

CEREBELLAR GLIOBLASTOMA MUTIFORME IN AN ADULT

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Eventhough supratentorial glioblastoma multiforme (GBM) is the most frequent primary brain tumor in adults, its localization in the cerebellum is extremely rare¹. It is very important for the neurosurgeon to make the differential diagnosis between the cerebellar GBM, and other diseases like metastasis, anaplastic astrocytomas, and cerebellar infarct because their treatment modalities, prognosis, and outcome are different². Clinical manifestations of these diseases are very similar, so making an appropriate diagnosis is fundamental for the management of each of them³.

We describe a case of cerebellar GBM managed at our institution reporting its clinical presentation, radiological and histological findings, and treatment instituted.

CASE

A 65 year-old male patient was admitted with a clinical history of headache, mental confusion, left hemiparesis, gait and trunk ataxia, and dismetry. The symptoms had been worsening on the last five months. Past medical history was significant for previous cerebral ischemic event without persistent deficits, and an emergency laparotomy for a bleeding gastric ulcer was done one year before and aortic ectasy.

An investigation with brain CT and MRI scan, showed an heterogenic contrast enhancing expanding process in the vermis and left cerebellar hemisphere. The MRI showed an intra-axial, infiltrative, heterogenic mass localized in the vermis with extension to left cerebellar hemisphere, left superior cerebellar peduncle and brainstem. The lesion was isointense in T1-weighted images, hyperintense in T2-weighted images, showing enhancement after paramagnetic contrast administration. Its dimensions were 4.8 cm × 3.6 cm × 2.6 cm. The lesion compressed the IVth ventricle, and caused hydrocephalus (Fig 1A).

Abdominal ultrasound, thoracic imaging and investigation for metastasis were negative.

The patient was submitted to microsurgical resection of the lesion through a cisternal craniectomy and transversian approach. The surgery was uneventful but a subtotal resection was performed because the tumor infiltrated the superior cerebellar peduncle and brainstem (Fig 1B). Postoperatively, the hydrocephalus diminished, but an occipital pseudo-meningocele

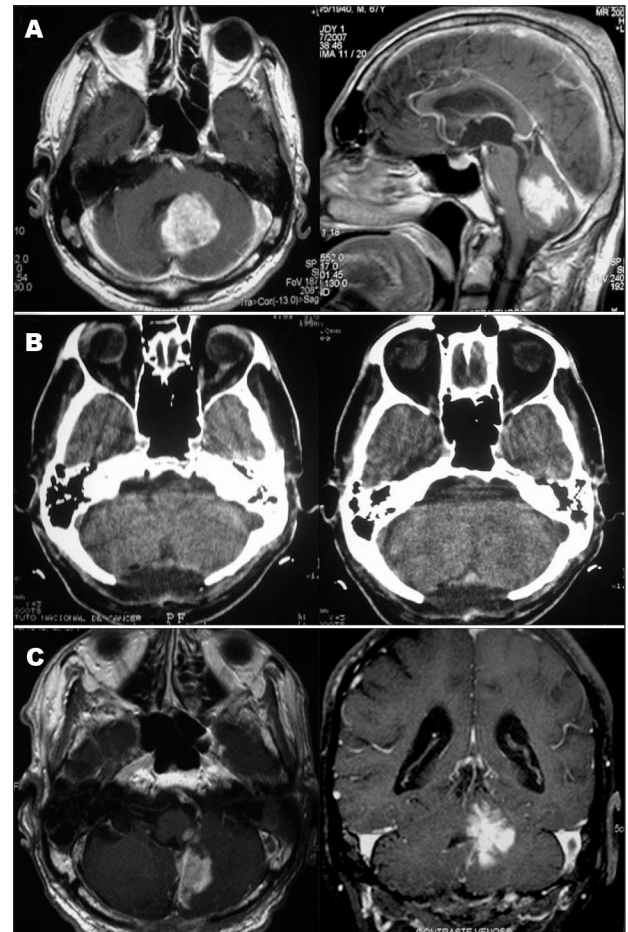


Fig 1. (A) Pre-operative MRI showing tumor infiltrating the left superior cerebellar peduncle, and compressing de IVth ventricle. (B) First day pos-operative CT showing the craniectomy site, and no hemorrhage on the tumoral bed. (C) Control MRI six months after surgery showing recurrent tumor in the left cerebellar hemisphere invading the superior cerebellar peduncle and brainstem.

originated another surgery for its closure. Control MRI six months after the first surgery shows residual tumor without signs of disease progression (Fig 1C).

