

Primitive Polar Spongioblastoma: A Rare Histopathological Entity

Shilpi Bhargava, MD*

Kumud Gangwal, MSc**

Shubha Gupta, MD***

Mansi Faujdar, MD****

Deepa Wadhvani, DPB*****

*Senior Resident, Department of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

**Senior consultant, Department of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

***Senior consultant, Department of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

****Consultant, Department of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

*****Senior resident, Department of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

Abstract

Primitive Polar spongioblastoma is a very rare histopathological entity characterized by bipolar tumor cells with palisading nuclei showing immunoreactivity for neuron-specific enolase. The entity was first described by Russell and Cairns in 1947. It is classified as a high grade glioma with a very poor prognostic outcome. However, similar histological profile is often seen in many neuroepithelial tumors, and this category was earlier excluded from the World Health Organization (WHO) classification raising the question of its existence as a tumour entity. Presently, it is included in WHO classification in the category of CNS tumour of uncertain origin. To our knowledge, only a few cases of Primitive polar spongioblastoma have been reported so far. The present case of a 65 yr old male clinically presenting with vomiting & altered sensorium. MRI findings were inconclusive suggesting a mass lesion in the temporal lobe & a presumptive diagnosis of tumoral bleed was given.

Keywords: Glioma; Neuroepithelial; Palisading; Spongioblastoma.

Introduction

Polar spongioblastoma was first described by Russell and Cairns in 1947. It is a high grade glioma characterised by the parallel palisading of spindle tumor cells without microvascular proliferation (MVP) and necrosis. Different

views have been suggested regarding the existence of this rare entity as nuclear palisades can be found as local architectural features in many neuroepithelial tumors like ependymomas and neuroblastomas.

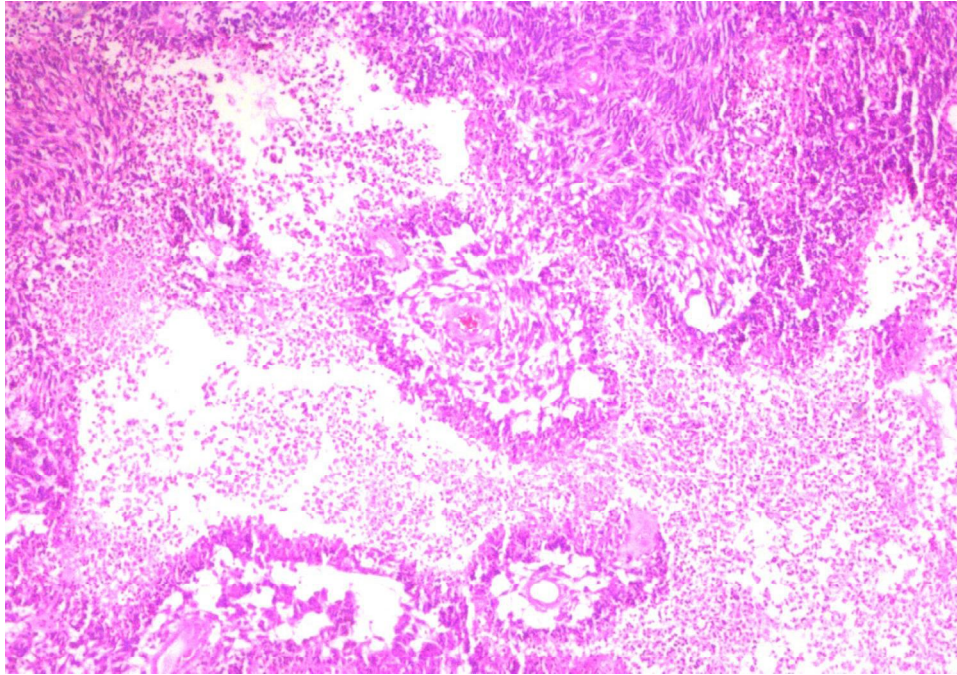
Case Presentation

A case of a 65 yr old male clinically presenting with frequent vomiting, altered sensorium & headache. CT scan findings were inconclusive. EEG was normal but MRI suggested a mass lesion in the temporal lobe,

Corresponding Author: Dr. Shilpi Bhargava, Department Of Pathology, Santokba Dhurlabji Memorial Hospital, Jaipur, Rajasthan,

E-mail: Drshilpi222@gmail.com

(Received on 08.04.2013, Accepted 18.04.2013)

