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Case Report

Extremely slow-growing cerebellar ganglioglioma in an elderly patient

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ABSTRACT

Background: Gangliogliomas account for 0.4% of primary brain tumors. They mainly occur in the supratentorial compartment and typically affect only children and young adults. We present an especially rare case of cerebellar ganglioglioma in an elderly patient.

Case Description: A 76-year-old Japanese woman presented with headache and nausea from 1 month previously. She had been diagnosed with a cerebellar tumor in her childhood, but the lesion was asymptomatic at that time, and there was no evidence of an increase in size, so it had been monitored without surgery. At the time of presentation, she had not been examined for approximately ten years. On admission, magnetic resonance imaging indicated a T2 hypertense cyst in the cerebellar vermis. Post-contrast T1 imaging showed an enhanced mural nodule in the cyst. Cerebral angiography showed that none of the vertebral arteries were significant feeders. The tumor was removed through posterior fossa craniotomy. The histopathological diagnosis was ganglioglioma. The patient's headache and nausea improved after surgery.

Conclusion: Our patient presented a very rare case of extremely slow-growing elderly ganglioglioma in the cerebellum. In patients with gangliogliomas, long-term follow-up is important because the disease may become symptomatic at an older age.

Keywords: Cerebellum, Cystic tumor, Elderly, Ganglioglioma, Slow-growing tumor

INTRODUCTION

Gangliogliomas are rare, slowly growing tumors composed of neoplastic mature ganglion cells in combination with glial cells. Gangliogliomas account for just 0.4% of primary brain tumors.[3,5] These tumors predominantly occur in children and young adults, with a median age of 12 years and with slight male predominance.[5] Gangliogliomas can arise anywhere in the central nervous system, but they are most commonly found in the temporal and frontal lobes, often leading to the manifestation of focal seizures.[3,5] Here, we present an especially rare case of ganglioglioma of the cerebellum in an elderly patient.

CASE DESCRIPTION

A 76-year-old Japanese woman presented with headache and nausea from 1 month previously. She had been diagnosed with a cerebellar tumor in her childhood, but the lesion was asymptomatic at

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that time, and there was no evidence of an increase in size, so it had been monitored without surgery. Her last magnetic resonance imaging (MRI) was performed approximately ten years before her presentation. On admission, her consciousness was alert, and physical examination revealed no ataxia or gait disturbance. Computed tomography showed a heterogeneous cystic mass in the left cerebellum without calcification [Figure 1a]. MRI indicated a T1 hypointense and T2 hypertense mass in the left cerebellar tonsil and a T2 hypertense cyst in the cerebellar vermis [Figures 1b-d]. Post-contrast T1 imaging showed an enhanced mural nodule in the cyst [Figures 1e and f]. Cerebral angiography revealed no mass stain from vertebral arteries [Figure 1g].

We performed tumor excision through posterior fossa craniotomy, incising the cyst and draining the xanthochromic fluid. The tumor was located within the cerebellar tonsil and was well-circumscribed from the surrounding parenchyma [Figure 2]. Hematoxylin and eosin staining of the surgical specimen showed binuclear atypical ganglion-cell-like cells and proliferation of atypical glial cells showing mild pleomorphism between hyalinized vessels, eosinophilic granular bodies and Rosenthal fibers [Figures 3a-c]. Immunohistochemically, the tumor cells were positive for GFAP and Olig2. Synaptophysin and neurofilament protein-positive ganglion cells were observed [Figures 3d and e]. V-Raf murine sarcoma viral oncogene homolog B (BRAF) V600E and CD34 were negative. The histopathological diagnosis was, therefore, ganglioglioma, the World Health Organization grade 1.

The patient's headache and nausea improved after surgery. She exhibited no cerebellar symptoms postoperatively and had a favorable postoperative course, so she was discharged to home two weeks after surgery. At the 1-year follow-up, postoperative MRI showed total tumor removal, and there has been no recurrence [Figures 4a and b].

