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A Rare Case of Adult-onset Gangliocytoma in the Parietal Lobe: Case Report and Surveillance, Epidemiology, and End Results Registry Data Analysis

Ryosuke IKEMACHI,[1] Yusuke TOMITA,[2] Yoshihiro OTANI,[1]
Joji ISHIDA,[1] Kentaro FUJII,[1] Fumiyo HIGAKI,[3]
Sawako ONO,[4] Yonehiro KANEMURA,[5] and Shota TANAKA[1]

[1]Department of Neurological Surgery, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Okayama, Japan
[2]Department of Neurosurgery and Neuroendovascular Surgery, Hiroshima City Hiroshima Citizens Hospital, Hiroshima, Hiroshima, Japan
[3]Department of Radiology, Okayama University Hospital, Okayama, Okayama, Japan
[4]Department of Pathology and Oncology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Okayama, Japan
[5]Department of Biomedical Research and Innovation, Institute for Clinical Research, NHO Osaka National Hospital, Osaka, Osaka, Japan

Abstract

We report a case of adult-onset gangliocytoma in the parietal lobe. A 54-year-old woman presented with sensory disturbance in her right upper limb. A computed tomography scan revealed a cystic and calcified lesion in the left parietal lobe. She underwent a left parietal craniotomy with gross total resection, and the pathological diagnosis was confirmed as gangliocytoma. Gangliocytomas are very rare tumors of the central nervous system, predominantly affecting children and young adults, and are often located in the temporal lobe. Reports of gangliocytomas developing after middle age are uncommon. To assess the epidemiology of gangliocytoma, we utilized data from the Surveillance, Epidemiology, and End Results database. From January 1, 2000, to December 31, 2021, a total of 74 cases were identified, 18 of which were in patients over 50 years of age. While 27 patients had tumors in the temporal lobe, the most frequent site, others had tumors in different locations. Notably, there were no patients over 50 with gangliocytoma in the parietal lobe in the Surveillance, Epidemiology, and End Results registry. These findings suggest that in older patients, although gangliocytomas located outside the temporal lobe are rare, they are kept in mind as one of the differential diagnoses.

Keywords: gangliocytoma, adult-onset, SEER

Introduction

Gangliocytomas are classified as glioneuronal and neuronal tumors in the 5th edition of the World Health Organization (WHO) classification of the central nervous system (CNS) tumors and are histologically benign, categorized as CNS WHO grade 1.[1)] They are considered distinct from tumors arising in the pituitary gland and from dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease). Gangliocytomas are very rare tumors of the CNS, accounting for less than 1% of primary brain tumors. Most cases are reported to occur in children and young adults, with a predilection for the temporal lobe.[2,3)] The main clinical presentation is epilepsy. Previous reports in epilepsy surgery series indicate that the relative incidence of gangliocytomas ranges from 0.4 to 3.2%.[2)] Due to its rarity, the

