ISSN: 2394-0026 (P)
ISSN: 2394-0034 (O)

Case Report

Dysembryoplastic neuroepithelial tumor: A rare case report

Kuntal Patel[1], Gunvanti Rathod[2*]

[1]P.G. Student, Pathology Department, SBKS MI & RC, Sumandeep Vidyapeeth, Vadodara, India
[2]Assistant Professor, Pathology Department, SBKS MI & RC, Sumandeep Vidyapeeth, Vadodara, India
[*]Corresponding author email: neempath@gmail.com

How to cite this article: Kuntal Patel, Gunvanti Rathod. Dysembryoplastic neuroepithelial tumor: A rare case report. IAIM, 2015; 2(5): 164-167.

Available online at www.iaimjournal.com

Received on: 29-03-2015 **Accepted on:** 11-04-2015

Abstract

Dysembryoplastic neuroepithelial tumor (DNET) is a rare benign tumor generally seen in children and adolescents with intractable epilepsy. This tumor demonstrates typical histological features such as glial nodules and the so called glioneuronal element. The diagnosis of DNET cannot rely just upon imaging features but needs a multidisciplinary contribution from the clinical and diagnostic department involving the clinician, radiologist as well as the pathologist to reach a definite and conclusive diagnosis. However histopathology is the gold standard for the final diagnosis.

Key words

Dysembryoplastic neuroepithelial tumor, Glioneuronal element, Histopathology.

Introduction

The term dysembryoplastic neuroepithelial tumor (DNET) was first coined by Daumas‐Duport and colleagues to describe a cortical lesion presenting in childhood [1]. It is a rare benign tumor generally seen in children and adolescents with intractable epilepsy. The typical radiological findings of DNET are supratentorial tumor generally affecting the temporal lobes and without any peritumoral edema or mass effect [2]. This tumor demonstrates typical histological features such as glialnodules and the so called glioneuronal element [3, 4]. Their favorable prognosis is also due to the fact that most lesions remain stable, yet rare cases with slow progression or haemorrhage due to hamartomatous vessels have been reported [5, 6]. Herein we have presented a case report of a patient with diagnosis of DNET where we are able to find and document the typical features of DNET.

Case report

A 13 year old male patient came in the neurosurgery outpatient department of Dhiraj Hospital with complaint of right sided headache

ISSN: 2394-0026 (P)
ISSN: 2394-0034 (O)

for 1 year and history of convulsion 2 months back. He also gave history of convulsion for few times 1 year back. His routine neurological examination and laboratory examination were within normal limit. CT scan of brain revealed focal edema in left posterior parietal region. **(Photo - 1)** MRI brain revealed a well defined wedge shaped cortical based bubbly appearing lesion seen in the left parieto-occipital region. T2W images showed a well defined hyper intense lesion. Overall appearance was suggestive of cortical neuroglial tumor with possibility of DNET. **(Photo - 2)** Excision biopsy was taken from the brain tumor and sent for the histopathological examination. The microscopic examination showed focal hypocellular area with oligodendrocytes like cells. **(Photo - 3)** The stroma showed microcystic pattern and myxoid background. **(Photo - 4)** The tumor cells express S-100 **(Photo - 5)** and negative for **GFAP. (Photo - 6)** The morphological and immunohistochemical features were those of DNET.

Photo – 1: CT scan of brain revealed focal edema in left posterior parietal region.

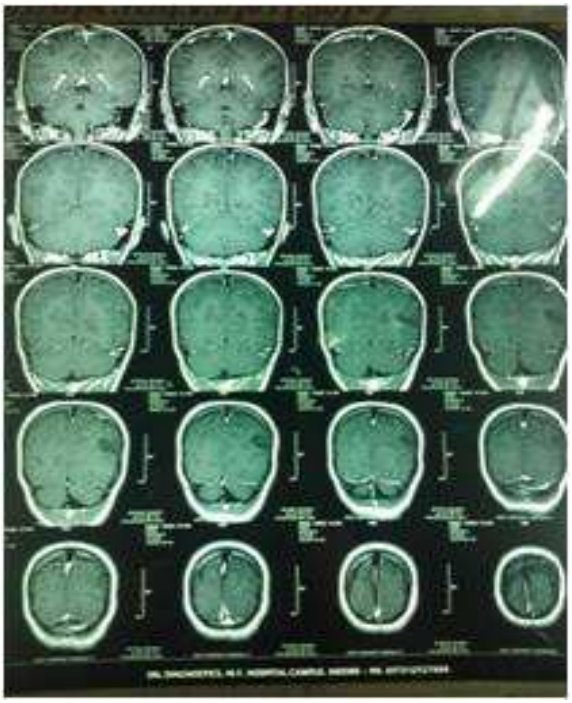


Photo – 2: MRI brain revealed a well defined wedge shaped cortical based bubbly appearing lesion seen in the left parieto-occipital region.