DISCUSSION

Gangliogliomas are well-differentiated, slow-growing glioneuronal neoplasms composed of a combination of neoplastic ganglion and glial cells, molecularly characterized by mitogen-activated protein kinase pathway alteration.[5] Gangliogliomas usually occur in the temporal lobes and are commonly associated with chronic epilepsy.[6] The median age at diagnosis for patients with ganglioglioma is 12 years, with 80% of cases being in patients under 30 years.[5] Our case is unusual in that it was located in the cerebellar tonsil and because it first became symptomatic when the

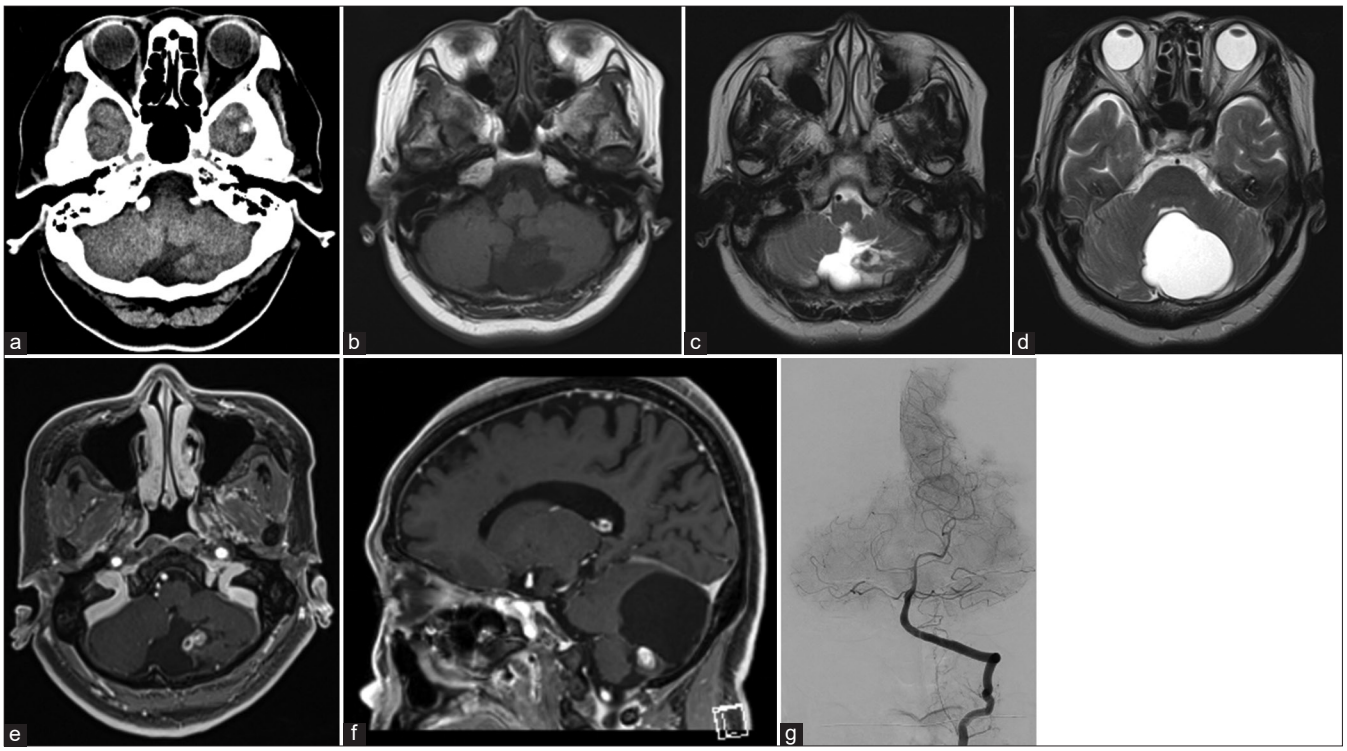

Figure 1: Preoperative images. Preoperative computed tomography (a) shows an isodense mass in the left cerebellar tonsil. In magnetic resonance imaging, the mass shows low intensity in (b) T1-weighted images and high intensity in (c) T2-weighted images. T2 hypertense cyst (d) is located in the cerebellar vermis. Post-contrast T1-imaging shows an enhanced mural nodule in the cyst (e, axial view, and f: sagittal view). The cerebral angiography (g) shows no tumor stains.

