CLINICAL CASE REPORT

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Gliomatosis cerebri

Rymantė Gleiznienė[1], Ugnius Bučinskas[1], Saulius Lukoševičius[1], Antanas Vaitkus[2], Simona Letautienė[3], Daiva Apanavičiūtė[1], Milda Galvonaitė[1]

[1]Department of Radiology, Kaunas University of Medicine, [2]Department of Neurology, Kaunas University of Medicine, [3]Vilnius Oncology Hospital, Lithuania

***Key words**: gliomatosis cerebri; magnetic resonance tomography; stereotaxic biopsy.*

***Summary.** Gliomatosis cerebri is a rare diffusely infiltrating glial tumor involving two or more lobes and is frequently is bilateral. Infiltrative extent of tumor is out of proportion to histological and clinical features. We present a case in which finally the diagnosis of gliomatosis cerebri was made. In this case, computed tomography showed that midline structures were insignificantly shifted to the left, there was a mild dilatation of lateral ventricles more expressed on the right, and no pathologic changes of brain tissue density were found. On magnetic resonance tomography, T2W/SE and T2W/FLAIR images revealed zones of hyperintense signal, spreading with time, through several lobes of the brain with no enhancement on T1W images. Diagnosis of gliomatosis cerebri was suspected, stereotaxic biopsy was performed, and pathological examination revealed changes typical of diffuse glial tumor. In this article, changes typical of gliomatosis cerebri seen in other radiological methods such as computed tomography, magnetic resonance spectroscopy, dynamic contrast-enhanced T2*-weighted magnetic resonance, and positron emission tomography also are discussed.*

Introduction

Gliomatosis cerebri (GC) is a diffusely infiltrating glial tumor involving two or more lobes and is frequently bilateral. Infiltrative extent of tumor is out of proportion to histological and clinical features (1).

Case report

A 43-year-old man complained of pain in the legs with shifting localization for 15 years and paroxysmal headache of various characters for several months, which subsided by itself or by changing position of the head. Clinical examination was unremarkable. The findings of laboratory tests were normal.

The patient was referred to the Department of Radiology for brain computed tomography. The midline structures were insignificantly shifted to the left. There was a mild dilatation of the lateral ventricles more expressed on the right. No pathologic changes of brain tissue density were found (Fig. 1). Then magnetic resonance imaging (MRI) of the brain was performed. At first, on T2W/SE and T2W/FLAIR images, zones of hyperintense signal and with no enhancement temporally in the right hemisphere including insular cortex were seen (Figs. 2 and 3). After two months on T2W/FLAIR images, these zones also appeared in the left hemisphere temporoanterobasally, frontobasally in medial structures, parietooccipitally and in the dorsal part of *corpus callosum* (Fig. 4 A–C). There were no changes in signal intensity on T1W/SE images without contrast media and no contrast enhancement. There was no change in signal intensity subtentorially. Basal cisterns and subarachnoidal convexital spaces were narrow; the course and signal intensity of the main blood vessels were unchanged. According to the MRI changes, cerebral gliomatosis was suspected. The stereotaxic biopsy was done, and pathohistological investigation revealed that changes are typical for diffuse glial WHO grade II tumor. So the diagnosis of gliomatosis was confirmed. The treatment with corticosteroids and radiotherapy was administered.

Discussion

It is a rare, highly aggressive primary diffusely infiltrating malignant glial tumor characterized by diffuse infiltration of the brain with neoplastic glial cells that typically involve multiple brain areas (2). Sometimes there is formation of a distinct tumor mass (3). Etiology of GC is controversial. It is classified as neoplasm of unknown histogenesis (1).

