Content available at: iponlinejournal.com

Indian Journal of Pathology and Oncology

Journal homepage: www.innovativepublication.com

Case Report

Small cell glioblastoma of basal ganglia: Case report

Barani Karikalan[1],*, Thanikachalam Pasupati[2]

[1]*Dept. of Pathology, Perdana University, Selangor, Malaysia*
[2]*Clinipath Pathology Laboratory, Klang, Malaysia*

ARTICLE INFO

Article history:
Received 12-04-2019
Accepted 18-11-2019
Available online 25-05-2020

Keywords:
Small cell glioblastoma
Basal ganglia

ABSTRACT

Small cell glioblastoma (SCGBM) is a rare variant with monomorphous cells and deceptively bland nuclei and are often misdiagnosed. Here, we present a case of small cell GBM in a 39-year-old male involving bilateral basal ganglia diagnosed as primary central nervous system lymphoma on MRI. After excision, the tumor showed features of small cell glioblastoma. Clinical features, morphological characteristics and immunohistochemical features of this rare lesion is hence discussed.

1. Introduction

Small cell glioblastoma (SCGBM) is a rare variant with monomorphous cells and deceptively bland nuclei and are often misdiagnosed. They account for 10% of all GBMs, with another 11% showing focal, small cell features.[1] Brain tumors that arise from deep brain are relatively rare with low grade astrocytomas being the commonest.[2] Here, we present a case of small cell GBM in a 39-year-old male involving bilateral basal ganglia.

2. Case Report

A 39-year-old female was found to have bilateral basal ganglion lesion diagnosed as primary central nervous system lymphoma on MRI. Stereotactic biopsy was taken. Biopsy specimen received as multiple fragments of grey brown tissue measuring about 20 mm in aggregate diameter grossly.

Microscopically, the lesion was seen to exhibit ganglia exhibit a loose, diffuse distribution of uniform, monotonous appearing cells with lymphoid configuration, small, medium, on large in size with indistinct cytoplasmic margins [Figure 1].

The scattered large cells and a few exhibiting cleaved or lobed appearance with small well defined prominent nucleoli or occasionally displaying coarse chromatic clumping was observed throughout the lesion [Figure 2]. A brisk mitotic activity of 1-2 per HPF was also noted. Occasional foci of closely packed large cells mimicking rosette formation were seen in a few foci. Trapped within, large areas of haemorrhagic congestion and foci of necrosis were observed.

An interpretation of high grade blue round cell tumor was made and further immunohistochemical studies were carried out.

The diffuse strong positivity noticed for GFAP, S100, and a proliferative index of 70% along with morphological features indicative of small cell glioblastoma (SCGBM) was noted [Figure 3]. Though morphologically the features mimicked a high-grade lymphoma the panel of lymphoma markers carried out were completely negative for the tumour component.

3. Discussion

Although SCGBM was defined and clarified already and its association with EGFR amplification was emphasized recently, SCGBM still remains as an under-recognised variant.[1]

* *Corresponding author*.
E-mail address: baranisri@gmail.com (B. Karikalan).

