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Central Neurocytoma: About A Case

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Abstract

Neurocytoma is a very rare brain tumour. Immunohistochemistry made it possible to take a considerable step forward in its positive diagnosis. We report a rare case of neurocytoma in a 50-year-old adult with local extensions contraindicating optimal surgery. The patient received exclusive radiotherapy with a good response. This case highlights the feasibility of radiotherapy in the context of inoperable brain tumors with the maintenance of an acceptable quality of life.

Keywords- *Neurocytoma, Brain Tumor, Radiotherapy.*

Introduction

Neurocytoma is an extremely rare brain tumor discovered by Hassoun and collaborator in 1982. It represents 0.1 to 0.5% of primary brain tumors. About 600 cases have been described in the literature since 1982 **(**1,2**)**. These are neurons that form from neuronal cells in the septum pellucidum or the ependymal cavity and not from glial cells as in most brain tumors. The majority of central neurocytomas grow inward into the ventricular system forming intraventricular neurocytomas. In the majority of cases, it is a benign tumor, but sometimes it can be malignant. Surgical treatment is the cornerstone of treatment for central neurocytoma, but due to the deep location of the tumour, and despite advances in surgery, resections are frequently incomplete, hence the interest of radiotherapy.

Observation

We report the case of a 50-year-old patient, with no medical or surgical history, who presented balance disorders with a short walk associated with vertigo evolving for three years. The clinical examination showed a drop in visual acuity in the left eye at 8/10, associated with slight ataxia when walking, segmental muscle function rated at 5/5 in all four limbs with retained sensitivity. Fundus examination revealed no papilledema.

Magnetic resonance imaging **(**MRI**)** of the brain showed an intraventricular tumor process of the 4th ventricle measuring 52 mm in height, 37 mm in anteroposterior diameter and 32 mm in width **(**Figure 1**)**. It has hemorrhagic and calcic stigmata. This intracerebral process has the following relationships:

- At the top, it continues in the aqueduct of Sylvius and in the 3rd ventricle up to the roof of the latter. It also comes into contact with the infero-internal wall of the right VL which is pushed back and invades the quadrigeminal cistern by including the pineal gland, it comes into contact with the quadrigeminal tubercles, without a separation border. It presents a bilateral contact with the free edge of the tent of the cerebellum. There is an intimate contact passing under the two internal cerebral veins. It drives back with contact outside with the posterior cerebral arteries.

- Below: there is no clearly visible extension in the foramen of Magendie.

Backward: the process has intimate contact with the upper vermis without invasion.

- Laterally: no clearly visible extension in the foramina of Luschka.

It presents a fuzzy and discreetly irregular interface in places with the cerebral parenchyma at the level of the brainstem without clearly visible signs of invasion **(**Figure 2**)**.

A stereotactic biopsy is performed when it is impossible to perform an excision. The anatomo-pathological report objectified round cells with a high nucleocytoplasmic ratio which are arranged in diffuse sheets or around vascular structures by outlining pseudo rosettes. Cytonuclear atypia was marked with dense chromatin and mitoses suggesting first a high-grade ependymoma **(**grade 3**)**.

An immunohistochemical complement carried out on the same samples had not objectified expression of Olig2 or of the epithelial membrane antigen or of GFAP in the presence of a positive internal control, on the other hand a strong expression of synaptophysin **(**Figure 1**)** on the tumor cells has been objectified as well as the presence of many cells that also express neurofilaments. Ki67 was weakly expressed by less than 10% of the cells.

The patient's file is then discussed in a multidisciplinary neuro-oncological consultation meeting and the decision was to perform exclusive radiotherapy.

The patient then benefits from a centering scan in a thin millimeter section going from the vertex to the second thoracic vertebra. The images are transferred to the TPS then the delineation of the organs at risk as well as the target volumes was carried out after fusion-registration with the initial MRI.

Our patient received three-dimensional conformal external radiotherapy at a total dose of 54 Gy in 27 fractions of 2 Gy **(**Figure 3**)**. Ballistics required photon beams of 6 MeV. The spread was 42 days including

2 days of stoppage for a breakdown of the treatment machine. We did not find any toxicity of grade greater than two during the treatment.

A post-therapeutic evaluation by MRI four weeks after the end of irradiation notes a regression of more than 80% of the tumor.