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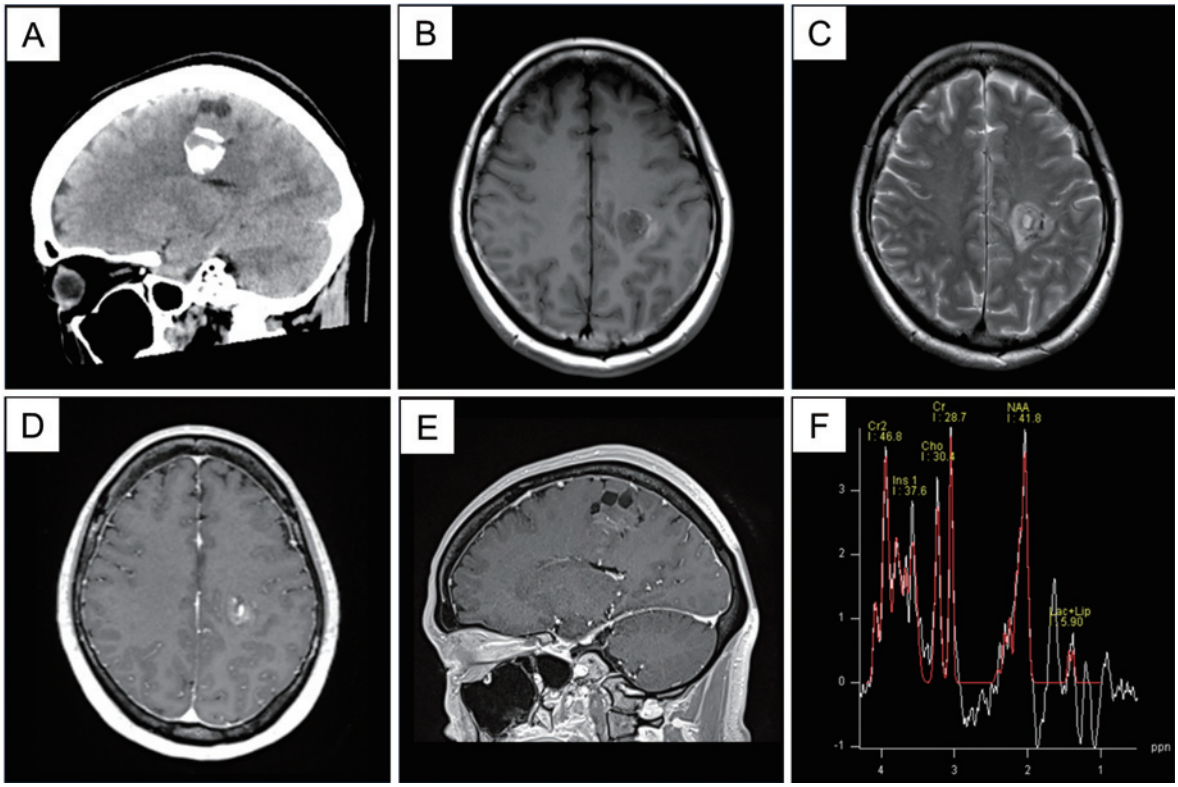

Fig. 1 Initial preoperative images.
A: Plain CT image, sagittal view. CT scan showed a mass with mixed calcified and cystic components in the left parietal lobe. B: T1-weighted MR image, axial view. C: T2-weighted MR image, axial view. D: Gadolinium-enhanced T1-weighted MR image, axial view. E: Gadolinium-enhanced T1-weighted MR image, sagittal view. MRI revealed a tumor with a maximum diameter of 45 mm, containing a mixture of solid and cystic components with gadolinium enhancement. F: MRS showed a relatively preserved N-acetyl aspartate peak and a mildly elevated choline peak, along with a mildly elevated lactate and lipid peak.
CT: computed tomography; MRI: magnetic resonance imaging; MRS: magnetic resonance spectroscopy

epidemiology of gangliocytomas is poorly understood.[4] The clinical and genetic landscape of gangliocytomas has not been extensively reported.

In this study, we reported an adult-onset gangliocytoma in the left parietal lobe in a 54-year-old female presenting with sensory disturbances. Reports of adult-onset gangliocytomas are limited, with only five cases documented in patients over 50 years old. To determine the clinical features of adult-onset gangliocytomas, we utilized the Surveillance, Epidemiology, and End Results (SEER) database, which represents approximately 47.9% of the United States (US) population.

Case Report

A 54-year-old female presented with sensory abnormalities in her right upper limb, and had a medical history of hypertension. A computed tomography (CT) scan of the head revealed a mass in the left parietal lobe with mixed calcified and cystic components (Fig. 1A). Magnetic resonance imaging (MRI) showed a relatively circumscribed lesion of 35 mm × 25 mm × 45 mm in the left parietal lobe, containing both solid and cystic components. The lesion appeared hypointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 1B and C). Gadolinium-enhanced T1-weighted images demonstrated heterogeneous enhancement of the solid component, while the cyst wall showed no enhancement (Fig. 1D and E). Magnetic resonance spectroscopy showed a relatively preserved N-acetyl aspartate peak and a mildly elevated choline peak, along with a mildly elevated lactate and lipid peak. (Fig. 1F). Diffusion tensor imaging indicated that the pyramidal tract was located just anterior to the tumor.

