

Primary cerebellar glioblastoma in a patient with neurofibromatosis type I - case report in military central hospital, Mexico City

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Abstract

Glioblastoma multiforme (GBM) is common in the cerebral hemispheres, most of them may be secondary to anaplastic astrocytomas, however, GBM in the cerebellum is a rare finding. We present the only case reported in our hospital of cerebellar glioblastoma in a 75 years old patient, the former was removed subtotally. We reviewed the literature, the association between NF1 and high-grade gliomas, aspects of Neuroimaging, as well as histopathological findings.

Keywords: glioblastoma, cerebellar, primary glioma, neurofibromatosis, posterior fossa glioma

Introduction

Glioblastoma represents 15-20% of all intracranial tumors and approximately 50% of all gliomas in adults,¹⁻³ the annual incidence of malignant gliomas is 3-5/100,000 with a slight predominance in the male sex. Malignant gliomas can develop at all ages with peak incidence in the fifth and sixth decade of life.^{4,5} These tumors are located more frequently in the cerebral hemispheres, basal ganglia, thalamus, corpus callosum, and they infiltrate the white matter and/or in the depth of the gray matter. They develop secondary to a diffuse anaplastic astrocytoma, however they can arise from primary form⁶ and rarely they can appear in the posterior fossa.^{1,7} It was confirmed by clinical history, physical data in the patient and literature that was revised the association between Neurofibromatosis type 1 and the risk of developing high-grade gliomas,⁸ which is why we will present the present case of cerebellar glioblastoma of the posterior fossa in an adult.

Case report

A 75-year-old male patient presented at our hospital with decreased alertness, drowsiness, nausea, vomiting and oral intolerance of 1 month of evolution, adding up inability to walk for a week of evolution. In patient's history there is Neurofibromatosis type 1 what is the base diagnosis for this, diagnosed at the age of 21 years and a radical prostatectomy for the treatment of prostate cancer in September 2013.

On physical examination, the patient appeared drowsy, dehydrated and was unable to walk. He also presented bilateral papilledema,

hyporeflexia of the pupils, dysarthria, generalized pallor, bilateral presbycusis, and multiple (numerous) fibroids (Figure 1).

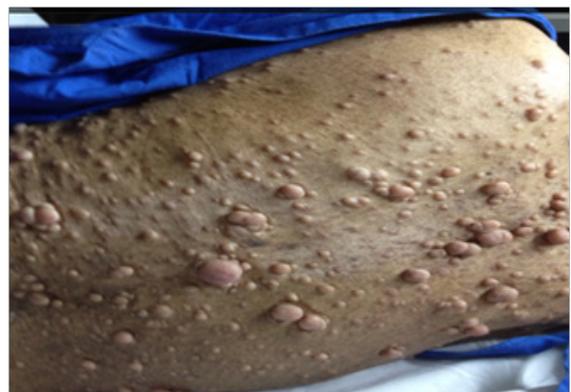


Figure 1 Patient with neurofibromatosis Type I; generalized neurofibromas are observed.

On admission, a simple skull computed tomography was performed, showing a hyper dense heterogeneous tumor in the posterior fossa, with peritumoral edema. In addition, a peripheral hypo dense area in the right cerebellar hemisphere was observed, which one that was making compression of the perimesencephalic cisterns. Likewise, an increase in the size of the ventricular system was observed, accompanied by subependymal edema (Figures 2) (Figure 3), leading to the radiological diagnosis of an acute non-communicating

hydrocephalus. Computed tomography with contrast of the skull was performed, showing heterogeneous lesion located in the cerebellar hemisphere (Figures 4) (Figure 5).

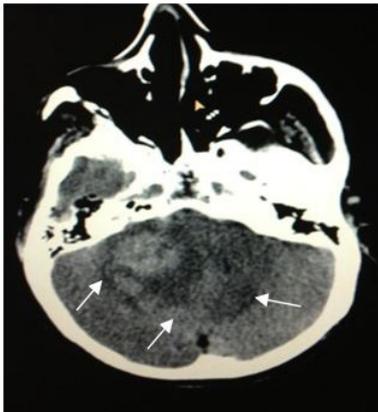


Figure 2 Computed tomography of the skull, axial section (5mm).A heterogeneous, irregular lesion, in the right cerebellar hemisphere. Presence of peri-lesional edema, as a result of the local volume increase.