patient was elderly. In adult gangliogliomas, only 19% of gangliogliomas are located in the posterior fossa.[6] Komboz *et al*. reviewed the literature and reported 26 patients with a mean age of 20 who had gangliogliomas involving the cerebellum.[4] Jang *et al*. reported the case of cerebellar gangliocytoma in a 68-year-old woman in Korea.[3] To the best of our knowledge, our case patient is the oldest among the previously reported cases of symptomatic cerebellar gangliocytoma. Notably, our patient had been diagnosed with a cerebellar tumor in her childhood, and the lesion became symptomatic in old age. Clinical manifestation in cerebellar ganglioglioma includes cerebellar or brain stem symptoms, headache, and seizures.[4] We presume that, in this case, the cyst enlarged and compressed the cerebellum, causing headaches and nausea. A cerebellar ganglioglioma can be an incidental finding, but long-term follow-up is important because the disease may become symptomatic at an older age.

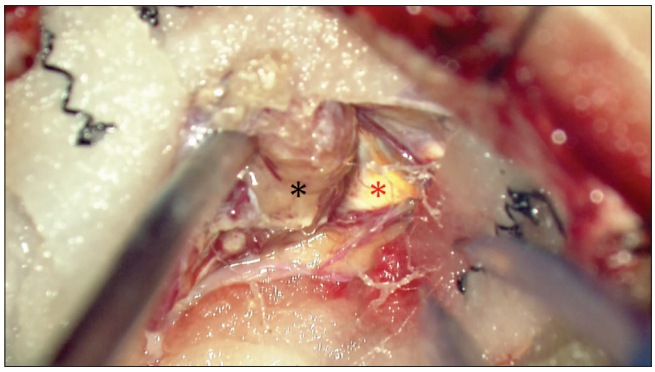

Figure 2: Surgical view. The surgical view shows a grayish tumor (black asterisk) located in the cerebellar tonsil (red asterisk).

This case involved a cystic tumor with nodules in the cerebellum of an elderly patient. The differential diagnoses include hemangioblastoma, pilocytic astrocytoma, and metastatic tumor.[2] Hemangioblastoma is often easy to diagnose preoperatively due to characteristic findings, including large feeding arteries, dilated draining veins, and a deep tumor blush on cerebral angiography. Pilocytic astrocytoma is the most common primary cerebellar tumor in children and young adults and cannot be distinguished from cerebellar ganglioglioma radiologically.[4] Metastatic lesions can be diagnosed by the patient's age and their association with systemic manifestation.[4] The gangliogliomas should also be considered in the differential diagnosis of cystic tumors with nodules in the cerebellum.

Gangliogliomas are well-differentiated and indolent tumors with excellent prognosis. Surgical resection is the standard of care for ganglioglioma and generally yields a good outcome.[6] The histological grade, age, and extent of resection are prognostic factors for progression-free survival.[6] BRAF V600E positivity in immunohistochemistry was around 40%, and BRAF V600E was associated with poor outcomes in pediatric ganglioglioma.[1] In this case, gross total resection was achieved, and BRAF V600E immunostaining was negative, suggesting a good prognosis.

