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Case Report

Non-pharmacological Management in Resistant Epilepsy with Intracranial Epidermoid Cyst: Case report

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ABSTRACT

Epilepsy is known to present with numerous neuro-psychiatric morbidities, one of which is intracranial Epidermoid Cyst. Lack of adequate knowledge about illness and treatment modalities is seen to be a cause of secondary emotional problems and poor medication adherence in the patients, causing further morbidity and poor prognostic outcome. This case reports and discusses the importance of timely detection of underlying organicity and optimal pharmacological management in a case of resistant epilepsy. It also aims to highlight the crucial role of non-pharmacological (psycho-educational) programs in maximising treatment outcome in these patients.

KEYWORDS: Epidermoid cyst, Resistant Epilepsy, Psychoeducation

1. INTRODUCTION

Seizure disorder (epilepsy) frequently presents with other neurological and psychiatric comorbidities, which can cause significant disability to patients and hamper their quality of life[1]. This includes the occurrence of intracranial tumors, of which Epidermoid tumors represent about 0.3 to 1.8% cases. Although epilepsy secondary to Epidermoid Cyst (EC) is relatively rare, it could be assigned to mechanisms of brain tissue infiltration, chemical meningitis or architectural change in epileptogenic areas[2]. EC commonly presents with symptoms of headache, seizures, raised intracranial pressure, cranial nerve deficits, focal cerebral deficits or cerebellar signs[3,4].

However, patients of epilepsy often lack intricate knowledge of their illness and available treatment modalities. As a result, they suffer from emotional distress as well as restriction in daily living and social interaction. Hence, psycho-educational programs curated with intent to improve knowledge and coping with epilepsy, and compliance to medication are being increasingly encouraged[5]. This case is presented to report the radiological evidence of an intracranial EC found while assessing a case of resistant epilepsy, and to highlight the role of psycho-education in long-term management.

2. CASE DETAILS

2.1 History and Examination

A right-handed female aged 13 years and weighing 33 kgs. was brought to Psychiatry OPD by her parents with complaints of episodic loss of consciousness associated with generalized jerky bodily movements lasting for about 30 – 40 seconds, up-rolling of eyeballs, frothing from mouth and followed by post-episode confusion and physical

weakness lasting about an hour. These episodes started 4 years back and have been more frequent and of longer duration over the years; also present during sleep. On enquiry, she also mentioned feeling fearful when alone, and asking her mother to accompany her during self-care routine. There was history of sleep initiation difficulties and occasional crying spells. The patient is first born of 3 siblings, born of a non-consanguineous marriage. There is no contributory family history.

Developmental history revealed history of delayed cry at birth during normal vaginal delivery at a hospital, with birth weight of 2.5 kgs and a history of 5 days NICU admission on day 3 of birth in view of postnatal jaundice. There was global delay in attaining milestones. She was enrolled in school at the age of 5 years, but had poor understanding of concepts and inability to perform satisfactorily on academic tasks. She dropped out in 2nd standard at the age 9 years after the onset of seizure episodes.

There were no additional behavioral complaints and she gradually learned activities of daily living and self-care with the help of her mother. Menarche was attained 6 months back (age 13 years) and so far, had been irregular. Her parents described her as a child with shy temperament, obedient and cheerful, with occasional irritability and anger spells on provocation.

On general examination she appeared pale, but her vitals were within normal limits. She had kyphoscoliosis of the spinal column with mild atrophy of left side of body, and a limp in gait. No obvious abnormality was noted on facial features to suggest a congenital anomaly or Syndrome, except for short frenum of tongue. An incidental finding of lump in left breast with skin excoriation and retraction of nipple was seen, with no discharge. CNS and other systemic examinations revealed no significant abnormality. On mental status examination, she appeared scared and shy, and was slow to warm up. There were no remarkable findings other than lack of clarity in pronunciations, increased reaction time to verbal responses and anxious affect.

2.2 Investigations

A battery of investigations including complete blood count, ESR, liver, thyroid and renal function tests, blood sugar and lipid profile was conducted and found to be within normal limits. Neuro-imaging (MRI brain) done to screen for underlying organicity revealed gliosis in bilateral parietal lobes, causing ex-vacuo dilatation of left lateral ventricle, like representing post-ischemic sequelae. An extra-axial hypointense (T1) cystic lesion measuring 27 x 13 x 12 mm (hyperintense on T2 and DWI) was seen involving right cerebello-pontine angle and prepontine cistern, causing indentations over right middle cerebellar peduncles, pons and lateral displacement of cisternal segment of right trigeminal nerve. These findings were suggested to be likely representing an Epidermoid Cyst [Fig 1-6]. Electro-encephalogram (EEG) showed generalized sharp waves and spikes suggestive of generalized seizure disorder [Fig 7]. The patient had earlier been evaluated for Intelligence Quotient and had documented an IQ of 58 suggesting Mild Intellectual Disability.

