggressive . A recent report suggests that fractionated radiotherapy after subtotal resection had a statistically significant higher tumor control rate and improved survival in adults (2). A higher 5-year progression free survival has also been shown for patients who received adjuvant fractionated radiotherapy after subtotal resection (67%) than patients without fractionated radiotherapy (53%) (10).

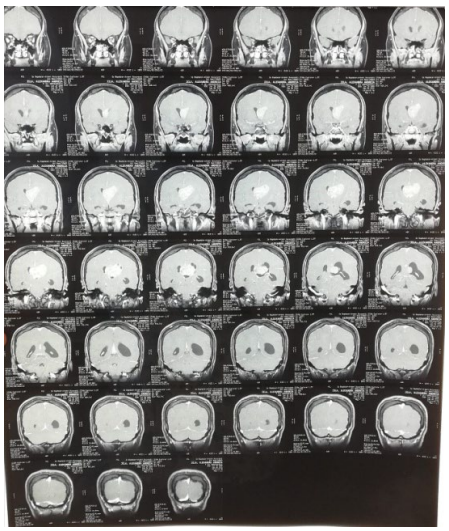
Prognosis: CN is a benign tumor, so it has an excellent prognosis. Surgery with gross total resection is the most preferable, correlated with the best long-term survival rates and local tumor control. Adjuvant radiotherapy may be considered for residual CN following subtotal resection, large CN size, or CNs near inoperable regions. Radiotherapy or chemotherapy the primary treatment for CNs has not been thoroughly examined.(2)

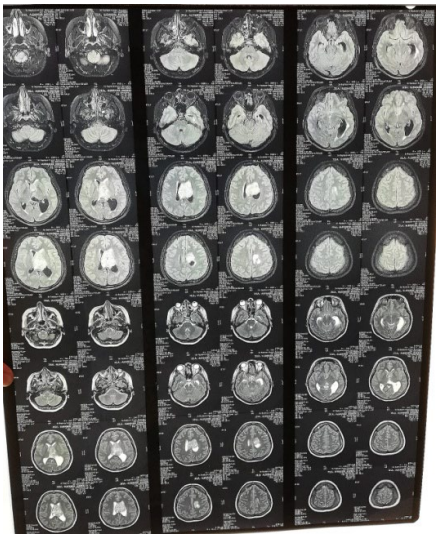
CASE PRESENTATION

This paper presents a case of a 21-year-old patient (having the approval of the Bioethics Commission no. 24389/ 28.06.2021) a personal history of headache, vomiting (appeared 2 weeks ago) and decreased visual acuity in both eyes was admitted on 07.09.2020 to the Neurosurgery Clinic III of THEBA. The patient was investigated both clinically and paraclinically.

IRM before surgery (09.09.2020) (Fig. 2, Fig. 3, Fig. 4.) revealed an extra-axial replacement process of space, developed intraventricular V III with extension in LLV and involving of the left foramen of Monro. The maximum dimensions are 52,6/39,3/25,4 mm (AP/TRA/CC). Mass effect on the lateral left ventricle that is dilated. The formation is MRI compatible with a thrid ventricle eppedimoma.

In 15.09.2020 total resection was performed, the patient having a favorable clinical evolution .





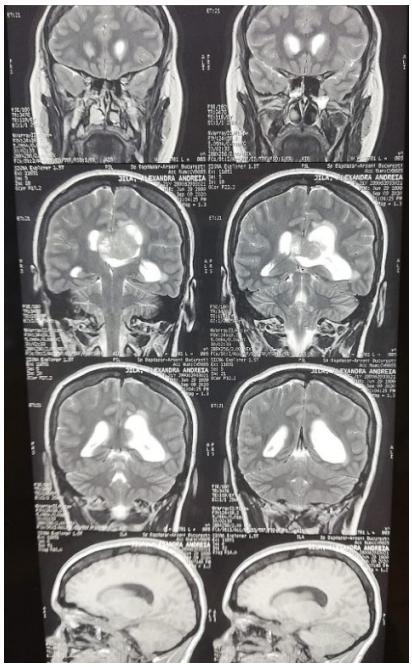


Fig. 2,3,4 IRM examination of our patient (from THEBA Neurosurgery Clinic III)

