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CASE REPORT - AN INTERESTING CASE OF ATAXIA

Neurology

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ABSTRACT

Steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT)/Hashimoto's encephalopathy (HE)is a rare clinical entity seen in paediatric population. It has a wide clinical spectrum ranging from myoclonus, seizures, behavioural changes, cognitive change, pyramidal tract dysfunction, involuntary movements, and cerebellar dysfunction to psychosis and coma, with relapsing and/ or progressive course.1
SREAT with acute ataxia as presentation is very rare in paediatric age group. We hereby report a case of an acute ataxia in a 12 year old girl, diagnosed as SREAT after detailed workup.

KEYWORDS

INTRODUCTION

Steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT) is a rare clinical entity seen in paediatric population.

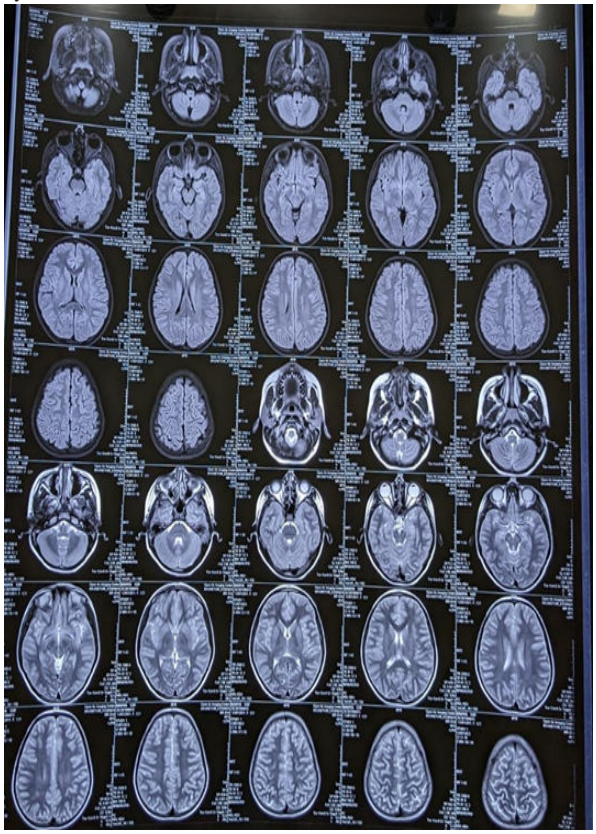
It has a wide clinical spectrum ranging from myoclonus, seizures, behavioural changes, cognitive change, pyramidal tract dysfunction, involuntary movements, and cerebellar dysfunction to psychosis and coma, with relapsing and/ or progressive course.[1]

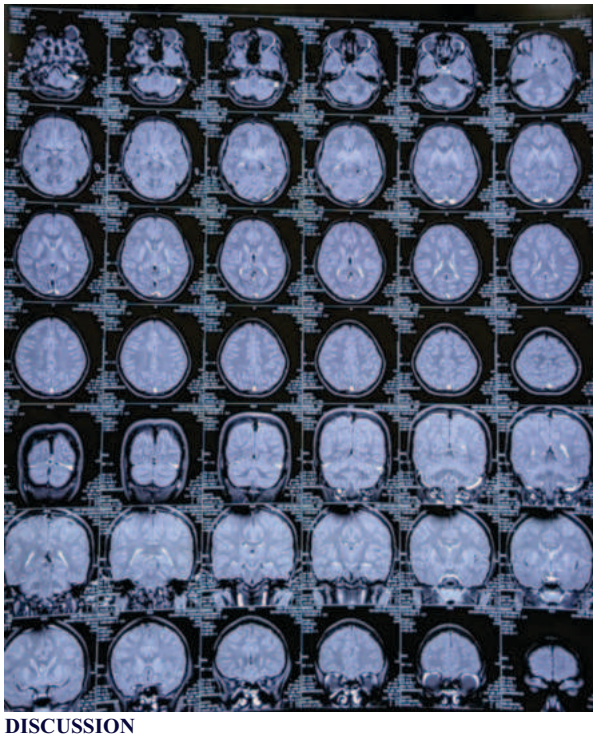
The estimated prevalence in adults is 2.1/100,000, the prevalence in children is unknown.[2]