Figure 1: H & E -100X-Tumour Cells In Palisading Pattern

measuring 3x2x2.5 cm, a provisional report of tumoral bleed was given. Per-operatively, a highly vascular, non suckable tumour mass was seen and excisional biopsy was performed. Histopathological examination was done. Grossly, the tumour consisted of multiple greyish white soft tissue pieces. On microscopic examination, a highly malignant neoplasm

composed of bipolar cells arranged in palisades of long columns was seen with intervening fibrillary zone in a linear arrangement (Figure 1, 2). Nuclei are hyperchromatic, round to oval. Mitotic activity is scant. IHC analysis revealed NSE positivity (Figure 3) & negative CK. The confirmed histopathological diagnosis of Primitive polar spongioblastoma was given.

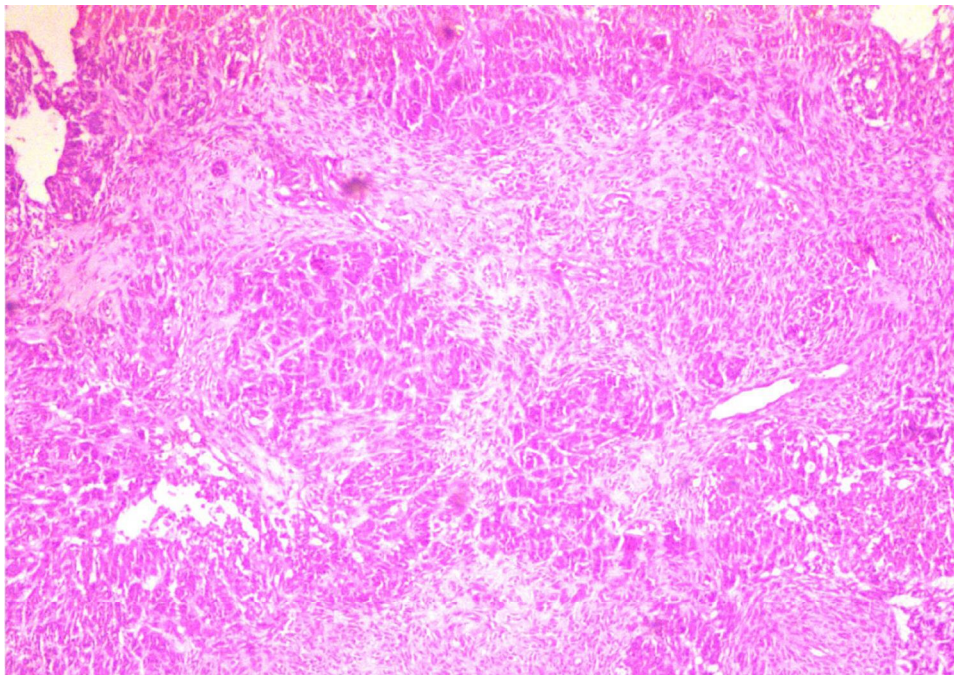
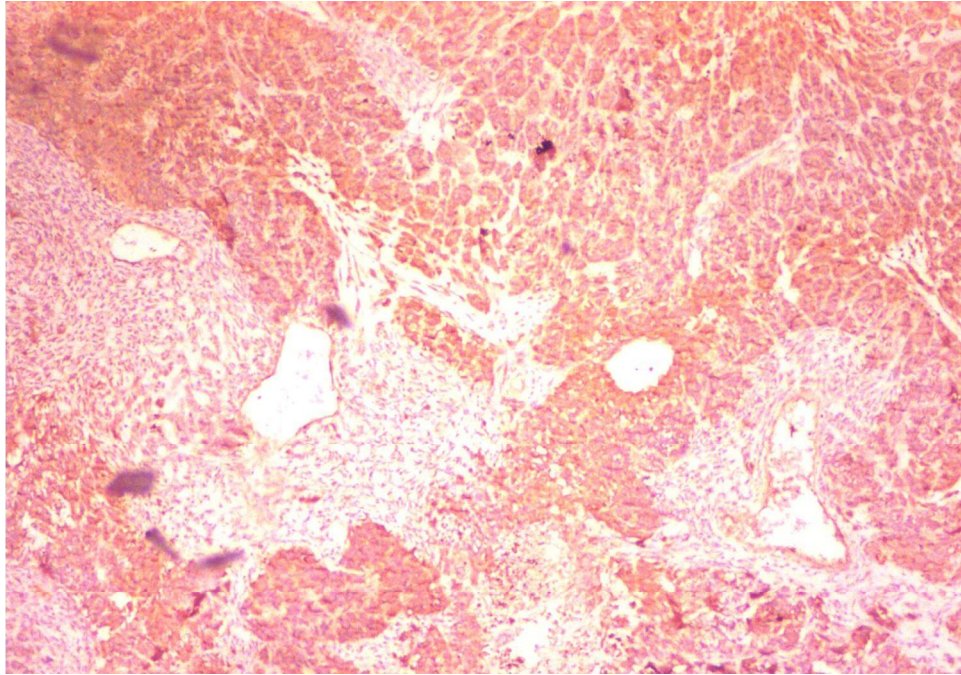
Figure 2: H & E-400X-Columns Of Bipolar Cells With Intervening Fibrillar Zone

