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DYSEMBRYONIC NEURO-EPITHELIAL TUMORS

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ABSTRACT

Introduction: Dysembryonic Neuro-Epithelial Tumors are mixed neuro-epithelial tumors affecting most commonly the temporal lobe. They usually result in epilepsy which is intractable in nature. Children and young adults are much more prone to exhibit these Dysembryonic Neuro-Epithelial Tumors.

Case presentation: Here, we present a case series of two male patients who presented to the department of radiodiagnosis at Dr. Vasant Rao Pawar Medical College and Research Centre for MRI evaluation with complaints of epilepsy.

Conclusion: Dysembryonic Neuro-Epithelial Tumors are a mixed neuronal glial neoplasm, mainly affecting the temporal lobe and is a very common cause of epilepsy. They are usually benign tumors with no or mild contrast enhancement.

Key words: Dysembryonic Neuro-Epithelial Tumors, Temporal lobe, Epilepsy, Pseudocystic

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INTRODUCTION

Dysembryonic Neuro-Epithelial Tumors (DNET) are a mixed neuronal-glial neoplasm affecting most commonly the temporal lobe. It is most commonly seen in children and young adults. Patients most commonly present with intractable seizures.[1]

CASE PRESENTATION

Two male patients came to the department of radiodiagnosis at Dr.Vasant Rao Pawar Medical College and Research Centre for MRI evaluation. Both these patients had intractable seizures refractory to treatment. Patient one was 20 years old and patient two was 44 years old. Both the patients had no significant past history, personal history or family history. Both the patients developed sudden onset seizures. The seizures responded poorly to regular anti-epileptic medications. There were no predisposing factors for the seizures. The seizures were mostly generalized tonic clonic type.

MRI Findings – MRI was performed on 1.5T magnet MR system (Siemens magnetomEssenza). Imaging was performed using a head coil.

Patient one –

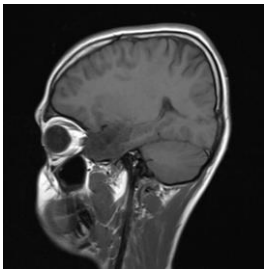


Fig 1

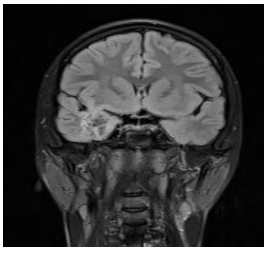
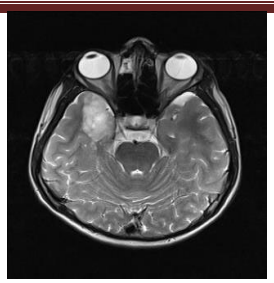


Fig 2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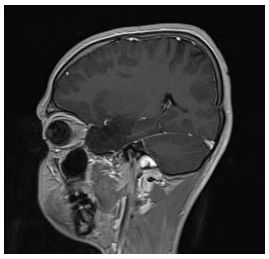
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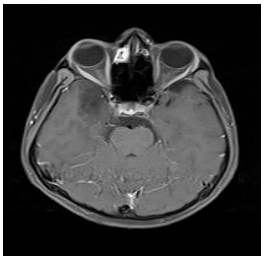


Fig 5

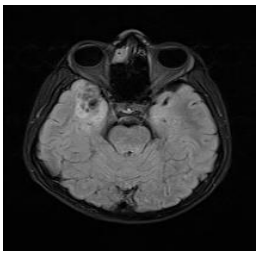


Fig 6

Patient two –

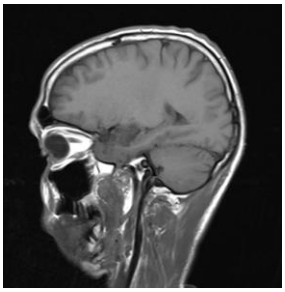
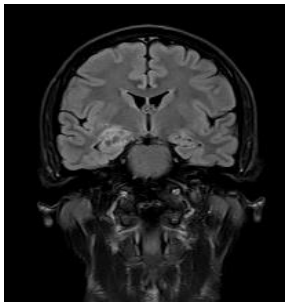