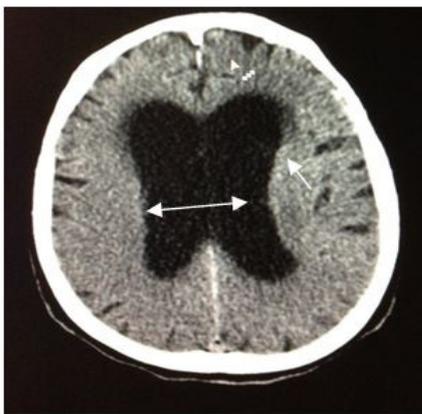


Figure 3 CT scan of the skull, axial section (5mm).Increase in the size of the ventricular system, with subependymal edema → Radiologic diagnosis of an acute non-communicating hydrocephalus.

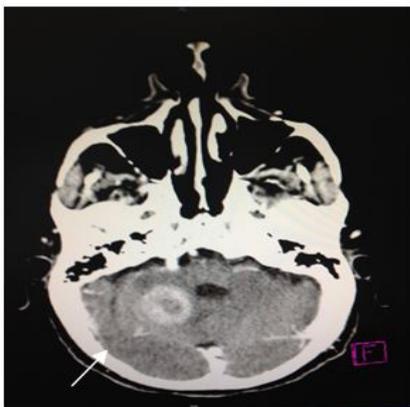


Figure 4 CT scan of the skull, axial section (5 mm).A heterogeneous image, with hypodense center and moderate reinforcement, in the right cerebellar hemisphere.

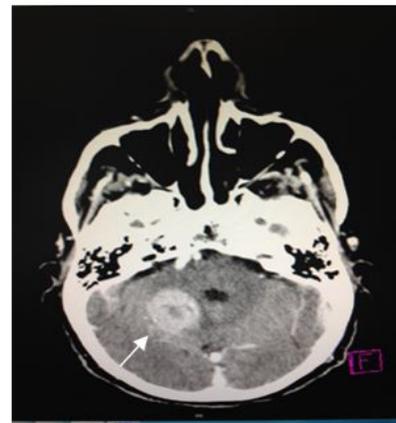


Figure 5 CT scan of the skull, axial section (5 mm).A heterogeneous image that reinforcement after the administration of contrast medium with necrotic center, it is pointed by the arrow.

A ventriculo-peritoneal shunt valve was placed without complications, resolving acute hydrocephalus and allowing the patient’s placement to the intensive care unit for postoperative monitoring.

Magnetic Resonance (1.5T) of the skull was performed with a contrast medium. Axial, coronal and sagittal slices, in sequence T1 and T2 were acquired (Figures 6–8). Axial T1 images with Gadolinium as contrast medium were also carried out (Figure 9). In addition it was made a spectroscopy with univoxel technique with TE of 144 msto see the peaks of N-acetylaspartate, creative, chorine and lactate (Figure 10).

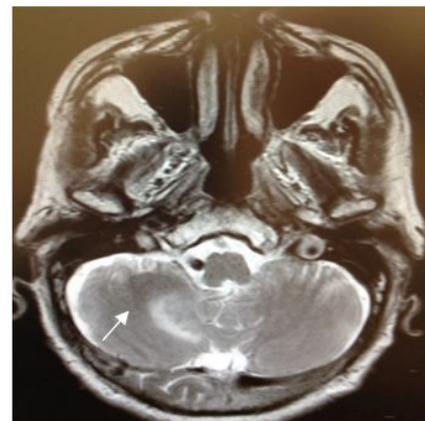


Figure 6 Magnetic Resonance imaging of the skull, in the axial T2 sequence, where a heterogeneous lesion with perilesional edema, located in the right cerebellar hemisphere, is observed.