Figure 3: Tumour Cells Showing Nse Positivity- (X400)

Discussion

A study by Narita Y and Fukushima S[1] in 2012 in a 44 yr old adult who presented with a headache exhibited a tumor of the right frontal lobe on MRI. Histological diagnosis of the tumor obtained by gross total resection was high-grade glioma, which was composed of the parallel palisading of spindle tumor cells, final diagnosis of a high grade glioma with histological pattern of polar spongioblastoma was made. It was suggested that this tumor might not be suited to any of the neuroepithelial tumors in the current WHO classification. Contrary to these findings, in a study by Schiffer D[2] in 1993, The distribution of cells in a parallel fashion with palisades of nuclei is common in neuroepithelial tumors and thus polar spongioblastoma does not exist as a tumor entity as pilocytic astrocytomas, oligodendrogliomas, medulloblastomas, cerebellar astrocytomas, neuroblastoma also present with similar histological profile. Concordance to the earlier views was seen in a case report of a 14 months old baby by Langford[3] (1987) stating that the palisading pattern in cerebral neuroblastoma mimicks juvenile polar spongioblastoma. Ultrastructural examination of this single case reveals cells that

range from the embryonal neuroepithelial cell to neurons with synapse formation. The diagnostic pattern characteristic of the polar spongioblastoma may be found in tumors of neuronal origin, although most polar spongioblastomas having the typical pattern of palisading seem to belong to the astrocytic cell line. A study by G.H. Jansen *et al*[4] in 1990, revealed juvenile polar spongioblastoma as a distinct electron microscopic entity with ultrastructural features of developing neuronal elements. These findings are in contrast with the longheld view that the polar spongioblastoma is cytogenetically related to the embryonal radial glial cells as revealed in a study by Chandarevian *et al*[5] 1984. The most important leading view in this respect comes from the current WHO classification which includes primitive polar spongioblastoma in neuronal tumours of uncertain origin.[6] The rhythmic pattern of uni and bipolar cell arrays is exclusively represented and aggressive biological behavior makes it a distinct entity.[7] Studies have proved that presence of a such palisading pattern mimicking polar spongioblastoma in an otherwise well differentiated tumour like cerebellar astrocytoma should be treated with aggressive chemotherapy & radiotherapy.[8]

References

1. Fukushima S, Narita Y *et al*. A case of unclassified high-grade glioma with polar spongioblastoma pattern. *Acta Neuropathology*. 2012; 32(6): 604-10.
2. Schiffer D, Cravioto H *et al*. Is polar spongioblastoma a tumor entity? *J Neurosurg*. 1993; 78(4): 587-91.
3. LA Langford, MH Camel. Palisading pattern in cerebral neuroblastoma mimicking the primitive polar spongioblastoma. *Acta Neuropathologica*. 1987; 73(2): 153-159.
4. GH Jansen, Troost *et al*. Polar spongioblastoma: an immunohistochemical and electron microscopical study. *Acta Neuropathologica*. 1990; 81(2): 228-232
5. Chadarévian *et al*. Neuroendocrine nature of Polarspongioblastoma: Case report. *Acta Neuropathologica*. 1984; 77(3).
6. Kleihuer P, Burger PC, Scheithauer BW. Histological typing of tumours of CNS. 2nd ed. New York: Springer -Verlag; 1993.
7. Russell DS, Rubinsten LJ. Pathology of tumours of nervous system. Baltimore; Williams and Wilkins; 1989, 169-172.
8. Schochet SS, Violet TW, Nelson J, Pelofsky S, Barnes PA. Polar Spongioblastoma of cervical spinal cord: Case report. *Clin Neuropathol*. 1984; 3: 225-7.