Case Report

Rare Case of Posterior Fossa Glioblastoma Multiforme

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ABSTRACT

In general, Glioblastomas multiforme frequently occurs in supratentorial region but in less than 4% occur in the posterior fossa predominantly in the cerebellum.We presents case of male with posterior fossa glioblastoma. MRI is the study of choice with high sensitivity and specificity for diagnosis, after that they underwent biopsy and the results of pathology described GBM. The treatment of election is surgerys,radiotherapy plus chemotherapy while the prognosis is poor even with treatment, thats is why we need to identify new therapeutic strategies; We currently believe it is necessary to use genetic platforms to identify possible therapeutic targets.

INTRODUCTION

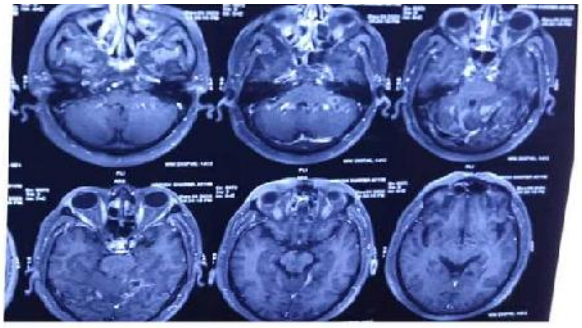
Glioblastoma (GBM) is the most frequent tumor of the central nervous system representing 50% - 60% of all brain tumors, they occur most frequently from 50 - 70 years and their most frequent location is supratentorial; posterior fossa glioblastomas are very rare and represent a challenge for diagnosis since they are confused with metastatic lesions, this location correspond to 0.24% to 1% of the total glioblastomas. GBMs located in the posterior fossa can cause various cerebellar symptoms such as headache, gait disorders, ataxia, nausea and vomiting. These findings may suggest the existence of a massive lesion in the posterior fossa. Surgery remains the treatment of choice if possible; followed by adjuvant treatment with Temozolomide (TMZ), -Radiotherapy in the case of a recurrence palliation remains the alternative of choice including biological therapy. The presentation of glioblastoma in cerebellum is extremely rare, and few reports of clinical cases with these characteristics have been published. Its etiology and prognosis are poorly clarified due to lack of data in the literature. We present clinical cases of glioblastoma with location in the posterior fossa.

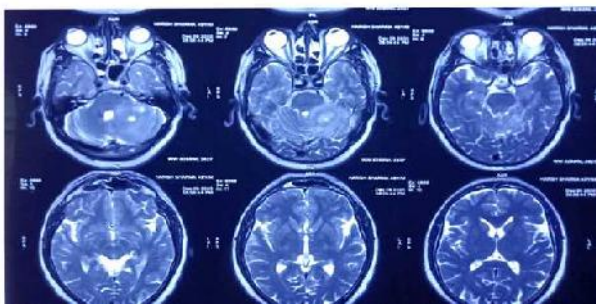
CASE

48 year male history of sudden imbalance while walking since 10days History of tremors in left hand since 10 days Neurology

- Gait having appendicular ataxia
- Pronator test left upper limb outwards
- Left side dysdiadochokinesia
- Dysmetria
- Intention tremors
- Horizontal nystagmus more on left side
- Tandem gait absent
- No staccato speech

All cranial nerves within normal limits





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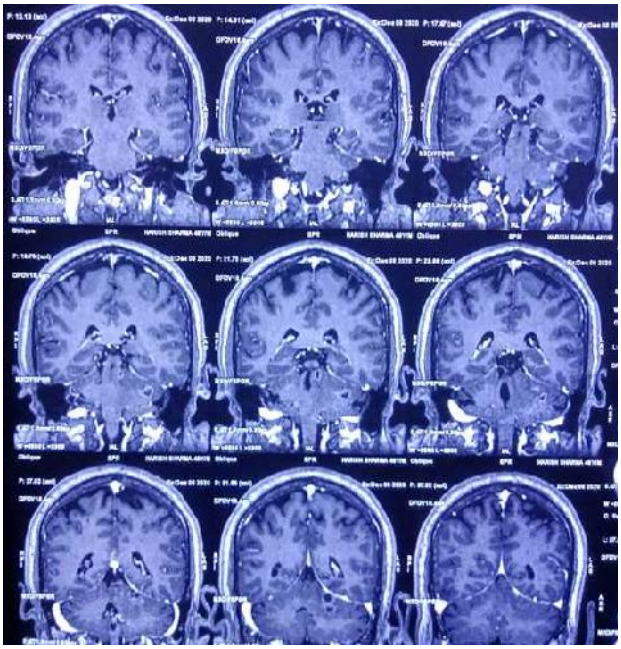
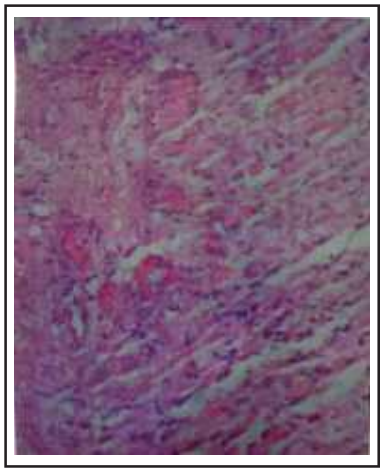


Patient was operated in prone position and skin incision kept over left paramedian region and after muscle incised and suboccipital craniectomy done,dura incised in cruciate fashion and transcerebellar approach was planned and tumor excision done gradually from petrosal part of cerebellum which seem intraoperatively to be either tuberculoma or metastasis; but on biopsy report came out to be glioblastoma multiforme.



