*IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*
*e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 18, Issue 1 Ser. 9 (January. 2019), PP 13-16*
*www.iosrjournals.org*

# The Girl Who Mistook Herself for a Boy- A Rare Case Report

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***Abstract****: We report a rare case of an 8year old female child who presented with 9months history of memory disturbances, behavioral and personality disturbances in the form of aggressiveness, lack of fear, disinhibition, auditory and visual hallucinations, gender misidentification, inability to recognize the parents, identifying everyone as transgender and using abusive language. Initially she was diagnosed as childhood psychosis/ conversion disorder by a psychiatrist and treated with antipsychotics. There was no response to treatment. Further evaluation revealed an interesting underlying organic cause [space occupying lesion of the temporal lobe] presenting with neuropsychiatric disturbances.*

***Key words-*** *Behavioral and personality disturbances, neuropsychiatric illness, gender misidentification, organic lesion, DNET, ICSOL of temporal lobe.*

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Date of Submission: 04-01-2019 Date of acceptance: 19-01-2019

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## I. Introduction

Patients presenting with behavioral and personality disturbances pose a diagnostic challenge to both the neurologists and psychiatrists. Often an organic lesion can present as a psychiatric disturbance. Accurate diagnosis and intervention are required to avoid unnecessary use of antipsychotic medication especially in children.

## II. Case Report

An 8year old female child presented with insidious onset gradually progressive behavioral and personality disturbances and memory disturbances of 9months duration. Altered behavior in the form of aggressiveness, arrogance, running away from home and injuring herself. She speaks irrelevantly, uses an abusive language. She identifies her parents and sibling as people living in neighborhood. She asks for different kinds of junk food at odd times in a day and if any of her wishes were not fulfilled she would beat the parents in rage. She winks at girls whom she encounters on her way. She has the urge of removing all the clothes over the body every night and prefers to sleep naked. She claims that she is a boy of age 6years and forced her family members to get her head shaved because of this gender misidentification. History of auditory and visual hallucinations were present in the form of hearing unknown voices and sounds and seeing some unknown people entering her room. Memory disturbances were in the form of difficulty remembering her family members, school teachers and friends. For these complaints, she initially presented to a psychiatrist and was diagnosed as early onset psychosis/conversion disorder and was started on antipsychotics. As her symptoms did not improve she was referred to our department to rule out any underlying organic cause.

On examination-Patient is conscious with elated mood. Not well behaved. Mini-Mental state Examination- She scored 18 out of 30. Using abusive language. Recent memory was impaired. Rest of the neurological examination was normal.

Routine blood investigations were normal. CT Brain revealed a hypodense lesion in right medial temporal lobe. MRI Brain revealed a 27×25×23mm ill defined non enhancing lesion which is hypointense on T1, hyperintense on T2 and FLAIR involving right antero-medial temporal lobe, right basal ganglia and posterior thalamus with no surrounding edema or mass effect. Based on these findings a radiological diagnosis of Dysembryoplastic NeuroEpithelial Tumor [DNET] was thought of. EEG showed right temporal slowing. Craniotomy and excision of tumor was done. Post operative period was uneventful. Histopathology examination showed myxoid cells with floating neurons and oligodendroglia like cells suggestive of DNET.

Patient's behavioral and psychiatric symptoms have subsided completely following surgery. She started going to school normally and her scholastic performance has improved.