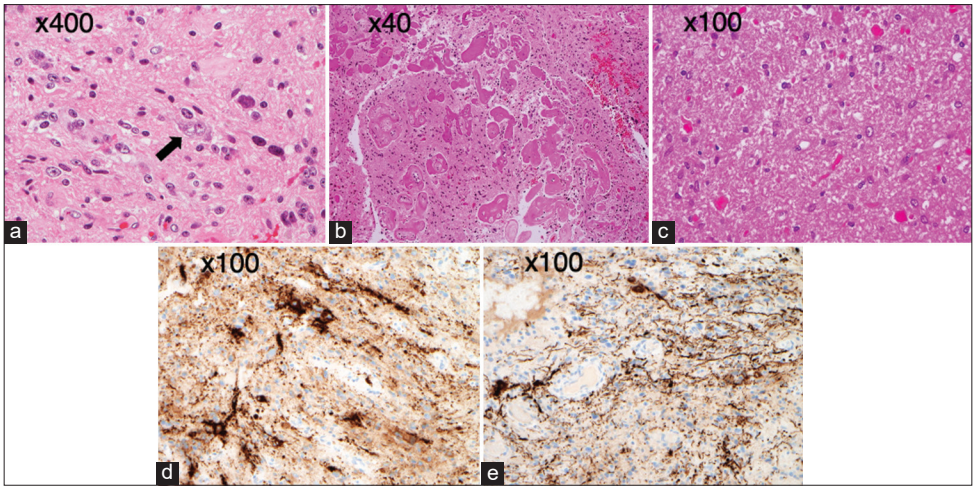

Figure 3: Surgical specimens. Photomicrographs of the surgical specimen stained with hematoxylin and eosin show binuclear atypical ganglion-cell-like cells (a black arrow) and (a) proliferation of atypical glial cells showing mild pleomorphism between (b) hyalinized vessels, (c) eosinophilic granular bodies, and Rosenthal fibers. Immunohistochemical stains show that tumor cells are immunoreactive for (d) synaptophysin and (e) neurofilament protein.

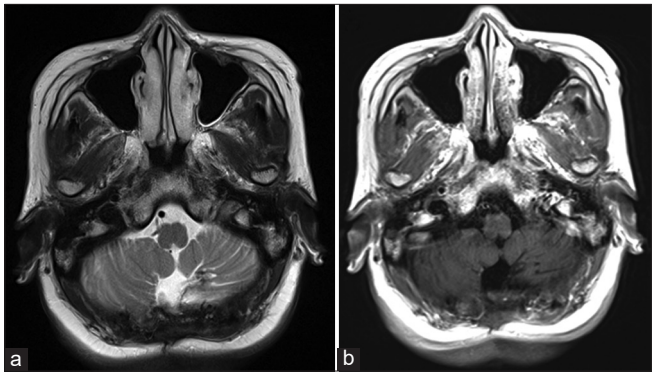

Figure 4: Postoperative images. (a) Postoperative T2-weighted images and (b) contrast-enhanced T1-weighted images show that the mass is totally excised.

CONCLUSION

We presented a rare case of extremely slow-growing ganglioglioma in the cerebellum in an elderly patient. Long-term follow-up of ganglioglioma is important because it may become symptomatic at an older age.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript, and no images were manipulated using AI.

REFERENCES

1. Dahiya S, Haydon DH, Alvarado D, Gurnett CA, Gutmann DH, Leonard JR. BRAFV600E mutation is a negative prognosticator in pediatric ganglioglioma. Acta Neuropathol 2013;125:901-10.
2. El Hamrouni I, Raftopoulos C, Duprez T, Wilms G. Cerebellar ganglioglioma in a young woman. Neurol Neurol Sci Open Access 2020;3:1014.
3. Jang EW, Cho JH, Chang JH, Ahn JY. Cerebellar ganglioglioma in an old patient. J Korean Neurosurg Soc 2007;42:53-5.
4. Komboz F, Zechel S, Malinova V, Mielke D, Rohde V, Abboud T. Infratentorial ganglioglioma mimicking a cerebellar metastasis. Int J Neurosci 2022;2:1-5.
5. WHO Classification of Tumours Editorial Board. WHO classification of tumors series: Central nervous system tumours. 5th ed., Vol. 6. Lyon, France: International Agency for Research on Cancer; 2021. Available from: https://publications.iarc.fr/601 [Last accessed on 2023 Sep 19].
6. Yust-Katz S, Anderson MD, Liu D, Wu J, Yuan Y, Olar A, *et al*. Clinical and prognostic features of adult patients with gangliogliomas. Neuro Oncol 2014;16:409-13.

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