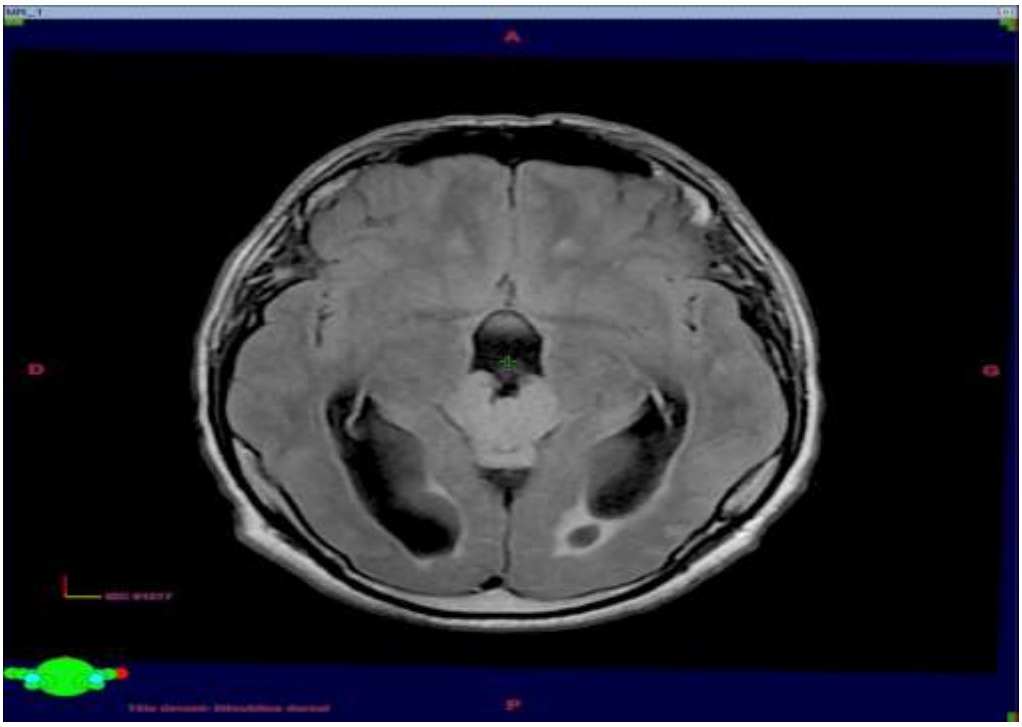

Figure 1: pre-therapeutic MRI image of our patient.

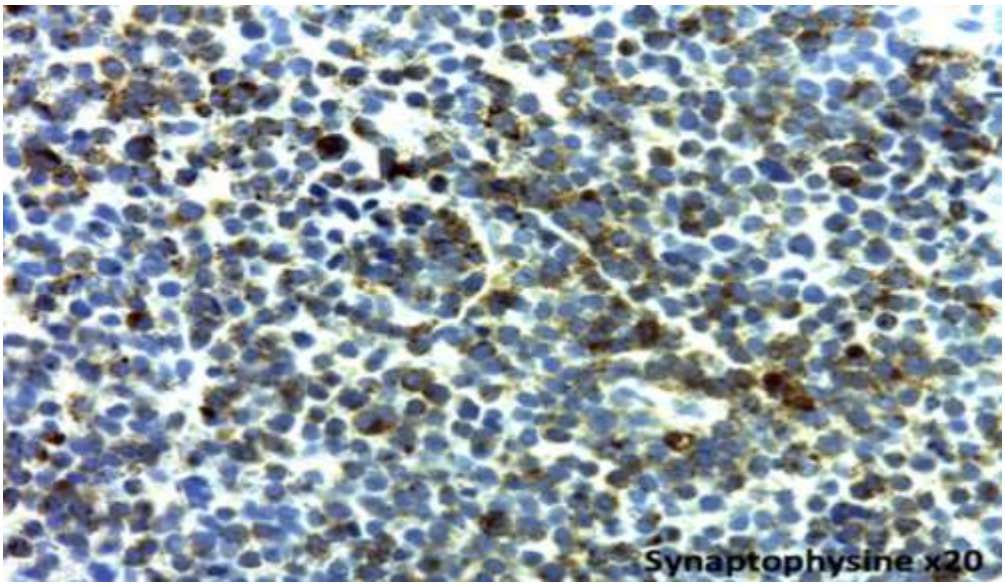

Figure 2: X20 magnification of synaptophysin labeling.