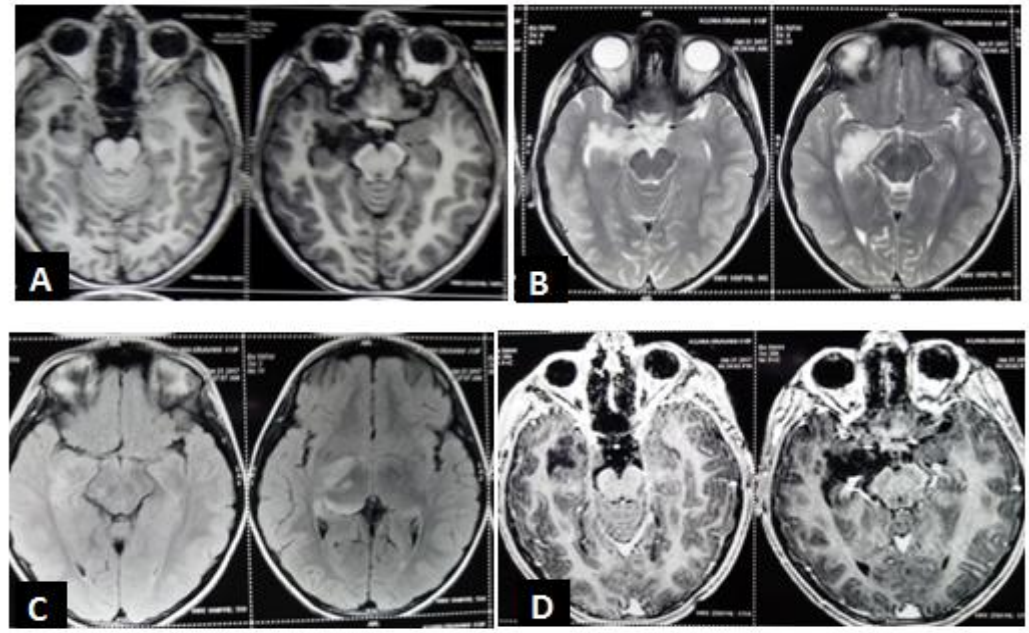


**FIGURE 1:** A)T1 axial B) T2 axial C) T2 FLAIR axial D) T1 contrast axial E) T1 contrast coronal images showing 27×25×23mm ill defined non enhancing lesion which is hypointense on T1, hyperintense on T2 and FLAIR involving right antero-medial temporal lobe, right basal ganglia and posterior thalamus suggestive of Dysembryoplastic NeuroEpithelial Tumor [DNET].

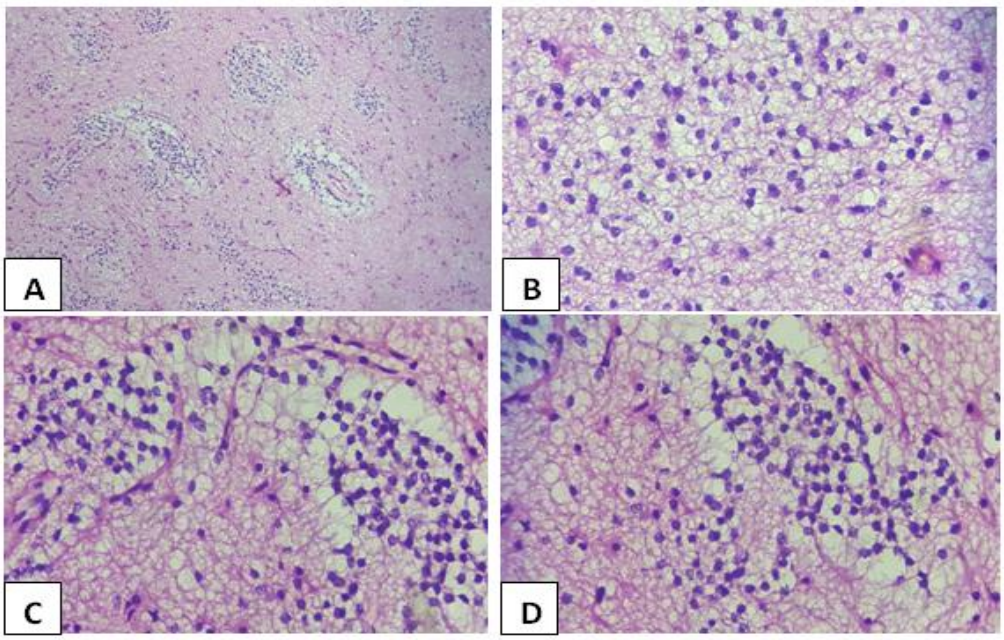


**FIGURE 2- A) B) C) D) Histopathological examination of the resected tumor showing Myxoid cells with floating neurons and oligodendroglia like cells.**