The preoperative diagnosis was supratentorial ependymoma or glioneuronal tumor. A left parietal craniotomy was performed, and after corticotomy of the superior parietal lobule, a grayish, well-demarcated tumor was exposed (Fig. 2A). The patient received 5-aminolevulinic acid preoperatively; however, the tumor did not exhibit red fluorescence under a 410 nm ultraviolet light source. The deep part of the lesion, appearing as a calcified component on CT, consisted of psammomatous calcifications that were easily aspirated (Fig. 2B). Intraoperative MRI confirmed that gross total resection was achieved (Fig. 2C).

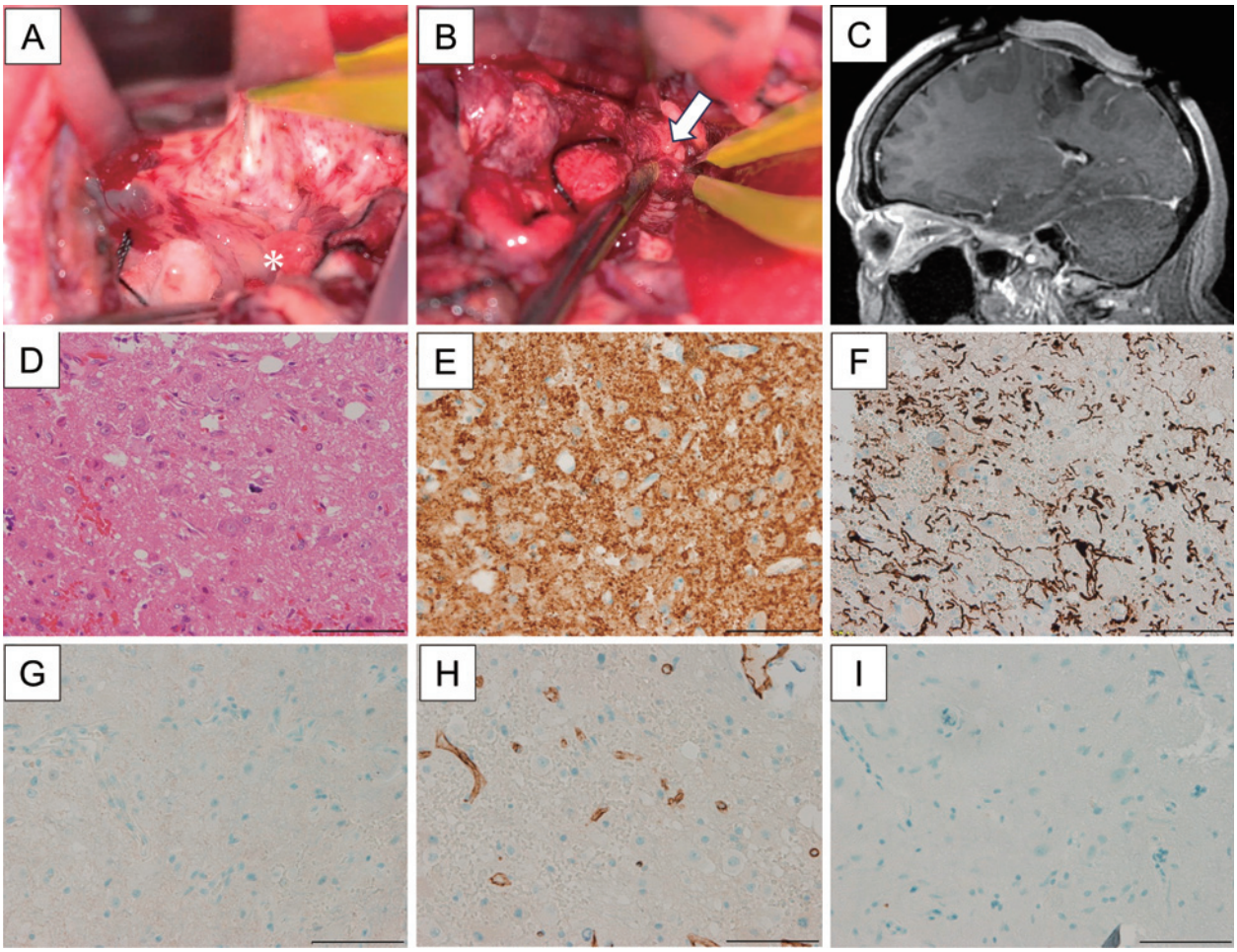

Fig. 2 Intraoperative findings and histopathological findings from surgical specimens.
The tumor (asterisk) was grayish, soft, and well-demarcated (A). The deep calcified areas consisted of psammomatous calcification (arrow), which was easily aspirated (B). Intraoperative MRI confirmed that gross total resection was achieved (C). Hematoxylin and eosin staining revealed clusters of mature and neoplastic neuronal cells, with glial elements present only sparsely and no atypia observed (D). Immunohistochemical staining demonstrated positive staining for synaptophysin (E) and negative results for GFAP (F), NeuN (G), CD34 (H), and BRAF V600E (I). Scale bar of A, B, C, D, E, and F = 100 μm.
GFAP: glial fibrillary acid protein; MRI: magnetic resonance imaging