https://doi.org/10.18231/j.ijpo.2020.069

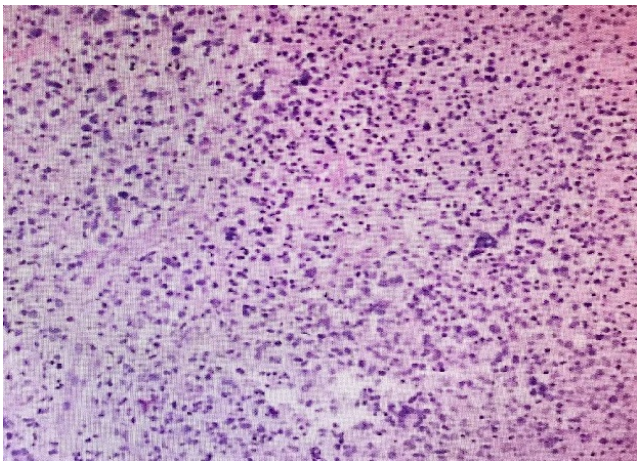

Fig. 1: Small cells 20X

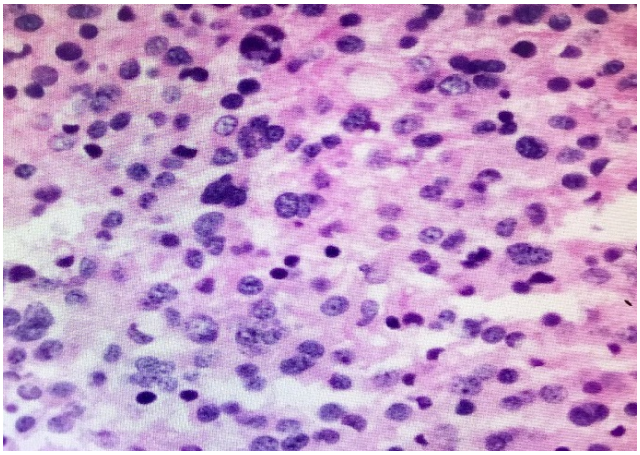

Fig. 2: Small cells 40X

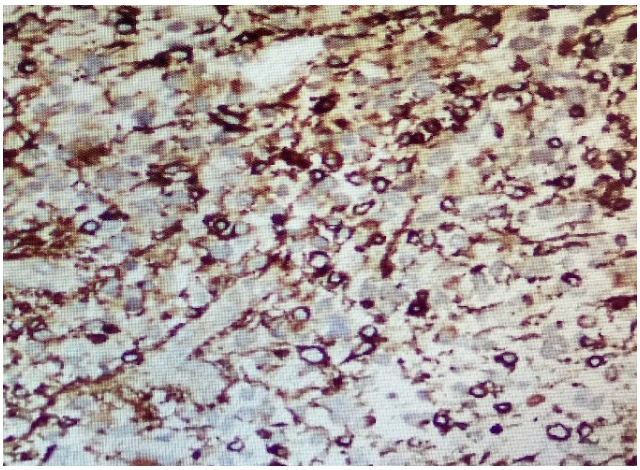

Fig. 3: GFAP positive

SCGBM is characterized by monomorphic proliferation of cells with small, round nuclei, scant cytoplasm, mild hyperchromasia, less stroma, and low mitotic index.[3] They account for approximately 10% of GBM diagnoses with another 11% showing focal areas of small cell configuration.[1] SCGBM are often misdiagnosed as high grade oligodendroglial tumors or low grade astrocytomas. They are aggressive lesions said to parallel grade 4 gliomas.

Many studies reported presence of foci of small cell morphology in lesions diagnosed as primary glioblastoma multiforme [GBM] and had EGFR amplification.[4,5] One study reported 88% of GBM with small cell features to have EGFR amplication when compared to 42% of control [GBM with no small cell features]. The results were also validated by larger set of neoplasms where 62% of primary GBM with small cell features showed EGFR amplification.[4] Another study that included 56 cases, showed 64% positivity for EGFR amplification as opposed to 23% in control via insitu hybridization.[5]

Another interesting fact about SCGBM is that its association with human polyomavirus JCV infection. A study showed that the glial and epithelial microdissected components of SCGBM showed presence of human polyomavirus JCV suggesting possible role of infection in the genesis of these lesions.[6]