The anatomo-pathological examination revealed fragments of neoplastic tissue consisting of a proliferation of round cells, monotonous in appearance, solid in disposition, with nucleus with granular chromatin ("salt and pepper" appearance), without the presence of mitoses, zonal with clear cytoplasms with the appearance of a perinuclear halo . The cells are supported by a fibrillar stroma with the presence of isolated dendritic glial cells. Immunohistochimical tests: Synaptophysin - positive diffuse in tumor cells, NSE - positive diffuse in tumor cells, Neurofilament - negative, GFAP - negative in tumor cells, positive in glial cells in stroma, Ki 67 - nuclear proliferation index of about 2%, EMA -negative.
Conclusions: Histopathological aspects and IHC tests support the diagnosis of central neurocytoma.
After the surgery the patient's neurological status improved and she was addmited in the Neuromuscular Rehabilitation Clinical Division 05.10.2020-07.01. In our clinic, the patient initially followed a complex nursing program and subsequently a rehabilitation adequate program.
The reasons for admission included stage rehabilitation treatment, motor deficit of flaccid right hemiplegia, speech disorders of mixed aphasia, sphincter disorders.
Objective examination upon admission: she was normal weight, afebrile, with a good general state. The blood pressure was 100/60 mmHg, heart Rate 60/min and oxygen saturation 99% spontaneously , important muscular hypotonia on right limbs, post-operatory plague at frontal level, controlled urinary.
Neurological examination: temporal-spatial oriented, conscious, cooperative (execute some simple commands), motor deficit of right hemiplegia, mixed aphasia, central facial right paresis, decreased osteo –tendinous reflexes on right side, plantar skin reflexes are not obtained.
The patient was assessed functionally using the following scales:

- Spasticity = 0 on Ashworth modified scale
- Glasgow Outcome Scale-Extended (GOS-E): 3 points
- modified Rankin scale (mRs) : 5 points
- FIM (Functional Independence Measure): motor 36 points; cognitiv 6 points
- Functional Ambulation Categories(FAC) International Scale (Fig. 5): 0
- QoL (Life Quality Assessment Quality of Life), Montreal Cognitive Assessement (MOCA), Aphasia Screening Test (AST-Whurr) – NT (mixed aphasia)

From functional point of view, the patient was immobilized in bed.

Nr.	Categorie	Caracterizare
0	Nefuncţionalitate	Pacientul nu poate merge sau necesită ajutor de la 2 sau mai multe persoane
1	Dependenţă - nivel 2	Pacientul necesită sprijin ferm şi continuu din partea unei persoane
2	Dependenţă - nivel 1	Pacientul necesită sprijin continuu sau intermitent din partea unei persoane pentru ajutor cu echilibrul sau coordonarea
3	Dependenţă - supraveghere	Pacientul necesită coordonarea verbală sau ajutor potenţial din partea unei persoane fără contact fizic
4	Independenţă - pe teren plan	Pacientul poate merge independent pe teren plan, dar necesită ajutor la scări, pante sau suprafeţe denivelate
5	Independenţă	Pacientul poate merge independent oriunde

Fig 5. FAC (Functional Ambulation Categories) international scale – utilised in our clinic devision

Clinical and paraclinical evaluation

During hospitalisation, the patient presents short episodes of: anaemia normocrome corrected by medication, left anterior epistaxis treated under guideliness specified by ENT doctor, inflammatory phenomena in the left genian region treated specific antibiotics according to guideliness specified by OMF doctor (increased inflammatory markers) and the patient has been a contact with one person SARS-COV2 positive, so the patient was clinically evaluated and tested, the result being negative.

Computed tomography performed on 07.12.2021 (Fig. 6) shows: Examination performed for evolutionary control. Postoperative porencephalic cavity that communicates with the left lateral ventricle, which are large. No signs of tumor recurrence.