Histological analysis revealed clusters of mature and neoplastic neuronal cells, with cytoplasmic ballooning and microcalcifications (Fig. 2D). Glial elements were sparsely present and showed no atypia. Immunohistochemical staining indicated that the large ganglion-like cells were positive for synaptophysin (Fig. 2E), while tumor cells were negative for glial fibrillary acid protein, NeuN, CD34, and BRAF V600E (Fig. 2F-I). Ki-67 positive cells were estimated to be less than 1%. Genetic analysis revealed no hotspot mutations for *IDH1*, *IDH2*, *H3F3A*, *HIST1H3B*, *TERT* promoter, or *BRAF*. Based on these findings, the diagnosis was gangliocytoma, CNS WHO grade 1. Postoperatively, the patient developed right-sided hemiparesis, which improved quickly. At 10 months postoperatively, MRI showed no tumor recurrence.

Analysis of the SEER registry data

Since gangliocytomas in elderly patients are rare, we utilized the SEER database to comprehensively analyze the location and age distribution of these tumors. The SEER database is a publicly available cancer registry maintained by the National Cancer Institute, covering approximately 47.9% of the US population and serving as a valuable resource for CNS neoplasm epidemiology studies.[5,6] We queried cancer statistics using SEER*Stat software version 8.4.2 (Information Management Service, Inc., Calverton, MD, USA) for data between January 1, 2000, and December 31, 2021. During this period, a total of 336,718 primary brain tumors were recorded, with 95 cases of gangliocytomas. The age-adjusted incidence rate of all primary brain tumors is 18 per 100,000 people, while the incidence rate for gangliocytomas is 0.005 per 100,000 people, ac-