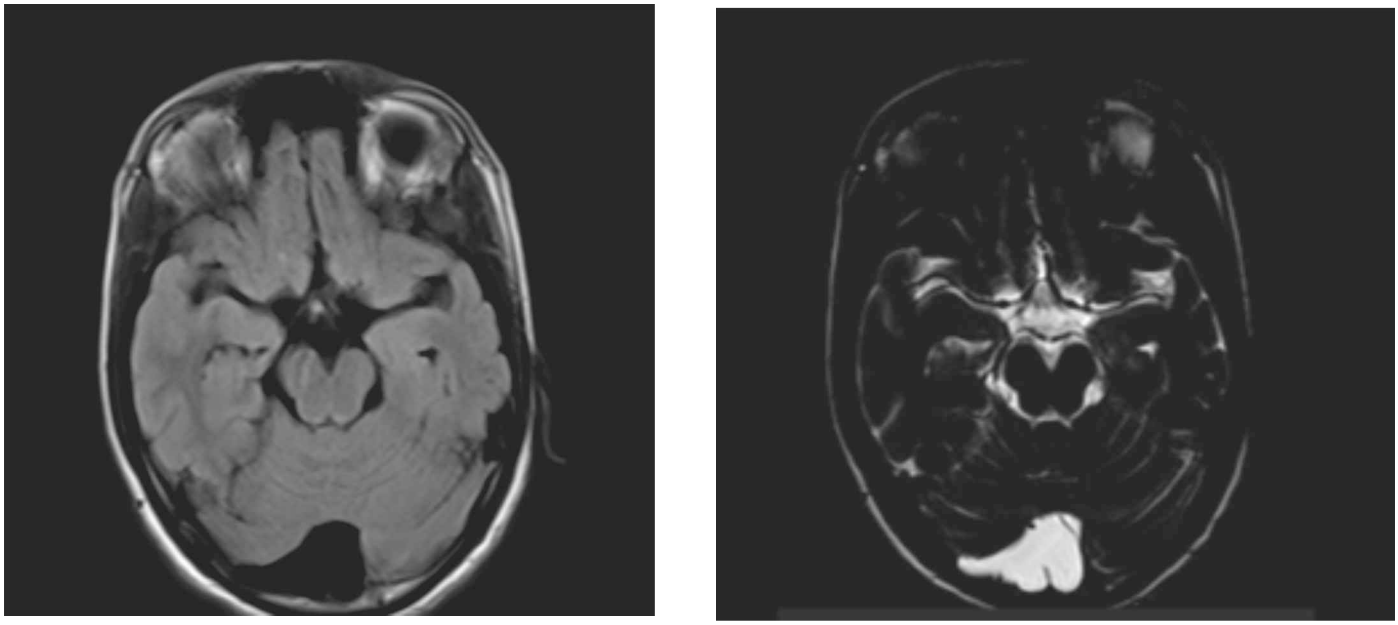


Figure 1,2: Axial section of brain (hyperintense on T2) showing a cystic lesion in posterior end of right hemisphere

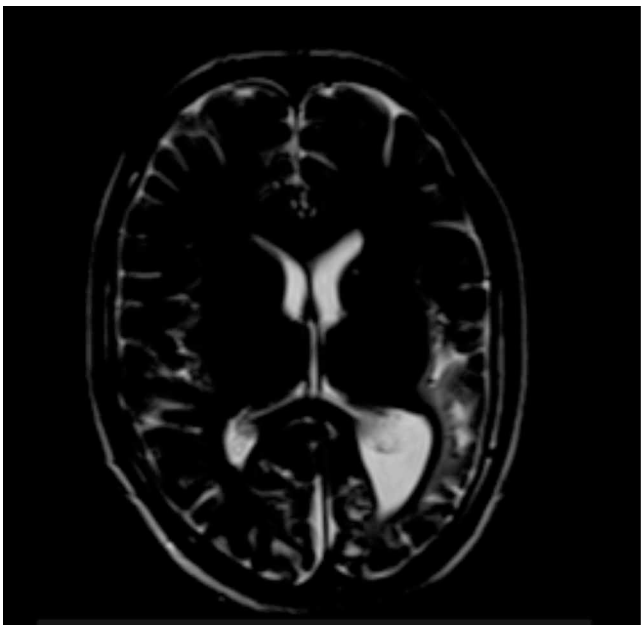


Figure 3: Axial section of brain showing gliosis in bilateral parietal lobes and dilated left lateral ventricle (T2)