The surgical procedure for tumor removal (partial resection of the tumor) and later on for histopathological examination, consisted of a subtemporal transtentorial approach with conventional microsurgical technique, without complications. However, 6 days after the surgical procedure, the patient presented generalized deterioration, becoming complicated with nosocomial pneumonia and despite treatment based on an antibiotic therapy, oxygen therapy with support of mechanical ventilation, and measures against cerebellar edema, the patient finally presented cardio respiratory arrest and died without having been given an adjuvant treatment. The histopathological examination of the surgical specimens diagnosed a Glioblastoma (Figures 11A–C).

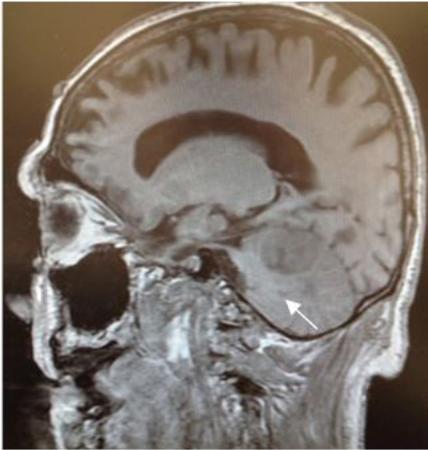


Figure 7 Simple sagittal T1 image with a hypointense tumor in the right cerebellar hemisphere

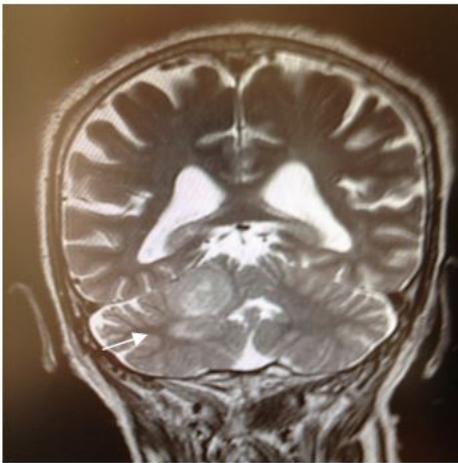


Figure 8 Coronal T2 image T2 showing a heterogeneous lesion with perilesional edema, located in the right cerebellar hemisphere, adjacent to the tentorium cerebelli.

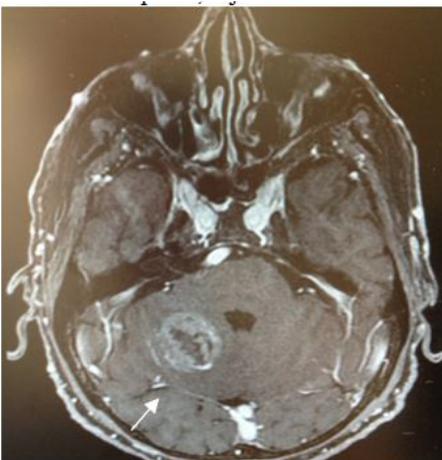


Figure 9 Axial T1 image after contrast enhancement. Shows an heterogeneous tumor reinforcement, in the right cerebellar hemisphere with hypointense and necrotic center.

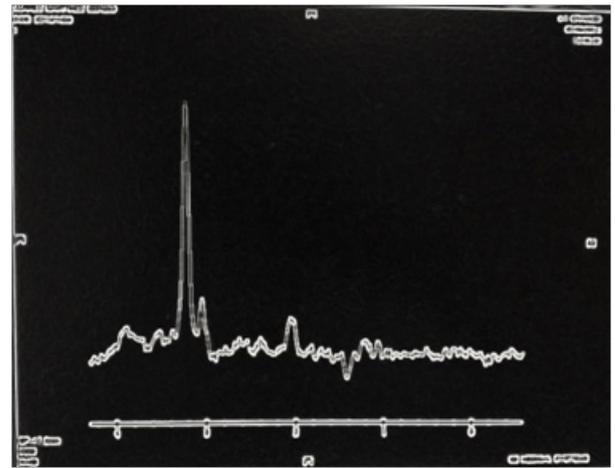


Figure 10 Spectroscopy with univoxel technique, where the elevation of the peak of choline (up to 5.1 times) and the decrease in the value of N-acetylaspartate, with a small peak of lactate, is observed.