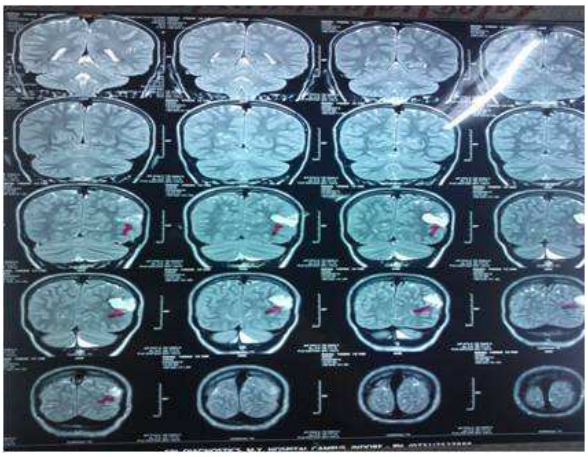
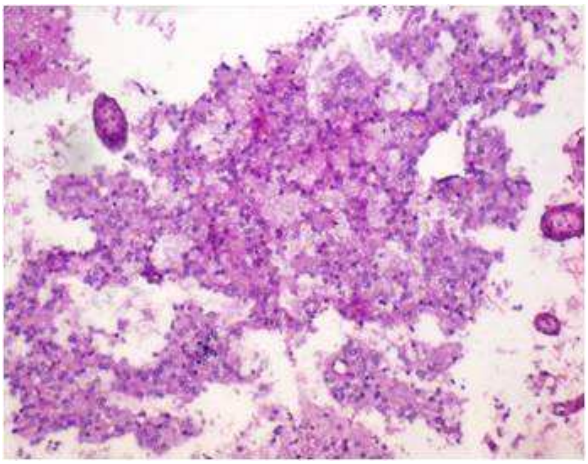


Photo – 3: Focal hypocellular area with oligodendrocyte like cells (H & E Stain, 4X).



Discussion

In 1988, Dumas-Duport, et al. first used the term dysembryoplastic neuroepithelial tumor to describe low-grade tumors found in young patients with intractable partial seizures [1]. In 1993, the distinct pathological entity known as DNET was given a place in the WHO classification of brain tumors as a grade I tumor of neuroepithelial origin [7, 8, 9]. Generally DNET is a benign tumor but there have been cases of malignant transformation [10].

ISSN: 2394-0026 (P)
ISSN: 2394-0034 (O)

Photo – 4: Microcystic pattern in dysembryoplastic neuroepithelial tumor (H & E Stain, 4X).

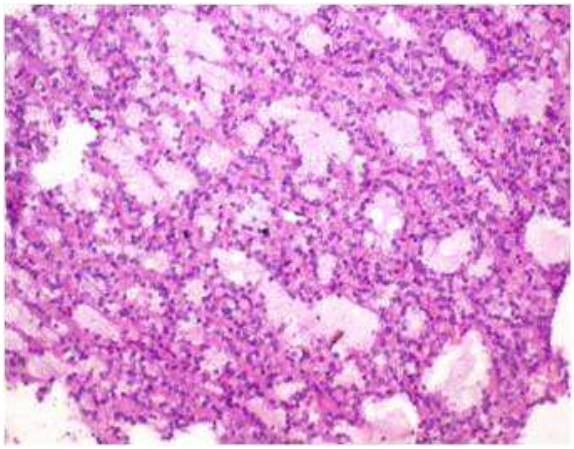


Photo – 5: Strong S-100 positivity in DNET.

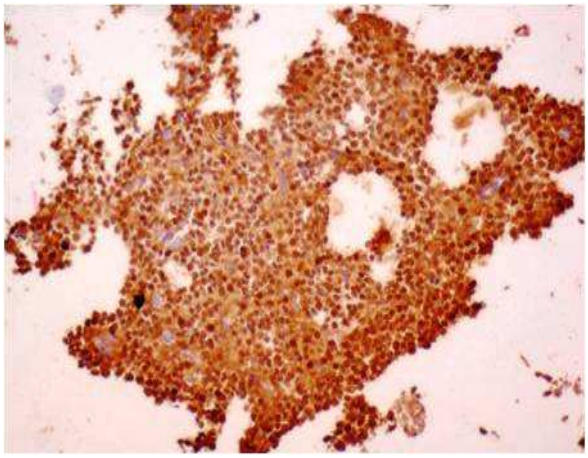
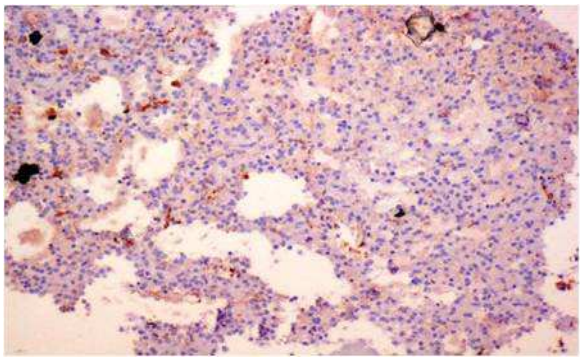


Photo – 6: GFAP negativity in DNET.



This tumor generally affects children and young adults and presents with a history of intractable seizures. Our patient was also a 13 year old male who presented with a history of headache and intractable seizures.