Table 1 Results of SEER Database Search for the Number of Cases by Age

Variables	50 > age (n = 56), n (%)	50 ≤ age (n = 18), n (%)	p-Value
Sex			0.79
Male	31 (55.4)	11 (61.1)	
Female	25 (44.6)	7 (38.9)	
Race			0.47
White	36 (64.3)	15 (83.3)	
Black	5 (8.9)	0	
Asian and Pacific Islander	4 (7.1)	2 (11.1)	
Hispanic	9 (16.0)	1 (5.6)	
Other	2 (3.6)	0	
Tumor location			0.95
Cerebrum	2 (3.6)	2 (11.1)	
Frontal lobe	8 (14.3)	2 (11.1)	
Temporal lobe	19 (33.9)	8 (44.4)	
Parietal lobe	5 (8.9)	0	
Occipital lobe	4 (7.1)	1 (5.6)	
Ventricle, NOS	2 (3.6)	1 (5.6)	
Cerebellum, NOS	6 (10.7)	2 (11.1)	
Brainstem	2 (3.6)	0	
Overlapping lesion	4 (7.1)	1 (5.6)	
Spinal cord	3 (5.4)	1 (5.6)	
Optic nerve	1 (1.8)	0	
Extent of resection			0.58
GTR	31 (55.4)	8 (44.5)	
STR	15 (26.8)	7 (38.9)	
No surgery	10 (17.9)	3 (16.7)	
Vital status			0.06
Alive	50 (89.3)	12 (66.7)	
Dead	6 (10.7)	6 (33.3)	

GTR: gross total resection; NOS: not otherwise specified; SEER: Surveillance, Epidemiology, and End Results; STR: subtotal resection

counting for approximately 0.03% of all primary brain tumors. Of the 95 cases of gangliocytomas, 86 were histologically diagnosed. Twelve cases were excluded because they were located in the pituitary gland, which is classified as a different entity in the WHO classification. Thus, 74 cases were further examined. Among these, tumor locations included the temporal lobe (n = 27, 36.5%), frontal lobe (n = 10, 13.5%), cerebellum (n = 8, 10.8%), parietal lobe (n = 5, 6.8%), occipital lobe (n = 5, 6.8%), spinal cord (n = 4, 5.4%), ventricle (n = 3, 4.1%), brainstem (n = 2, 2.7%), and optic nerve (n = 1, 1.4%). Eighteen of the 74 cases (24.3%) occurred in patients aged 50 and older, with none located in the parietal lobe. In addition, clinical characteristics were compared between the patient groups aged 50 years and older and those younger than 50 years (Table 1). Fisher's exact test was used to compare categorical variables, and two-sided p-values less than 0.05 were considered statistically significant. There are no significant differences observed. Although the mortality rates appeared relatively high, 10.7% for individuals under 50 years old and 33.3% for those over 50 years, the majority of these deaths were unrelated to gangliocytoma.

Discussion

Gangliocytomas are rare tumors and are classified in the same group as gangliogliomas in the WHO classification. Radiologically and histopathologically, gangliocytomas share similar features to gangliogliomas and are often discussed together.[7] Radiological imaging features are mostly

Table 2 Review of Reported Cases of Gangliocytomas over 50 Years of Age

Author, year	Age, sex	Location	Symptoms	Radiological findings	Treatment	Outcome
Beal MF, 1981	55, M	Third ventricle	Cognitive decline	Nonenhancing mass with hydrocephalus	VP shunt and radiotherapy	Died 2 years after radiographic diagnosis[#]
Kim HS, 2001	59, F	Parietal lobe	Headache and dizziness	Homogeneous enhancing dural-based mass with calcification, mimicking a meningioma	Resection	Not described
Mirzai H, 2004	52, M	Frontal lobe	Nothing	Cystic and calcified mass	Resection	Not described
Ratilal B, 2007	60, F	Temporal lobe	Seizure	FLAIR high lesion in the white matter	Resection	Not described
Alarifi N, 2022	66, F	Lateral ventricle	Fatigability, headaches, and confusion	Diffuse enhancing mass with hydrocephalus	Resection	No recurrence in 12 months

F: female; FLAIR: fluid-attenuated inversion recovery; M: male; VP shunt: ventriculoperitoneal shunt
[#]Histological diagnosis of this case was performed by necropsy.

similar, typically presenting a mixture of cystic and solid components, with calcification observed in approximately 30% of cases. The solid components exhibit various enhancements with gadolinium.[7] Histopathologically, gangliocytomas consist of clusters of large neuronal cells with atypia, with binucleation and cytoplasmic ballooning being common. Glial elements are present only sparsely, and no atypia is observed. The key difference from gangliogliomas lies in the presence of neoplastic glial elements in the latter. Recent molecular diagnostic studies have shown that approximately 50% of gangliogliomas harbor *BRAF V600E* mutations;[8] however, specific genetic alterations in gangliocytomas have not been identified. Due to their radiological and histopathological similarities, distinguishing between the two can sometimes be challenging.