Histological examination showed a poor-differentiated glial neoplasm with intense pleomorphism, nuclear hyperchromasy, and necrosis (Fig 2). The histological diagnosis was GBM.

The case was eligible for cranial radiotherapy starting for-

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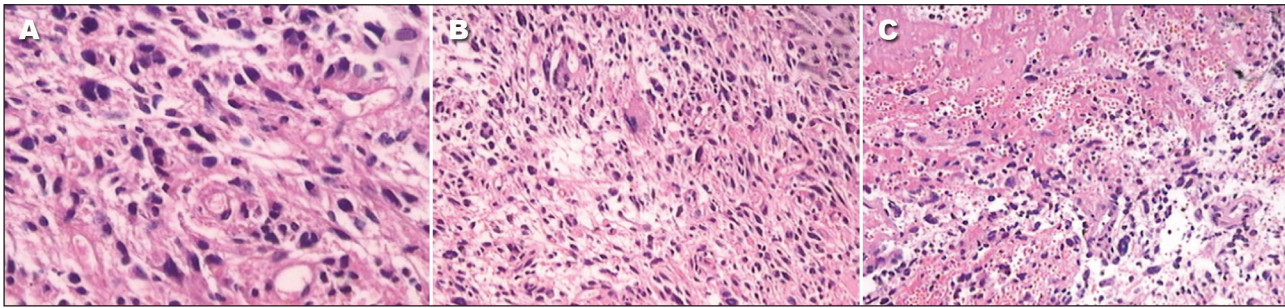


Fig 2. Light microscopy of the surgical specimen. H-E staining. (A) Glial neoplasm with intense cellular pleomorphism (100x objective). (B) Intense nuclear pleomorphism and hyperchromasia (200x objective). (C) Tumor areas with necrosis (100x objective).

ty days after surgery. Considering the age and the performance scale the patient was not considered candidate to chemotherapy in our Institution. Since then, the patient remains on ambulatory follow-up. Eight months after tumor resection he is clinically stable. The mental confusion and left motor deficit diminished. Severe trunk and gait ataxia however still remains.

The patient provided informed consent agreeing with this case publication and the case study was approved by the Ethic Committee of the National Institute of Cancer – Brazil.

DISCUSSION

Primary cerebellar GBM in adults is a rare entity. It accounts for less than 1% of all GBM⁴. The most common clinical findings include headache, nausea, and vomiting, because of intracranial hypertension, and manifestations of cerebellar dysfunction⁵. Adults with infratentorial GBM have a median overall survival (OS) of approximately 10 months, with a 1-year OS of 38%⁶. In children the prognosis is also poor, with a mean survival time of 9.9 months. This grim prognosis resembles that related to the supratentorial GBM. The patient presented here is still under ambulatory follow-up, eight months after surgery. The MRI demonstrates recurrent infiltrative tumor, and his clinical condition continued stable.

Considering the different treatment modalities available it is very important to make a proper differential diagnosis between infratentorial GBM and metastasis, hemangioblastoma and even cerebellar infarct with contrast enhancement³. In our case we initially suspected of metastasis but the histological examination confirmed the diagnosis of the rare neoplasm presented here.

Considering malignant glial tumors, it would be always reasonable to attempt a gross-total tumor resection; however the infiltrative nature of the GBM makes it difficult to achieve it many times⁷. The infratentorial compartment has the special problem of brainstem invasion, which is a known independent prognostic factor affecting survival. In our patient we did not tried a gross-total resection because of superior cerebellar peduncle and brainstem infiltration

by the tumor. With this strategy we did not added deficits to the patients, even though severe ataxia persisted.

Tumor recurrence is almost always local. In very few cases reported there are also extra-cerebellum failure, including spinal seeding through the CSF. The use of radiation therapy for malignant gliomas is a well established treatment; however it remains a matter of debate whether it should be delivered only to the posterior fossa, to the whole brain, or to the neuraxis⁸. The role of chemotherapy is not fully established, but is the only reasonable complementary therapy for younger children. Considering the fact that the infratentorial GBM histological characteristics are equal to the supratentorial ones, it seems reasonable to use concomitant and adjuvant chemotherapy in its treatment⁹.

Even though difficult to confirm, there is a theory that the rarity of cerebellar GBM can be explained by a lesser tendency of cerebellar astrocytes to suffer malignant transformation¹⁰. Notwithstanding these observations, the fact is that infratentorial GBM is a very malignant tumor, with rapid onset of symptoms, and poor prognosis. Best treatment for infratentorial GBM seems to be the same for supratentorial GBM: the most radical surgery possible, followed by radiotherapy, and chemotherapy.

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