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Case Report

2014 Posterior Fossa Ganglioglioma: An Unusual Neoplasm in a Rare Location

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Abstract

Introduction : Ganglioglioma is a primary neoplasm of the central nervous system that typically affects young individuals, accounting for 0.4 to 9% of all brain tumors. Gangliogliomas are rarely found in the infratentorial compartment. In this article, we report on this case and make a brief review of the literature on the subject.

Case Report : We present a case of a 36 year-old female patient with an 18-month history of headache and dizziness. Imaging studies showed a cerebellar lesion, with compression of the fourth ventricle. The patient underwent surgical excision of the lesion, of which histopathological analysis disclosed a ganglioglioma. The patient presented good evolution and was discharged with no new deficits.

Conclusion : The ganglioglioma represents an uncommon neoplasia. Its occurrence in the posterior fossa is even more striking. The best treatment consists of total excision of the lesion.

Keywords : Brain neoplasms; Infratentorial neoplasms; Ganglioglioma; Posterior cranial fossa

Introduction

The ganglioglioma is an infrequent primary neoplasm of the central nervous system (CNS) which typically affects young adults [1]. It most frequently affects the temporal lobe [2,3] and its occurrence in the posterior fossa represents a rare event [4].

We report on a case of a patient with a ganglioglioma in the posterior fossa who underwent surgical treatment at our facility and conducted a brief review of the literature on the subject.

Case Report

A 36-years-old female patient was admitted to the Neurosurgery Service due to an insidious picture of headache and dizziness that had started 18 months before, occasionally associated with nausea and vomiting. At physical examination, the patient was in good general status, conscious, oriented, with no neurological deficits or papilledema.

Upon further investigation, magnetic resonance imaging (MRI) of the brain showed a solid/cystic lesion hypointense on T1-Weighted Image, hyperintense on T2-Weighted Image, with homogeneous gadolinium enhancement (Figure 1).

The patient underwent suboccipital craniotomy and complete excision of the lesion. She was discharged after 5 days, with no neurologic deficits. Histological analysis showed proliferation of bipolar astrocytes amid multiple ganglion cells. The diagnosis of ganglioglioma was confirmed by immunohistochemical analysis (Figure 2).

Discussion

Gangliogliomas are mixed neoplasms of neuronal and glial

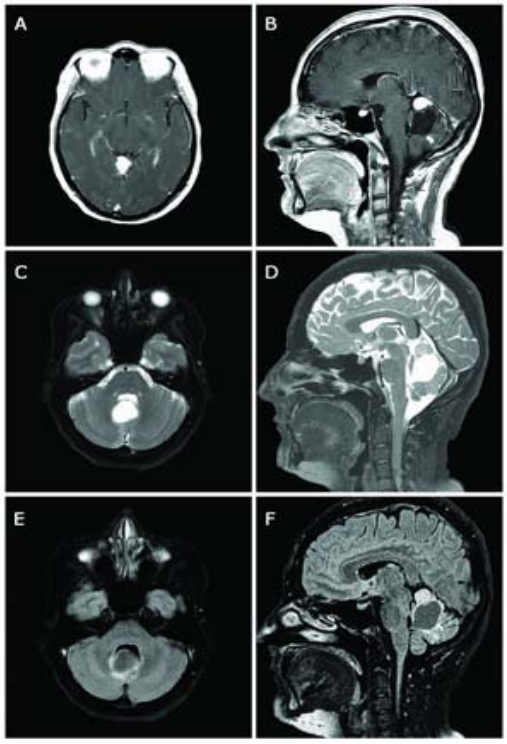

Figure 1: Magnetic resonance imaging of the brain showing solid/cystic cerebellar vermis lesion with gadolinium enhancement and compression of the fourth ventricle (A - Axial T1 with gadolinium enhancement, B - Sagittal T1 with gadolinium enhancement, C - axial in T2; D - sagittal in T2; E - axial FLAIR and F - sagittal FLAIR). The entire solid/cystic structure had approximate dimensions of 4.6 X 3.0 X 2.7 cm.

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cells, classified as grade I or II by the WHO (5), with rare reports of anaplastic gangliogliomas in the literature [2,4]. They represent approximately 0.4 to 9% of all CNS neoplasms [1,2,4] and only 15% of these involve the posterior fossa [4]. When in the supratentorial compartment, they are more commonly located in the temporal lobe [2,3] and manifest by generalized or focal seizures [2-4]. In the posterior fossa, they can manifest as hydrocephalus, cranial nerve palsies, gait and speech disorders and even myoclonus [4].

The MRI is the most sensitive study for the diagnosis of these lesions [2], which usually be present as isointense or hypointense lesions on T1-Weighted Image and hyperintense on T2-Weighted Image [2,4]. The contrast uptake may be heterogeneous to intense and homogeneous [4,5]. In the present case, the MRI showed an extensive solid/cystic formation in the cerebellar vermis, compressing the fourth ventricle and intense gadolinium enhancement, as can be expected for theses lesions.

The histological analysis shows neoplastic glia and neurons as intermixed components [3,4]. Immunohistochemistry analysis and the presence and expression of neural markers can be used to complement the histological analysis [3]. These histological features and synaptophysin labeling were present in the analyzed specimen.

Regarding prognosis, a 5-year survival is possible in up to 73% of cases and a 3-year disease-free follow-up in 53% of cases of posterior fossa gangliogliomas [4]. Factors related to poor prognosis are older age at diagnosis, male gender, histological features of malignancy and high Ki67 index [1]; good prognosis is related to its manifestation with seizures, its location in the cerebral hemisphere and complete resection of the lesion [1].

The best treatment for gangliogliomas is the complete excision of the lesion [1]. Unfortunately, this cannot be achieved in all cases. The adjuvant treatments are controversial, reserved for cases of extensive residual tumors [1,2,5]. Multiple cycles associated with chemotherapy have been described [4,5], which can lead to hematological aplasia and death [5]. There have been reports of lesion reduction with palliative radiation and localized radiotherapy [4].

Conclusion

The ganglioglioma represents an uncommon neoplasia. Its occurrence in the posterior fossa is even more striking. The best treatment consists of total excision of the lesion, with neurosurgical planning being an essential step for better patient outcome.

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