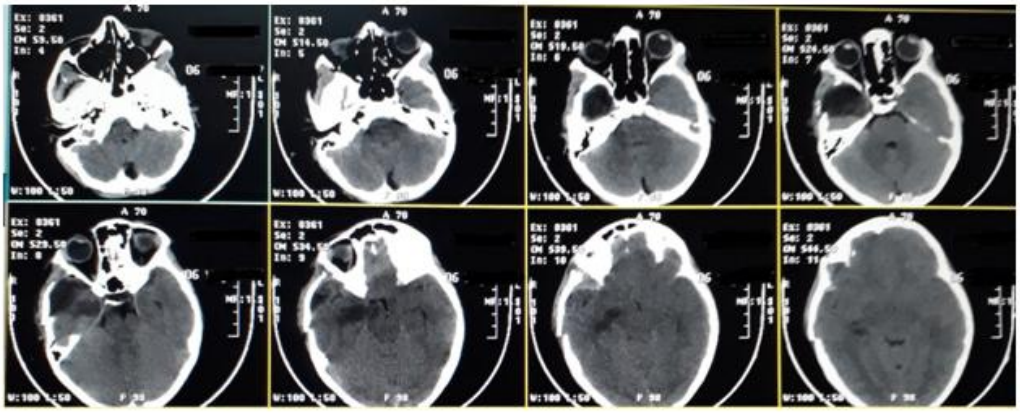

**FIGURE 3- Post Operative CT Scan of the patient showing complete excision of the tumor.**

## III. Discussion

The term DNET was proposed by Daumas-Duport[1]. Dysembryoplastic neuroepithelial tumors (DNETs) represent approximately 1% of all neuroepithelial brain tumors in patients younger than 20 years of age. It is a unique, surgically curable low-grade brain tumor which is included in the latest WHO classification as neuronal and mixed neuronal-glial tumor[2]. Two-thirds of DNETs are located in the temporal lobes, and 5% to 15% of temporal lobe resections for intractable epilepsy show DNETs[3].

Patients present with a long history of complex partial seizures, which are often drug resistant. The average age of the onset of seizures is 9 years. Contrast-enhanced cranial MRI shows absence of edema and only minimal if any enhancement. Pathological findings include oligodendroglia like cells and neurons in a mucinous eosinophilic background that give the appearance of floating neurons. The differential diagnosis includes oligodendroglioma, mixed oligoastrocytoma, and ganglioglioma. DNETs have a benign course, and gross total resection is often curative[2]. Adjuvant chemotherapy and radiation therapy are not recommended. The oncogenic BRAFV600E mutation can be found in 30% of DNETs.[2]

The prevalence of inter-ictal psychiatric disorders appears to be high in epileptic patients with a temporal lobe DNET primarily in relation to personality and behavioral problems with some degree of impulsivity and verbal aggressiveness. The improvements after surgery suggest that severe psychiatric disorders do not contra-indicate this procedure[4].

The basal ganglia play an important role in psychomotor behavior. Dysfunction of basal ganglia has been implicated in psychosis, obsessive compulsive disorder and depression. They also have a role in reward process.[5] Improvement in the neuropsychiatric disturbances was observed in our case as the temporal lobe tumor compressing the basal ganglia was excised.

## IV. Conclusion

DNETs are a group of rare and unique tumors primarily seen in younger population. Although the tumors may be located in any part of the brain, there is a proclivity for the superficial (just subcortical or intracortical) lateral or medial temporal lobe. Patients with these tumors typically present with a long history of complex partial seizures, which are often drug resistant. In the present case it is the behavioral and psychiatric manifestations that were predominant in the clinical picture. Most of the behavioral and personality disturbances have so far been attributed to the dysfunction of the frontal lobe. In this patient it was the involvement of basal ganglia responsible for the altered behavior and personality. Treatment is surgical resection and no adjuvant chemotherapy or radiotherapy is required. Usually carries excellent prognosis.

Financial support- None
Conflict of Interest- None

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