Shortly after SCGBM was accepted as a distinct variant, many studies were done retrospectively on GBMs to study the prevalence of this newly recognised variant. In a study that included 229 GBMs, 71 tumors had more than 80% of the samples showing small cell morphology and so were concluded to be SCGBM. Another 11% of the tumors had significant small cell morphology but in less than 80% of the samples. About 37% of these SCGBMs showed minimal or no lesion enhancement in radiology and 33% showed no necrosis or endothelial proliferation and hence were classified as grade 3 astrocytomas. But the mortality of these lesions were similar to grade 4 SCGBMs. The study also suggested SCGBM mimicking brain tumor to be oligodendrogliomas, due to some of the features including chicken wire capillary networks, perinuclear clearing, per neuronal satellitosis and microcalcifications found in SCGBMs. However, oligodendrogliomas can be distinguished by their unique presence of 1p/19q deletion which was consistently absent in SCGBMs. Despite, the use of these molecular markers for distinction of SCGBM from similar lesions, the authors also gave a list of histological features that can be used to define SCGBM that includes ring enhancement, pseudopalisading necrosis, oval nuclei, high proliferation index and thin GFAP-positive cytoplasmic processes. The survival rate for SCGBM is not different from that of classic GBM which is 14.3 months, after age and type of surgery adjustments.[7]

The other important non-glial tumor that has to be differentiated from SCGBM are primary central nervous

system lymphomas (PCNSL) because of the morphological resemblance of small cells to the lymphoid cells. PCNSLs account for 2-4% of primary brain tumors and 35% of them are known to occur in basal ganglia.[8] It is easy to distinguish lymphomas by immunohistochemical markers but has to be suspected while dealing with small cell morphology.

Tumors arising from deep brain like basal ganglia and thalamus are quite rare. Most common are low grade astrocytomas. Primitive neuroectodermal tumors, ganglion cell tumors, oligodendrogliomas, lymphomas and germinal neoplasms are other tumors that are known to arise from deep brain.[2] Our case is unique in a way to report bilateral involvement of basal ganglia by SCGBM.

4. Conclusion

Thus, a case of SCGBM is reported for its rarity and unique location in this case.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Perry A, Aldape KD, George DH, Burger PC. Small cell astrocytoma: An aggressive variant that is clinicopathologically and genetically distinct from anaplastic oligodendroglioma. *Cancer*. 2004;101(10):2318–26.
2. García-Santos J, del Río ST, Sánchez A, Martínez-Lage J. Basal ganglia and thalamic tumours: an imaging approximation. *Childs Nerv Syst*. 2002;18(8):412–25.
3. Louis DN, Ohgaki HH, Wiestlerod, Louis DN, Ohgaki HH, Wiestler OD, et al. Astrocytic tumors. In: WHO classification of tumours of the central nervous system. Albany: WHO Press; 2007.
4. Burger PC, Pearl DK, Aldape K, Yates AJ, Scheithauer BW, Passe SM, et al. Small cell architecture – a histological equivalent of EGFR amplification in glioblastoma multiforme? *J Neuropathol Exp Neurol*. 2001;60(11):1099–1104.
5. Quezado M, Ronchetti R, Rapkiewicz A, Santi M, Blumenthal DT, Rushing EJ. Chromogenic in situ hybridization accurately identifies EGFR amplification in small cell glioblastoma multiforme, a common subtype of primary GBM. *Clin Neuropathol*. 2005;24:163–169.
6. Piña-Oviedo S, León-Bojorge BD, Cuesta-Mejías T, White MK, Ortiz-Hidalgo C, Khalili K, et al. Glioblastoma multiforme with small cell neuronal-like component: association with human neurotropic JC virus. *Acta Neuropathol*. 2006;111(4):388–96.
7. Birner P, Toumangelova-Uzeir K, Natchev S, Guentchev M. Expression of mutated isocitrate dehydrogenase-1 in gliomas is associated with p53 and EGFR expression. *Folia Neuropathol*. 2011;49:88–93.
8. Malikova H, Koubska E, Weichet J, Klener J, Rulseh A, Liscak R, et al. Can morphological MRI differentiate between primary central nervous system lymphoma and glioblastoma? *Cancer Imaging*. 2016;16(1):40.

Author biography

Barani Karikalan Senior Lecturer

Thanikachalam Pasupati Consultant Pathologist

Cite this article: Karikalan B, Pasupati T. Small cell glioblastoma of basal ganglia: Case report. *Indian J Pathol Oncol* 2020;7(2):347-349.