In a study done by R. Boelen and T. de Vries , epilepsy and behavioural cognitive problems were the most common clinical presentations. Ataxia as a presentation was uncommon.[3]

CASE REPORT

A 12 year old child was admitted in our hospital on 28/07/2023 with history of acute onset of unsteadiness with giddiness and vomiting since 22/06/2023, with worsening of symptoms since 1 week .On examination she had gait ataxia,stance ataxia, appendicular ataxia, scanning speech and gaze evoked nystagmus. Her birth and development history was normal. There was no history of febrile illness, drug exposure or toxin exposure prior to the onset of illness. About a week after onset of symptoms, she was admitted and evaluated in a private hospital. MRI brain with contrast showed T2 hyperintense area which showed blooming in left occipital lobe with no surrounding edema or contrast enhancement - possibility of cavernoma/chronic bleed.Blood investigations including complete blood count, renal and hepatic parameters, peripheral smear were within normal limits. HIV and hepatitis B virus, hepatitis C virus serology were negative. CSF analysis was normal. Nerve conduction study was normal. She was given supportive treatment for the same and discharged on 17/07/2023. The course of the disease was static with no improvement/worsening since the onset of the disease. She was admitted in our hospital with worsening of symptoms since 21/07/2023. Serum T3,T4 levels were normal. Serum TSH was high with a value of 100 mIU/L. Serum Anti-TPO titre was high(27.57 IU/ml). ANA profile was negative. A diagnosis of SREAT /HE with ataxia was made and she was started on prednisolone (1mg/kg body weight)and was found to have significant symptomatic improvement. She was discharged on a maintenance dose of oral steroids. On discharge, she had no stance ataxia, gait ataxia, appendicular ataxia or dysarthria.





DISCUSSION

Steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT) is a rare clinical entity seen in paediatric population. It has a wide clinical spectrum ranging from myoclonus, seizures, behavioural changes, cognitive change, pyramidal tract dysfunction, involuntary movements, and cerebellar dysfunction to psychosis and coma, with relapsing and/ or progressive course.[1]The estimated prevalence in adults is 2.1/100,000 , the prevalence in children is unknown.[2]

In a study done by R. Boelen and T. de Vries, epilepsy and behavioural cognitive problems were the most common clinical presentation. Ataxia as a presentation was uncommon (seen only in 11 out of 100 cases studied).[3]

In another study done by Bayram et al in Turkey, only 2 out of 15 patients had ataxia.[4]

Hashimoto's encephalopathy (HE)/ SREAT is an autoimmune disease related to thyroid antibodies and was first described by Brain et al. in 1966. It is believed that the immune inflammatory reaction during the pathogenic process may lead to focal or diffuse brain damage in the brain, leading to clinical symptoms such as cognitive impairment, tremor, altered consciousness, transient aphasia, seizures, myoclonus, gait disorder/ataxia.

Till date, clinical reports of cerebellar ataxia as the main symptom of HE are rare. Disease is commonly seen in fifth to sixth decade of life, with female population predominantly affected.[5]

Pathogenesis is not clear; however immune mediated cerebral vasculitis, toxic effects of thyroid-stimulating hormone on the central nervous system, global hypoperfusion, or the role of an antigen common to the brain and thyroid (e.g., the antigen a-enolase) are the possible mechanisms. However no correlation was found between severity of disease and antibody titre. Patients show good response to steroid and slow tapering of steroid is suggested. If there is a relapse during tapering or insufficient response, long term immunosuppressive drugs like azathioprine, cyclophosphamide, methotrexate is used. If the patient also has hypothyroidism, thyroxine supplementation has to be given[1]

CONCLUSION

SREAT/HE is rarely seen in paediatric population and presentation with isolated acute ataxia is very rare. It is very important to recognise SREAT in patient presenting with acute or subacute ataxia/ behavioral or cognitive disturbances or seizures, since it is a treatable condition and shows very good response to steroids.

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