On neuroimaging DNETs are cortical lesions with little mass effect and a predilection for the temporal lobes. On computed tomography DNETs are typically well-demarcated, hypodense, cortical lesions that can be associated with deformation of the overlying skull. MR images often show a solid and cystic mass with the cystic portions appearing slightly more intense than cerebrospinal fluid. The solid components often appear multinodular, hypointense on T1-weighted MR images, hyper intense on T2-weighted MR images, and occasionally weakly enhancing [11, 12].

Three histologic forms of DNET have been described: complex, simple, and nonspecific [13]. The histopathological hallmark is bundles of axons lined by oligodenroglia-like cells, forming columns in a pale mucoid matrix in which isolated neurons float. These so called glioneuronal elements are observed both in simple and complex forms of DNET. The heterogenous appearance of the latter is due to additional glial or neuronal cell populations which mimic low- grade gliomas. Our patient was presented with a history of intractable seizures and histology showed the typical features of DNET including glioneuronal elements.

Since most patients with DNET following surgery have shown excellent recovery and the seizure in them are more or less fully controlled. These patients need not to be subjected to any radiation or chemotherapy .therefore it is essential that DNET should be accurately diagnosed [14]. The diagnosis of DNET cannot rely just upon imaging features but needs a multidisciplinary contribution from the clinical and diagnostic department involving the clinician, radiologist as well as the pathologist to reach a definite and conclusive diagnosis [11]. However, histopathology is the gold standard for the final diagnosis.

ISSN: 2394-0026 (P)
ISSN: 2394-0034 (O)

Conclusion

DNET is a rare benign glioneuronal tumor of the central nervous system. The patients of DNET with seizures are excellent surgical candidates and having low recurrence rate. As the lesion carries a favorable prognosis and these patients do not require radiation following surgery, it becomes very essential that this lesion should be accurately diagnosed by the surgical pathologist.

References

1. Daumas‐Duport C, Scheithauer BW, Chodkiewicz JP, Laws ER Jr, Vedrenne C. Dysembryoplasic neuroepithelial tumor: A surgically curable tumor of young patients with intractable partial seizure. Report of thirty‐nine cases. Neurosurgery, 1988; 23: 545‐56.
2. Daumas-Duport C, Varlet P. Dysembryoplastic neuroepithelial tumors. Rev Neurol (Paris), 2003; 159: 622-36.
3. Daumas-Duport C, Varlet P, Bacha S, Beuvon F, Cervera –Pierot P, Chodkiewicz JP. Dysembryoplasic neuroepithelial tumor: Nonspecific histological forms - A study of 40 cases. J Neurooncol., 1999; 41: 267-280.
4. Prayson RA, Morris HH, Estes ML, Comair YG, Dysembryoplasic neuroepithelial tumor: A clinopathologic and immunohistochemical study of 11 tumors including MIB1 immunoreactivity. Clin Neuropathol., 1996; 15: 47-53.
5. Ostertum B, Wolf HK, Campos MG. Dysembryoplasic neuroepithelial tumor: MR and CT evaluation. Am J Neuroradiol., 1996; 17: 419-430.
6. Thom M, Gomez-Anson B, Revesz T. Spontaneous intra lesional haemorrhage in Dysembryoplasic neuroepithelial tumor: A series of five cases. J Neurol Neurosurg Psychiatry, 1999; 67: 97-101.
7. Dumas-Duport C. Dysembryoplastic neuroepithelial tumours. Brain Pathol, 1993; 3**:** 283-295.
8. Kleihues P, Burger PC, Scheithauer BW. The new WHO classification of brain tumors. Brain Pathol, 1993; 3**:** 255-268.
9. Louis DN, Ohgaki H, Wiestler OD, et al. The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol, 2007; 114: 97-109.
10. Ray WZ, Blackburn SL, Casavilca-Zambrano S, Barrionuevo C, Orrego JE, Heinicke H, et al. Clinicopathologic features of recurrent dysembryoplastic neuroepithelial tumor and rare malignant transformation: A report of 5 cases and review of the literature. J Neurooncol, 2009; 94: 283-92.
11. Fernandez C, Girard N, Paz PA, et al. The usefulness of MR imaging in the diagnosis of dysembryoplastic neuroepithelial tumor in children: A study of 14 cases. Am J Neuroradiol, 2003; 24: 829-834.
12. Stanescu CR, Varlet P, Beuvon F, et al. Dysembryoplastic neuroepithelial tumors: CT, MR findings and imaging follow-up: A study of 53 cases. J Neuroradiol, 2001; 28: 230-240.
13. Chan CH, Bittar RG, Davis GA, et al. Long-term seizure outcome following surgery for dysembryoplastic neuroepithelial tumor. J Neurosurg, 2006; 104: 62-69.
14. Burger PC, Scheithauer BW, editors. Tumors of the central nervous system. Atlas of Tumor Pathology, 3rd series, Washington, DC: Armed Forces Institute of Pathology; 1994, p. 184-187.

Source of support: Nil
Conflict of interest: None declared.