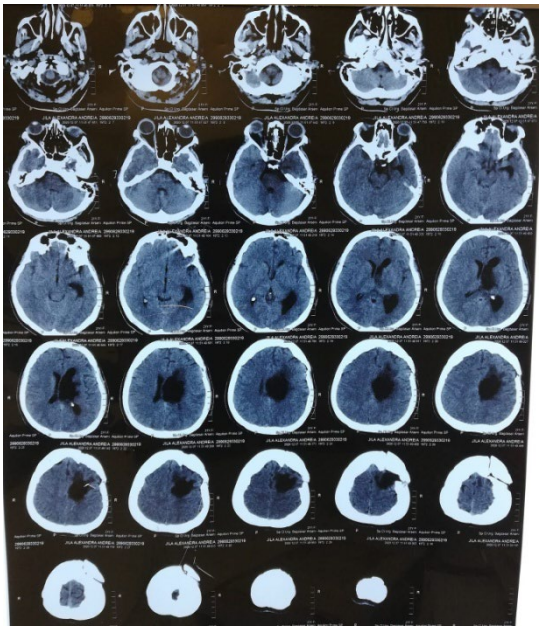


Fig. 6. Control CT examination on 07.12.2020 (from THEBA Neuro -Rehabilitation Clinic Devision)

Interdisciplinary evaluation

- ORL examination Left anterior epistaxis (vascular stain). Dapping with gelospon. Recommendations: blood count, blood coagulation. Avoid hot foods. Tarosin 4 cp / day. Emofix nasal ointment twice a day
- Ophtalmological examination: Eye bottom examination shows proeminent papillae discolored with blurred outline, emergence of vessels covered by edema extending peri- and parapapillary. Diagnsotic: papilloedema, optic atrophy, astigmatism, myopia. Recommendations: Mirtilene ginkgo 1 tb/day, Epinerv 1 tb/day, Acetazolamide 1 tb/day for 1 month after 1 tb/2 days for 1 month, Multiminerals 1 tb/day
- BMF examination: the left genian cellulite with starting point 26. Recommendations: Antibiotics: Amoxiplus 1,2 g/12 h iv for 5-7 days, Metronidazol 1g/12 h iv for 5-7 days and NSAID 1tb of 400 g x 2/day. Rigorous oral hygiene (brushing teeth after every meal).

Diagnosis

Based on the anamnesis data, on the clinical examination,and parclinical investigation the diagnosis is the following:

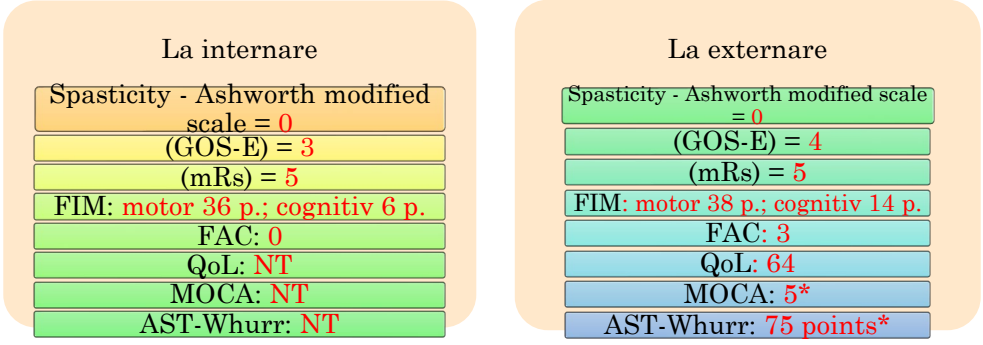
- *Right fllacid hemiplegia,*
- *Mixed aphasia after*
- *Surgical removal of left lateral ventricular tumor with extension in the right lateral ventricle and the third ventricle*
- *Partial optical atrophy*
- *Left genian cellulite*

Treatment

During hospitalization, the patient received complex treatment with: drugs (Injectable anticoagulant, anticonvulsant, cerebral anti-oedema, cerebral trophic, antibiothics, anti-inflammatory, symptomatics), physiotherapy: initially only in bed with a personalised program : passive movement at the joints level, active ones, active with left limb resistance, correct positioning in bed for prevention of vicious joints positions or thrombophlebitis, and after that at the physical therapy room: exercises at MotoMed bycicle, exercises for upper limb at pully (cuffs) and MotoMed, vertical support at wall bars, walking training with self-support in tetrapod walking stick, occupational and speech therapies, psychotherapy.