The morphology of tumor cells is diverse, taking on the appearance of astrocytes, oligodendrocytes, or Schwann cells with variable mitotic activity. GC represents an extreme form of diffusely infiltrating glioma (2). Both white and gray matters are infil-

Correspondence to U. Bučinskas, Department of Radiology, Kaunas University of Medicine, Eivenių 2, 50009 Kaunas, Lithuania. E-mail: ugnius77@yahoo.com

Adresas susirašinėti: U. Bučinskas, KMU Radiologijos klinika, Eivenių 2, 50009 Kaunas
El. paštas: ugnius77@yahoo.com

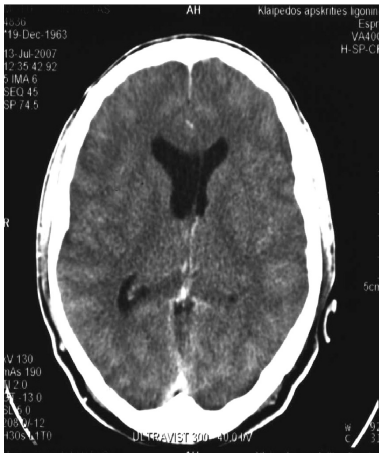


Fig. 1. Contrast-enhanced computed tomography of the brain

The midline structures are insignificantly shifted to the left, mild dilatation of lateral ventricles is observed, no pathologic changes of brain tissue density are seen.

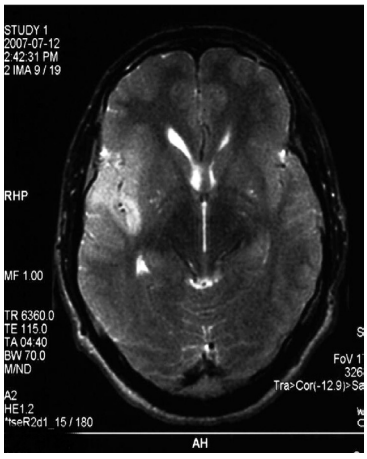


Fig. 2. Magnetic resonance tomography of the brain, T2W axial image

Hyperintense signal temporally in the right hemisphere including insular cortex.

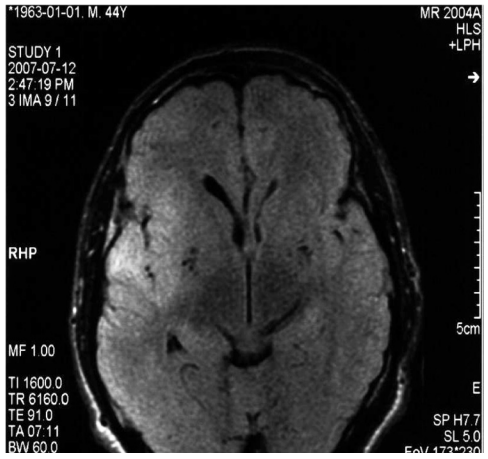
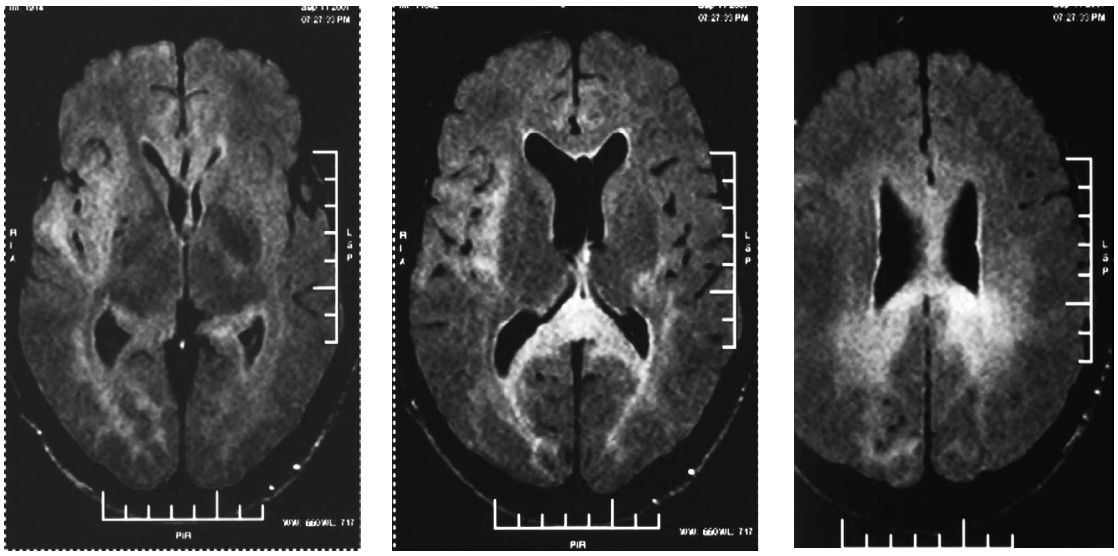


Fig. 3. Magnetic resonance tomography of the brain, T2W/FLAIR axial image

Hyperintense signal temporally in the right hemisphere including insular cortex.