HISTOPATHOLOGY
(GLIOBLASTOMA MULTIFORME WHO GRADE 4)

DISCUSSION

Glioblastoma, the most frequent tumor among all primary tumors of the central nervous system in adults, has a frequency of 50%. However, adult cerebellar is extremely rare, accounting for 0.24% to 3.8% of all intracranial glioblastomas. From 1975 till 190 articles and abstracts about cerebellum glioblastoma were published, according to a search of the Medline database. The male-to-female ratio is 2:1. Cerebellar glioblastoma can be seen in all age groups. About 70% of lesions occur in adults with a median age of 46.7 years while 30% were noted in children (Demiret al2005; Mattos et al 2006). As with our patient, localization is generally median or paramedian with a possible extension to the fourth ventricle.

The clinical features of patients with cerebellar GBM are similar to those of other aggressive fast growing infratentorial tumors. Signs and symptoms include headache, nausea, vomiting, and cerebellar dysfunction including ataxia, imbalance and unsteady gait. Non-enhanced CT scan findings of GBM may include a heterogeneous poorly marginated mass; internal areas of low or fluid attenuation that are the foci of necrosis (present in as many as 95% of GBMs); internal areas of high attenuation that are the foci of hemorrhage or, rarely, calcifications. There may be significant mass effect and perilesionaledema. Enhanced CT scans display significant enhancement with findings such as irregularity and heterogeneity.

The imaging features of cerebellar GBM are described as nonspecific. Lesions may occur laterally in the cerebellar hemispheres or in the midline within the vermis. The lesions are typically infiltrating with indistinct margins. Signal characteristics are heterogenous, often with necrotic and cystic components. A thick and irregular wall is commonly seen. However, irregular peripheral enhancement is consistently described following contrast administration. Edema is usually present and obstructive hydrocephalus is common. This is in contradistinction to the imaging findings in our case where the features of perilesional edema and contrast uptake were sublte.

Additionally, MRI has a highest degree of confidence in the diagnosis of glioblastoma multiforme (GBM; malignant glioma). MRI findings demonstrate a heterogeneous mass that is generally of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.There are internal cystic areas, areas of high signal intensity on T1 (hemorrhagic foci), neovascularity, necrotic foci, significant peritumoralvasogenic edema, and significant mass effect. Irregular but intense enhancement after the administration of gadolinium-based contrast material (same pattern as with enhanced CT scanning) is also

found. However, the patient being presented did not conform to these described findings. Magnetic resonance imaging (MRI) demonstrated a midline cerebellar mass that was hyperintense on T1 with minimal adjacent edema, blooming on gradient sequence and no restriction with diffusion. The mass had minimal enhancement with gadolinium contrast.

The histology and biology of cerebellar GBM is similar to that of cerebral GBM . This includes malignant tumor cells, mitoses, hypercellularity, pleomorphism and neoangiogenesis. The presence of necrosis helps differentiate GBM from anaplastic astrocytoma or from well-differentiated astrocytoma (Luccarelli et al 1980; Georges et al 1983; Katz et al 1995; Rizket al 1994). The case being presented exhibited these features as well as considerable nuclear and cytoplasmic pleomorphism, with multinucleated, giant cells and hemosiderin-laden macrophages.

As with any GBM and any malignant brain tumor, cerebellar GBM has a very poor prognosis. This is attributed to rapid tumor progression, locally aggressive behavior as well as the common findings of CSF pathway spread. Early intervention including aggressive surgery as well as aggressive radiation and chemotherapy have been advocated to increase the disease free interval and to prolong survival. Despite these measures; however, survival of patients with cerebellar GBM is very poor, in the range of 3-22 months.

This case is an unusual presentation due to the presence of significant hemorrhage, well-defined margins, minimal contrast enhancement and minimal edema. There were few features helpful in making the correct specific prospective diagnosis of glioblastoma multiforme. However, GBM should be included in the differential diagnosis of a hemorrhagic infratentorial mass with rapid progression of clinical findings as well as imaging findings considered atypical for the common entities that occur in the posterior fossa.

CONCLUSION

In general, the presentation of glioblastomas occurs in the supratentorial region but in less than 4% of cases occur in the posterior fossa particularly in the cerebellum, there are very few cases reported. Its pathogenesis and prognosis are not yet fully clarified, there are some data that suggest a worse prognosis. The differential diagnosis with metastatic posterior fossa tumors or cerebellar absces is a challenge for the doctor due to the similarity in the location, for this the magnetic resonance imaging (MRI) is mandatory as well as the pathological anatomy and the molecular diagnosis. The treatment for these lesions is like the supratentorial GBM, knowing that surgical resection is conditioned by its location and the implications in neurological sequelae, which is why it is very limited.

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