Evolution and clinical-therapeutical / recovery results

Regarding the clinical-therapeutic evolution, following the complex program of neuromuscular recovery, the patient presented a favorable evolution as can be seen from the comparative analysis we performed below through the evaluation scales, scales that allowed quantification. spasticity, motor deficit, ability in daily activities, ambulatory capacity, but also language disorder. It is noted that the 3 scales related to quality of life and language disorder, untestable at hospitalization, could be further tested subject to the fact that the patient has right hemiplegia and optic atrophy, which prevented her from obtaining a maximum score at written.



Evolution

The two photos (Fig. 7, Fig. 8) outline the patient's evolution. The first, the one on the left, was performed in November 2020, when the patient was mobilized in a wheelchair at the physiotherapy room. The positioning of the right upper limb in order to prevent edema is noted here. In the second photo, the one on the right, the patient also practices walking in a tetrapod-type cane, with the mobilization of the right upper limb, an exercise better highlighted in the videos that will follow.

Fig 7, 8. Evolution of our patient (November 2020 and June 2021) (from THEBA Neuro -Rehabilitation Clinic Devision)

CT examination 9 months after surgery (Fig.9) reveals a left frontal operative flap. The cortico-subcortical and deep porencephalic area is highlighted, which determines ventricular asymmetry with moderate left ventricular dilation - in a postoperative context. No signs of local tumor recurrence.

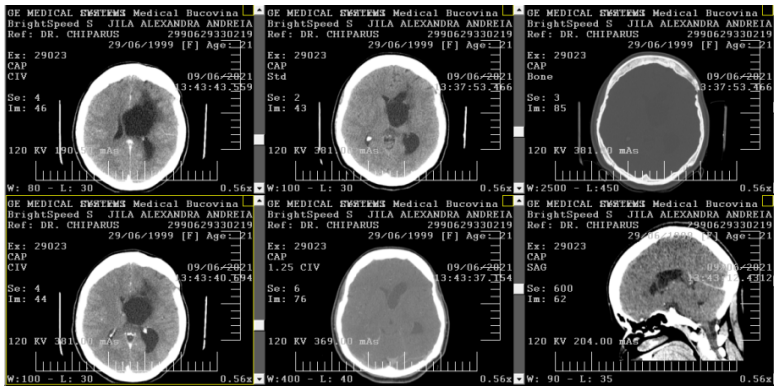


Fig. 9. CT examination at 9 months after surgery (from the patient's imaging investigations)

The patient had 3 admissions in our Clinic division (05.10.2020-04.11.2020, 06.11.2020-18.12.2020 and 21.12.2020-07.01.2021) where she benefited from a complex therapeutic program carried out by a multidisciplinary team presenting a favorable evolution proven especially after the evaluation through the scales mentioned above, with aphasia in remission and neurological improvement, being possible practicing walking with a tetrapod-type walking stick with supervision from another person. At the same time, the patient can use her right upper limb to perform feeding activities. Returned to the control 9 months after the operation in the Neurosurgery and Neuromuscular Recovery Clinics, we

mention the fact that a series of objectives were achieved such as the integration of the patient in the family, the possibility of household chores, but also the patient's contribution to the child's education process.

Prognosis

In our case the prognosis is favorable, except the at laborem prognosis who is reserved because the patient is currently practicing walking with a tetrapod-type walking stick, with the supervision of another person.

Ad vitam and ad functionem prognosis are favorable, provided that the recommendations from the discharge will be respected (drug treatment: anticonvulsant, antiaggregatory agent, cerebral trophic, recommended treatment by ophtalmologist, symptomatics, hygienic-dietary diet , avoiding emotions and conflicting states and limiting the use of telephone and television, she will continue the recovery treatment learned in the hospital (physiotherapy, speech and occupational therapy) (11-15).

Conclusion

It should be noted that neurosurgical intervention and pharmacological treatment, associated with an individual rehabilitation program consisting of: physical, occupational and speech therapies and also rehabilitation nursing interventions in a patient diagnosed with flaccid right hemiplegia and mixed aphasia after total surgical resection of the intraventricular central neurocytoma has improved the control of symptoms and the patient's quality of life

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