These ganglion cell tumors have a predilection for children and young adults, often occurring in the temporal lobe.[2,3] They have also been reported in various locations within the CNS, including the spinal cord, brainstem, basal ganglia, thalamus, and ventricles.[9-12] Hirota et al.[13] noted that 60% (8 of 12 cases) of gangliocytomas occurring in the cerebral hemispheres were found in individuals aged 30 or younger. According to the Japanese Brain Tumor Registry (2005-2008), there were 12 cases of gangliocytoma (0.1% of all primary brain tumors), with only one patient being over 50 years old.[14] Adult-onset gangliocytomas have been reported in limited numbers, with only five cases documented in individuals over 50 years of age (Table 2).[15-19] One case involved a 59-year-old woman with a tumor in the parietal cortex that exhibited exophytic extension and was attached to the dura mater.[15] Other reports included temporal lobe lesions in a 60-year-old woman,[16] third ventricle lesions in a 55-year-old man,[17] frontal lobe lesions in a 52-year-old man,[18] and lateral ventricle lesions in a 66-year-old woman.[19] Among these five cases, genetic analysis was conducted for only one, which showed negative results for IDH1R132H and BRAF V600E through immunohistochemistry. No remarkable clinical similarities were identified among these six cases, including our own.

To clarify the detailed incidence of gangliocytomas in elderly patients in the real world, we utilized the SEER database. This study represents the largest comprehensive analysis of the epidemiology of gangliocytoma. While previous reports estimated that nearly 80% of cases occurred in the temporal lobe,[2,3] our results indicate that only approximately 40% occur in this region, with the remaining cases distributed across various other areas of the CNS. Previous reports, which analyzed cohorts of patients undergoing epilepsy surgery, may have skewed the incidence in the temporal lobe. Similar to our findings, a previous analysis of pediatric ganglioglioma and gangliocytoma using the SEER database showed an incidence of 47.41% for ganglioglioma and gangliocytoma in the temporal lobe.[20] When comparing patients over 50 years of age to those under 50 years, no significant epidemiological differences in tumor locations were observed. However, there were no recorded cases of parietal lobe occurrence in patients over 50 years, suggesting a potential tendency for fewer parietal lobe cases in older patients compared to younger patients. In addition, a quarter of the cases in our study involved patients older than 50 years of age. Generally, gangliocytomas are known to be more prevalent in younger people, but no study has specified the exact frequency. The strengths of this SEER database study include its representation of the US population, making the findings potentially generalizable, as well as its long data collection period and large number of cases. Compared to previous notable observational studies, this population-based observational study has the potential to more accurately reflect the actual epidemiology of gangliocytomas.

Conclusions

In this report, we presented a case of adult-onset gangliocytoma arising in the parietal lobe. In addition, we

analyzed the epidemiology of gangliocytomas using the SEER database, which encompasses the largest number of gangliocytoma cases to date. Our analysis revealed that adult-onset gangliocytoma in the parietal lobe is indeed very rare. Although the occurrence of gangliocytomas in older age groups is uncommon, it is kept in mind as one of the differential diagnoses, as these tumors can develop at any age and in any part of the CNS.

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Ethics Approval

All procedures performed in studies involving human participants were conducted in accordance with the ethical standards of the Okayama University Graduate School of Medicine (No. 1608-026) and the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Informed Consent

Informed consent for publication was obtained from the patients.

Conflicts of Interest Disclosure

There are no conflicts of interest.

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Corresponding author: Yoshihiro Otani, MD, PhD
Department of Neurological Surgery, Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences, 2-5-1 Shikata-cho, Kitaku, Okayama 700-8558, Japan.
e-mail: yotani@okayama-u.ac.jp