Fig 1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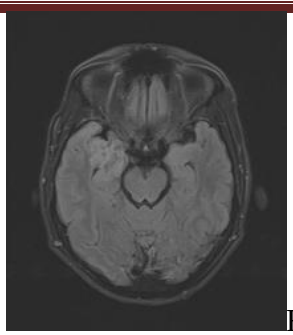
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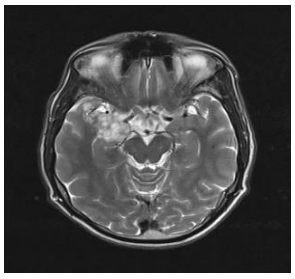
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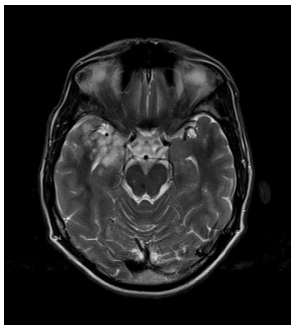
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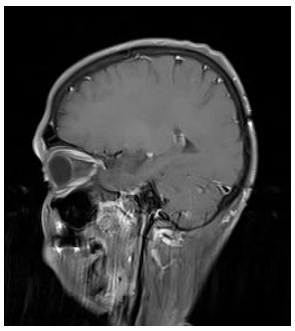
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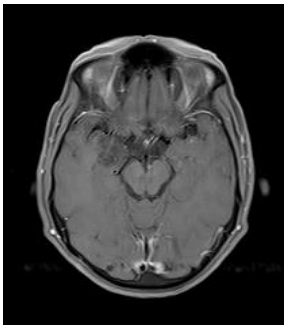
Fig 7

The imaging findings in our patients corraborated with those given in the literature. The lesions were iso to hypointense on T1 and hyperintense on T2 and FLAIR. The lesions did not show contrast enhancement. The lesion were multi-cystic in

nature with typical bubbly appearance. In both the patients the lesions were seen at the temporal lobe.

DISCUSSION

DNETs are mostly benign tumors with rare malignant transformation, first described by Daumas Duport et al. [2-4] They are a tumor of mixed neuronal and glial origin.[5] They are mucinous or gelatinous tumors with friable consistency.[5] It is mostly a tumor of pediatric population but can also be seen in adults. They show hypointense signal on T1WI and hyperintense on T2WI.[1] The tumor does not exhibit mass effect, edema, hemorrhage or calcification.[1,6] The patients present with intractable epilepsy without any neurological deficit.[7] The patients usually have a normal intelligent quotient.[7] It is more commonly seen in males.[8]

The most common site affected is temporal lobe followed by frontal lobe.[8] Other sites are caudate nucleus, cerebellum and pons.[8] These tumors are derived from secondary germinal layers. It is sometimes associated with neurofibromatosis I and Klinefelter's syndrome.[4]

The tumor consists of nodular architecture and a specific glioneuronal element with oligodendrocyte like cells.[7]

The matrix of the tumor consists of abundant interstitial myxoid content and this gives the tumor a multi-cystic appearance. Usually there is no contrast enhancement or mild contrast enhancement. On FLAIR images, there is a hyperintense rim surrounding the tumor, this is sensitive and specific for DNET.[1] It generally has a soap bubble appearance at the cortical margin.[8]

DNETs are of two types – Simple and Complex. Simple forms donot contain glial nodules and hence donot have nodular architecture while complex forms have glial nodules and multinodular architecture.[7-9] Surgery is required to cure epilepsy and prevent hemorrhagic complications.[2] However, there is high chance of recurrence.[3] The main differential diagnosis of DNETs are Oligodendroglioma and Gangliogliomas.[5,10]

As DNETs originate from the dysembryoblastic cells, therefore, radial glial fibres pathway give it a triangular pattern.[2] The other features seen are septations.

CONCLUSION

DNET is a mixed neuronal glial neoplasm, mainly affecting the temporal lobe and is a very common cause of epilepsy. They are usually benign tumors with no or mild contrast enhancement.

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