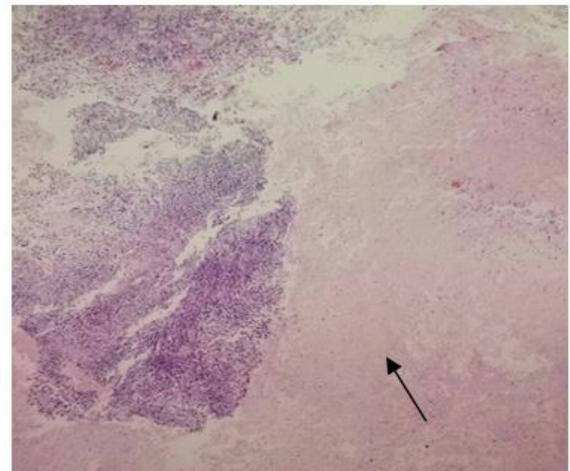


Figure 11A H&E stain of the tumor showing extensive tumor necrosis (H&E; X40).

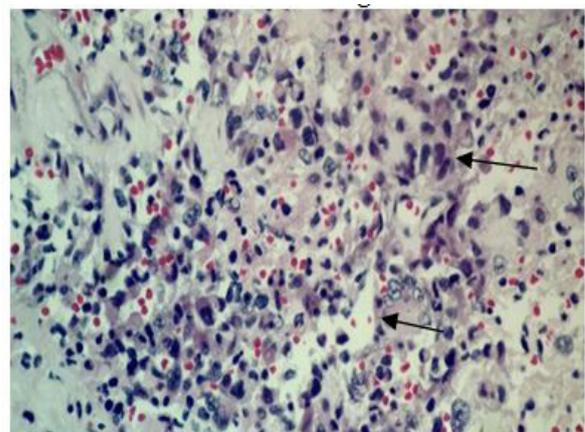


Figure 11B H&E stain of the tumor showing multinucleated neoplastic cells along with vascular proliferation (H&E; X400).

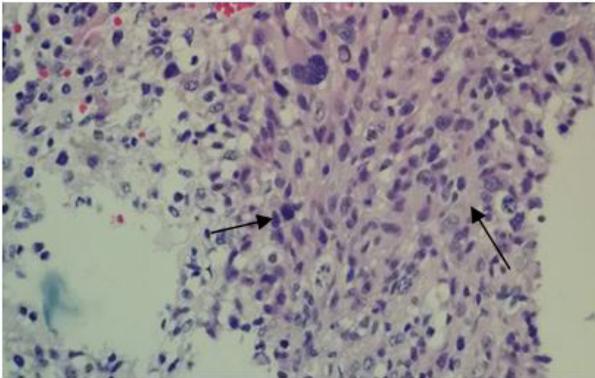


Figure 11C H&E stain of the tumor showing neoplastic tapered cells with eosinophilic fibrillary cytoplasm. Moderate nuclear pleomorphism can also be observed.

Conclusion

Glioblastomas of the posterior fossa remain very rare tumors.^{1,9} Our team takes the opportunity to present this case publication, since this is the first report (in our database) of a primary Glioblastoma (WHO Grade IV) in an adult with a cerebellar localization. In our case, the time between diagnosis and the onset of complications was very short, thus could not be put in the average survival time of 6-8 months as mentioned in some publications.⁵ The symptoms presented by the patient were more frequent (nausea, headache, vomiting, ataxia, confessional state) as mentioned in the literature.^{10,11}

Differential diagnoses in posterior fossa lesions should consider metastatic lesions, cerebellar infarction, cystic astrocytomas, abscess, hemangioblastoma, and entities such as encephalitis, tuberculoma, or multiple sclerosis.^{12,13} Patients with a history of NF1 have a 5 times higher risk of developing glioblastoma than the general population.⁸

Spectroscopy of astrocytomas shows a reduction in NAA due to loss of neuronal elements, and elevation of the peak of Choline that may reflect the increase in membrane synthesis, cell membrane proliferation. The presence of the lactate peak correlates with the high degree of malignancy observed in Glioblastomas.^{14,15}

Acknowledgements

None.

Conflict of interest

The author declares no conflict of interest.

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