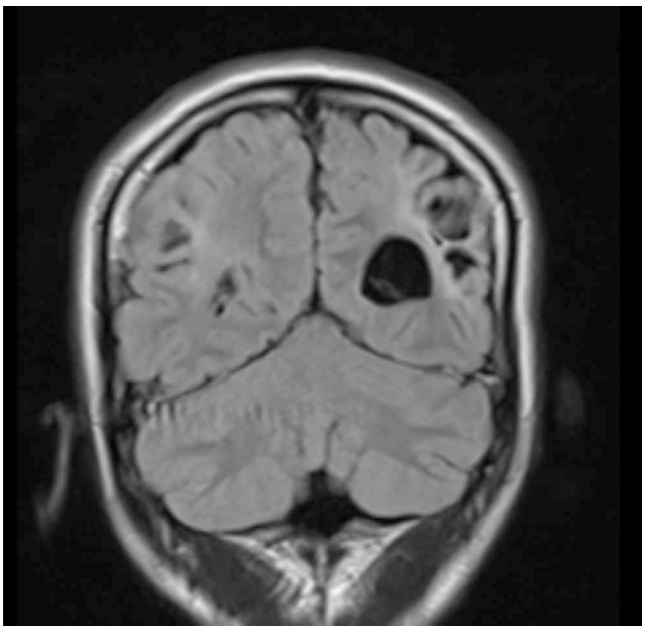
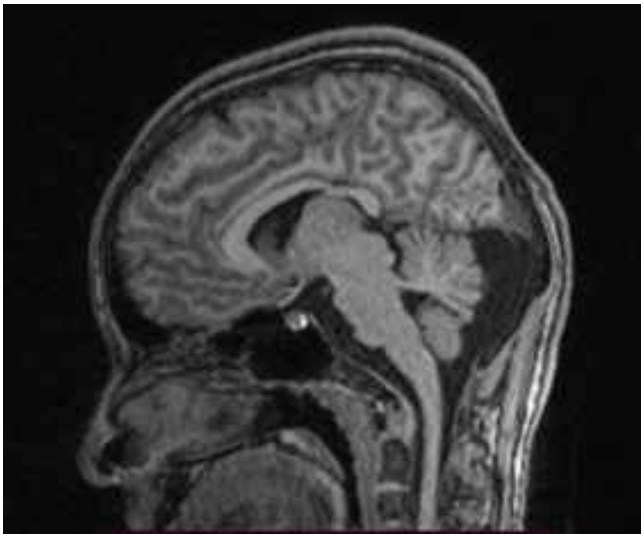


Figure 4: Coronal section of brain showing gliosis of parietal lobes and dilated left lateral ventricle (T1)



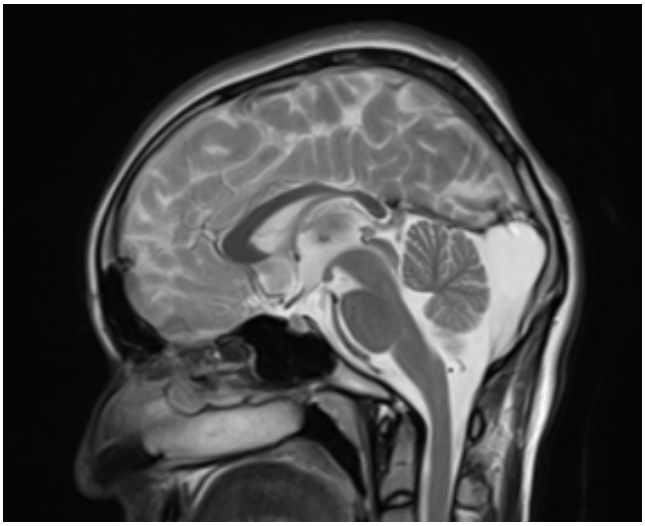


Figure 5,6: Sagittal section of brain showing cystic lesion in cerebello-pontine angle (hyperintense on T2) causing indentation over underlying structures