A B C

Fig.4. Magnetic resonance tomography of the brain, T2W/FLAIR axial images

Hyperintense signal remains temporally in the right hemisphere including insular cortex (A) and new hyperintense zones appeared in the left hemisphere temporoanterobasally, frontobasally in medial structures (A), occipitally, in dorsal part of *corpus callosum* (B) and parietally (C).

trated. Macroscopically the tumor appears as an increased volume of the affected hemisphere, particularly white matter (3).

One of the accepted hallmarks is increased cellularity without destruction of the infiltrated parenchyma (4). There is no necrosis or neovascularity. There are two gross pathologic types: type I, neoplastic overgrowth, expansion of existing structures without circumscribed tumor mass; type II, a diffuse lesion with a focal neoplastic mass with malignant features (1). GC usually is WHO grade III, but also may be WHO grade II.

Although GC can occur at any age, it generally affects individuals in their third and fourth decades of life with no gender predominance (2).

GC may affect any part of the brain or even the spinal cord, optic nerve and compact white matter. Clinical manifestations are nonspecific and include headache, seizures, visual disturbances, corticospinal tract deficits, lethargy, and dementia (2, 5, 6). These symptoms are as a result of increased pressure within the head (headache and vomiting), as well as more localizing symptoms as a function of specific tumor location, rate of growth and associated in-

flammation (weakness and other motor dysfunction, neuroendocrine abnormalities, changes in behavior or thought processes) (6). Rarely GC is complicated by hydrocephalus or herniation and extremely rarely by hemorrhage (1). Very rarely GC may present as a parkinsonian syndrome (7). Symptoms appear late since the tumor is slow-growing and does not disrupt the brain tissue until late. Personality changes are more common than focal signs (3).

On MRI, it typically appears as a diffuse, poorly circumscribed, infiltrating nonenhancing lesion that is hyperintense on T2-weighted images and expands the cerebral white matter (2). GC infiltrates, enlarges yet preserves underlying brain architecture. Typically, it involves hemispheric white matter, basal ganglia and thalami (75%), *corpus callosum* (50%), brainstem and spinal cord (10–15%), cerebellum (10%). MRI shows expanded hemisphere, with sulcal effacement and ventricular compression as well. Usually there is no enhancement on MRI until late in the course of the disease (1, 3).

MRI spectroscopy might be used to classify GC as a stable or progressive disease indicating its potential therapeutic relevance (8). GC shows a marked elevation of myo-inositol, normal or mildly increased choline, decreased N-acetyl-aspartate concentration. Diffusion tensor imaging shows the preservation of nerve fibers in GC compared with other tumors (1). Dynamic contrast-enhanced T2* weighted MRI shows a low relative cerebral blood volume, which correlates with lack of vascular hyperplasia. PET with FDG shows marked hypometabolism (1).

The diagnosis of GC should be confirmed by biopsy. The pathological grade of GC is not always established, because only a fraction of these tumors is biopsied (6).

GC can be mistaken for nonneoplastic white matter disease. It is difficult to distinguish GC from highly infiltrate anaplastic astrocytoma or glioblastoma multiforme (2).

In addition, it must be differentiated from vasculitis, anaplastic astrocytoma, viral encephalitis, demyelinization, progressive multifocal leukoencephalopathy, lymphoma, and inherited or acquired metabolic disorder (1).

Usually there is a relentless progression, and prognosis for GC is generally poor with a median survival time of 38 months. Surgery is not practical considering the extent of the disease, standard chemotherapy (nitrosourea) is shown to be unsuccessful, and although brain irradiation can stabilize or improve neurological function in some patients, its impact on survival has yet to be proven (1, 2, 5).