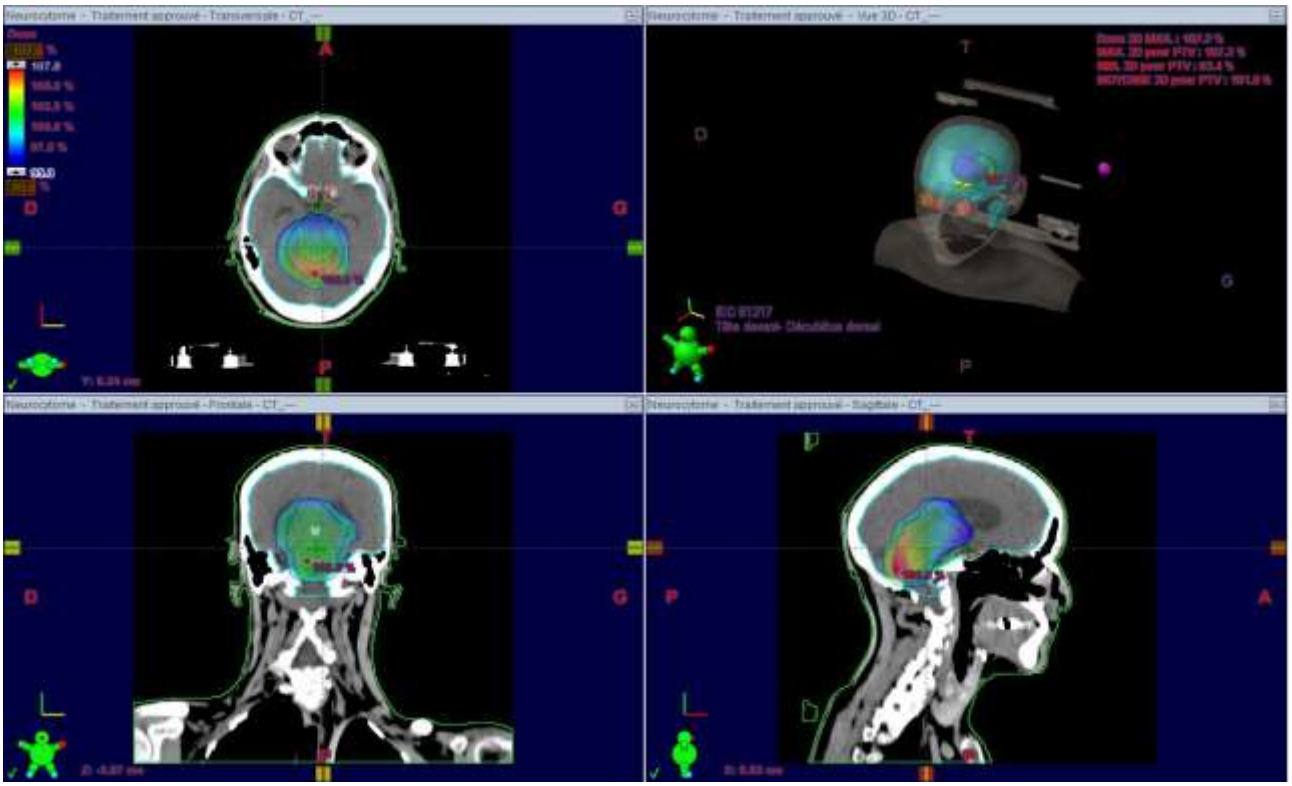

Figure 3: dosimetric image of our patient.

Discussion

Neurocytomas are usually discovered by a syndrome of intracranial hypertension secondary to obstructive hydrocephalus **(**3**)** as in the case of our patient. Other signs can be observed: a decrease in visual acuity with papilledema, gait disorders, neurological deficit or epileptic seizures **(**4**)**, which shows an aggressive tumor.

Histologically, the central neurocytoma can pose diagnostic problems given the resemblance to other clear cell brain tumors. In optical microscopy, it presents small cells with rounded or regular nuclei, with clear cytoplasm with absence of mitosis **(**4**)**. However, in atypical forms, mitotic activity is variable, vascular proliferation and tumor necrosis can be observed **(**4**)**.

Definitive diagnosis is only made using immunohistochemistry, which confirms the neuronal origin of the tumor by the expression of synaptophysin, neuron-specific enolase **(**NSE**)**, microtubule-associated protein 2 and glial fibrillary acidic protein **(**GFAP**)**. The use of neuronal nuclear antigen **(**NeuN**)** has been described and appears to be sensitive and specific in the diagnosis of central neurocytoma **(**5**)**. Electron

microscopy confirms the diagnosis by showing neurosecretory granules, neuronal extensions, the presence of microtubules and synaptic formations.

On computed tomography, the central neurocytoma is described as a well-circumscribed isodense or hypodense lesion with mild to moderate contrast enhancement [6]. Classically, there are intratumoral calcifications as well as cystic areas. MRI shows in T1 sequence a tumor in isosignal and heterogeneous in T2 with variable intensity as well as calcifications and cysts in hypersignal [7]. Contrast uptake after injection of gadolinium is variable [8]. SpectroMRI shows in the central neurocytoma an increase in the peak of glycine, choline and alanine [9] and a decrease in the peak of N-acetylaspartate **(**NAA**)**, and seems interesting for the differential diagnosis [10]. In comparison with gliomas. For the central neurocytoma, the choline/creatinine ratio is higher, the NAA/choline ratio lower. A correlation between the choline peak and the degree of malignancy has been described [11]. Diffusion MRI shows a lower apparent diffusion coefficient value [12]. This dynamic study could not be done in our patient and would have made it possible to better refine the diagnosis from the outset.

The treatment is surgical and the most complete resection possible must be performed without causing great morbidity, which could not be achieved in our patient.

Exclusive external radiotherapy has been little studied. No study has compared surgery alone and external radiotherapy after biopsy with the idea of a less invasive strategy. Published data on external radiotherapy modalities are based solely on three-dimensional conformal irradiation studies with a total dose of 50 Gy in conventional fractionation.

Conclusion

Central neurocytoma is a rare tumor of neuroepithelial origin. It is increasingly reported in the literature since the introduction of immunohistochemistry in the diagnosis of brain tumors. Radiotherapy is an option of choice in the face of unresectable neurocytoma to avoid complications related to surgery.

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