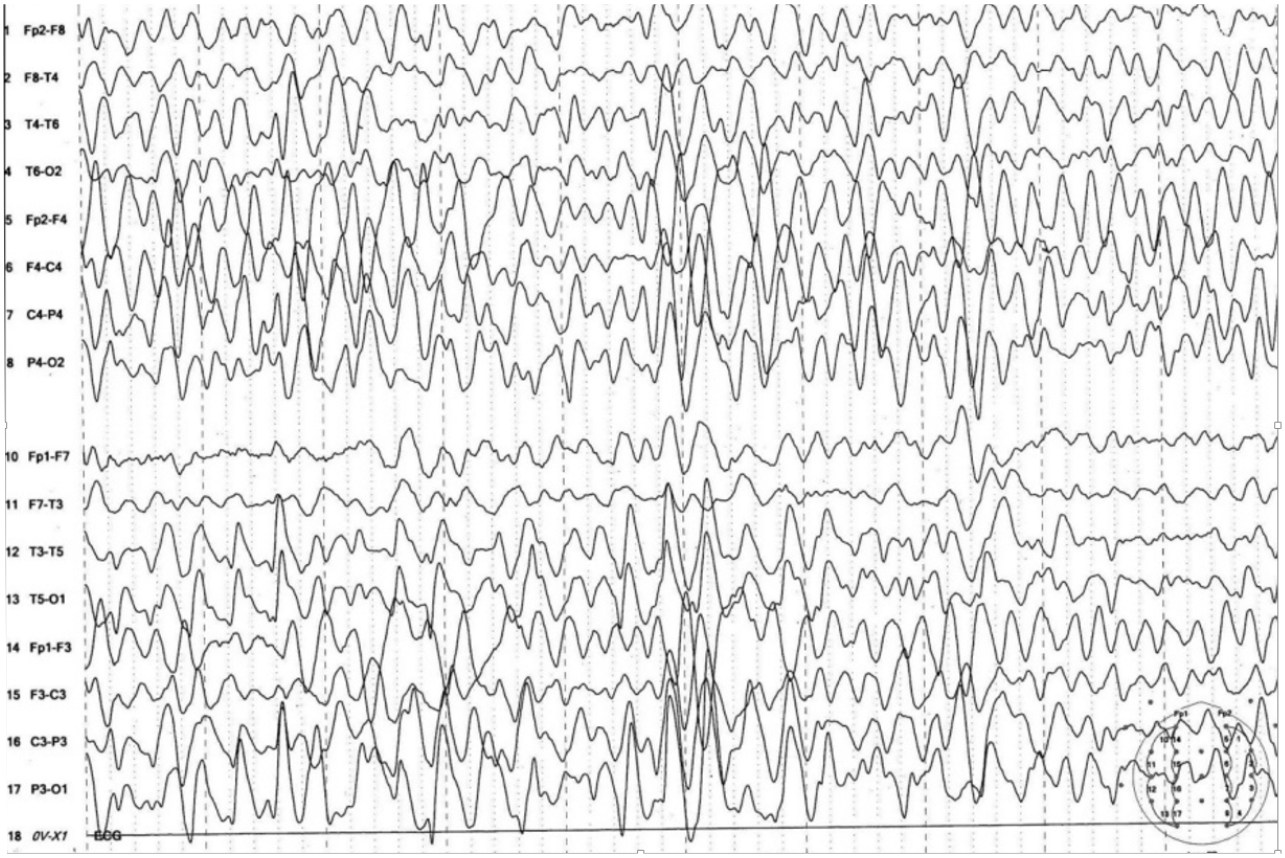


Figure 7: Awake EEG record showing generalized epileptiform activity in the form of spikes, sharp and slow waves

2.3 Management

Based on the clinical history and examination, and as per findings of investigation reports, the patient was diagnosed as Intellectual Disability Disorder with Seizure Disorder, secondary to underlying organicity (intracranial Epidermoid cyst). Her neurotic symptoms were concluded to be reactive to physical illness and its resultant emotional and social repercussions over time. The patient had so far received trials of Levetiracetam (up to 1000 mg/day), and another trial of Phenobarbitone (60 mg/day) in combination with Carbamazepine (up to 600 mg/day) in the past, both of which she had tolerated well, but had shown partial response to.