Galvos smegenų gliomatozė

Rymantė Gleiznienė[1], Ugnius Bučinskas[1], Saulius Lukoševičius[1], Antanas Vaitkus[2], Simona Letautienė[3], Daiva Apanavičiūtė[1], Milda Galvonaitė[1]

[1]Kauno medicinos universiteto Radiologijos klinika, [2]Kauno medicinos universiteto Neurologijos klinika, [3]Vilniaus onkologijos ligoninė

Raktažodžiai: galvos smegenų gliomatozė, magnetinio rezonanso tomografija, stereotaksinė biopsija.

Santrauka. Galvos smegenų gliomatozė yra retas infiltratyviai augantis navikas, kuris pažeidžia dvi ar daugiau galvos smegenų skilčių, dažnai yra abipusis. Naviko infiltracinis išplitimas nėra proporcingas histologiniams ir klinikiniams požymiams. Šiame straipsnyje pateikiamas klinikinis atvejis, kai buvo diagnozuota galvos smegenų gliomatozė. Kompiuterinės tomografijos metu pastebėta nežymi galvos smegenų vidurinės linijos struktūrų dislokacija į kairę, švelnus šoninių skilvelių išsiplėtimas, ryškesnis dešinėje, tačiau patologinių židininių smegenų tankio pokyčių neaptikta. Magnetinio rezonanso tomografijos metu T2W ir T2W/FLAIR vaizduose nustatytos keliose galvos smegenų skiltyse plintančios hiperintensinio signalo zonos be kontrastinės medžiagos kaupimo požymių T1W vaizduose. Įtarta galvos smegenų gliomatozė. Atlikta stereotaksinė biopsija bei patologinis tyrimas ir nustatyta difuziniam glialiniam navikui būdingų pokyčių. Šiame straipsnyje yra aptariami ir šiai patologijai būdingi pokyčiai, rasti kitų radiologinių tyrimų metu, tokių kaip kompiuterinė tomografija, magnetinio rezonanso spektroskopija, dinaminis kontrastinis T2*W magnetinio rezonanso tyrimas ir pozitronų emisijos tomografija.

References

1. Osborn A, Blaser S, Salzman K. Diagnostic imaging: brain. 1st ed. NY: Amirsys Inc; 2004. I:6-26-29.
2. University of Rochester Medical Center [homepage on the Internet]. New York: University of Rochester Medical Center. ©Copyright University of Rochester Medical Center, 1999-2006. Matthew Cham, Manoj Ketkar, P-L Westesson. Neuroradiology Case of the Week Case 1. [Last updated: 03/26/2007] Available from: URL: http://www.urmc.rochester.edu/smd/Rad/neurocases/Neurocase01.htm
3. Medcyclopaedia [data base on the Internet]. Copyright General Electric Company 1997-2007. Available from: URL: http://www.medcyclopaedia.com/library/topics/vol-

ume_vi_1/g/gliomatosis_cerebri.aspx
4. MedPix™–2. Medical Image Database, Atlas, and Teaching File. Topic 923. Copyright © 1999–2007, Smirniotopoulos JG, Irvine H. Gliomatosis cerebri. Modified: 2003-12-22. Available from: URL: http://rad.usuhs.edu/medpix/radpix.html?mode=single&comebackto=mode%3Dgeo_browse&recnum=923
5. Wikipedia free encyclopedia [data base on the Internet]. Wikimedia Foundation, Inc. Nervous tissue tumors, ICD-O 9381/3. Gliomatosis cerebri. Available from: URL: http://en.wikipedia.org/wiki/Gliomatosis_cerebri
6. Children's Hospital Boston [homepage on the Internet]. Boston: Children's Hospital Boston. ©Copyright Children's Hospital Boston, 2005-2006. Gliomatosis cerebri. Available from: URL: http://www.childrenshospital.org/az/Site963/mainpageS963P0.html
7. Asada T, Takayama Y, Tokuriki Y, Fukuyama H. Gliomatosis cerebri presenting as a parkinsonian syndrome. J Neuroimaging 2007;17:269-71.
8. Bendszus M, Warmuth-Metz M, Klein R, Burger R, Schichor C, Tonn JC, et al. MR spectroscopy in gliomatosis cerebri. AJNR Am J Neuroradiol 2000;21(2):375-80.

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