Considering inadequate response to previous treatment regimen, it was decided to admit the patient for close monitoring and comprehensive management. A trial of Phenytoin (200 mg/day) and Phenobarbitone(120 mg/day) in divided doses was initiated and Carbamazepine was slowly cross-tapered with Valproate (400 mg/day). For one off seizure episode during this cross-over, she was administered Clobazam 10 mg on SOS basis. With gentle rapport-building and after appropriate psycho-education and supportive counselling of both parents, it was observed that her anxiety and withdrawn behavior slowly receded and the patient seemed comfortable during inpatient stay. Over the first week, two episodes of seizures were observed, each lasting less than 3 seconds and with less than 2 minutes of post-ictal confusion. This was a relatively dramatic control over seizure intensity which had not been previously attained.

An opinion from Gynecology team was sought for the lump in breast and treatment initiated in view of an inflammatory lesion. Neurosurgical consultation was done with reference to management strategy for intracranial EC and it was opined that in view of satisfactory improvement in symptoms, surgical resection was not imminent at present. Observation for symptom worsening was suggested and a review was planned over next six months.

On her recent follow up visit no seizure episode has been reported for the past one month. Her mother also reported an improvement in her overall functioning, including emotional responsiveness and cognitive functioning as observed through her active participation in household chores and social interactions.

3. DISCUSSION

The term 'resistant' also known as refractory or intractable epilepsy, appears to be self-explanatory, but its precise definition remains elusive. As per consensus fromThe International League Against Epilepsy (ILAE), drug resistant epilepsy is defined as failure of adequate trials of two tolerated, appropriately chosen and used antiepileptic drug schedules (as monotherapies or in combination) to achieve sustained freedom from seizure episodes. Although lifelong seizure freedom without adverse effects would be the most clinically relevant outcome of any intervention for epilepsy, some breakthrough seizures could be provoked by sleep deprivation, menstruation, fever or other external factors[6,7]. This case was labelled as resistant epilepsy based on the above criterion.

Psychosocial problems in treating epilepsy are seen to be been a major unmet need. A review article stated that specific interventions for these problems can improve quality of life, social adjustment, and adjustment to seizures in these patients, with outstanding results for refractory cases. Integration of psychosocial management into the mainstream treatment of specialty clinics like Neurology has also been proposed[8]. In line with these recommendations, a study testing efficacy of structured educational programs on drug adherence in epilepsy reported significant improvement in medication adherence ($p = 0.001$), as measured by Morisky Medication Adherence Scale (MMAS)[9].

In another controlled study, researchers conducted analyses with a questionnaire specifically based on epilepsy, and found significant improvement on measures of knowledge ($p<0.001$), coping and adaptation ($p<0.01$), illness related anxiety ($p<0.05$), and seizure management ($p<0.05$), when implementing the educational program called FAMOSES (modular service package epilepsy for families). It was also found to be helpful in reducing seizure frequency in children ($p < 0.05$)[10]. Advantages of psycho-education in epilepsy are emphasized as lack of perceived stigma in comparison to forceful participation in psychotherapy, and an opportunity for providing basic medical information, triggering mechanisms in epilepsy and various coping skills. Delivery of this information in regional dialects and promoting self-management skills are also highlighted as therapeutic principles[11]. Thus, misconceptions about epilepsy can be reduced by imparting correct information to patients and their families, in turn improving course of illness, optimizing long-term management and preventing complications[5].

Psychosocial outcomes in children and adolescents with epilepsy are particularly compromised due to perceived stigma, behavioural symptoms, academic difficulties and comorbid depression. In such cases, psycho-educational interventions including positive peer support are highly encouraged[12].

This case highlights the much-needed role of relevant, adequate and timely psycho-education of patient and care givers in minimizing emotional health consequences in neuro-psychiatry. Given the adverse long-term implications of such diagnoses on the patient's social and personal life, it is vital to share difficult information sensitively and empathically to the family. Also, effective management of primary presenting problem such as refractory seizures in this case, is crucial for alleviating their immediate stress response.

It is the second case in a series of EC cases being assembled in view of presentation with psychiatric symptoms and psychosocial aspects of management in addition to pharmacotherapy and / or surgical intervention. One such case reported by our team elaborated on psychosis as a manifestation of EC with resistant epilepsy[13].

4. CONCLUSION

Sensitive and appropriate psycho-education is a critical aspect of symptom management in neuropsychiatry. An index of suspicion for underlying organicity is equally important in cases presenting with difficult to treat symptoms like resistant epilepsy.

5. CONFLICTS OF INTEREST: None

6. FINANCIAL SUPPORT: None

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