Chorea: An Atypical Manifestation of Coeliac Disease

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ABSTRACT

Chorea is a rare neurological manifestation of coeliac disease. A 35-year-old male presented with a nine month history of chorea, starting from the right little finger and spreading over the next few months to involve the right upper and lower limbs. History and physical examination were not suggestive of an underlying cause. CT scan and MRI scan did not reveal any structural abnormality. Levels of serum IgA anti-tissue transglutaminase antibodies were very very high (396EU/ml). Duodenoscopy and duodenal histology were normal. Marked symptomatic improvement was seen with gluten free diet, along with disappearance of anti-tissue transglutaminase antibodies. Chorea can occur as an atypical manifestation of coeliac disease. This possibility must be kept in mind when the cause of chorea is not easily identifiable.

Keywords: Chorea; Gluten; Coeliac disease

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INTRODUCTION

Chorea refers to brief abnormal involuntarily non-rhythmic writhing movements of the limbs. The underlying etiology is quite broad and a large number of conditions can cause chorea. Some of these are common while others are seen rarely. The search for the cause in a given patient can at times be challenging even for the astute physician. No underlying cause can be found in a significant proportion of individuals [1]. Coeliac disease is one of the rare causes of chorea. This case report describes one such case.

CASE REPORT

A 35-year-old male presented to us with abnormal involuntary movements involving the right little finger, starting around nine months ago. These movements progressively involved the right hand followed by the right upper limb and then the right lower limb during the following three to four months. There was no history of any other neurological symptom or gastrointestinal complaint. Drug and family history were not contributory. On examination, the patient had chorea involving the right half of his body. Kayser- Fleischer rings were not seen on slit lamp examination. There was no ataxia or cerebellar signs and the rest of the systemic examination was unremarkable. The patient was further evaluated in detail by a neurologist at a tertiary care center.

Baseline hematological investigations that included complete blood count, platelets, liver function test, serum urea, creatinine and fasting serum lipid profile were normal. Peripheral blood smear did not reveal any acanthocytes. No abnormality was detected on CT scan brain and MRI brain. Further investigations revealed significantly high levels of IgA anti-tissue transglutaminase (tTG) antibodies (396EU/ml). Antigliadin antibodies were negative. Upper gastrointestinal endoscopy was normal as was duodenal mucosa on histopathological examination. There was no evidence of an increase in intraepithelial lymphocytes, villous atrophy or crypt hyperplasia. Serum ceruloplasmin, copper and creatinine kinase were also within normal limits. Syphilis serology (serum Venereal Disease Research Laboratory and Treponema Pallidum Haemagglutination Assay) was negative. Cerebrospinal fluid routine exam, electromyogram/ nerve conduction studies and echocardiogram did not show any abnormality. The patient was started on a gluten free diet, with which he clinically improved remarkably. Serum IgA anti tTG antibodies were retested after three months and were not detectable. The patient was advised to continue with the dietary restrictions and follow up regularly.

DISCUSSION

Coeliac disease in adults can have a number of

neurological manifestations; common ones being peripheral neuropathy and cerebellar ataxia, seen in around 10% of patients [2]. Chorea is a rare association, with only a few cases described in literature [3]. All of these patients had clinical evidence of typical gastrointestinal (GI) involvement due to coeliac disease. This patient is different from others since he did not have any evidence of the disease in his GI tract. It is well known that these extra-GI complications may precede the diagnosis of coeliac disease. Despite extensive workup, no other cause of chorea was identified. This, coupled with clinical improvement and disappearance of serum anti tTG antibodies on a gluten free diet, proves that chorea was due to gluten intolerance and not idiopathic.

Anti tTG antibodies were used for the diagnosis in this patient because they have the highest sensitivity, specificity as well as positive and negative predictive values out of all the antibodies that may be positive in coeliac disease. Chorea limited to one side of the body should generally raise suspicion of a structural brain lesion, but fortunately, neuroimaging was normal in this patient. Since he had progressive disease, it is possible that chorea would have progressed to involve the left half of his body.

The possible mechanism for neurological manifestations including chorea in coeliac disease is still a matter of debate. It is thought to have an immunological basis, with antigliadin and other antibodies cross reacting with unknown neuronal targets sharing the same epitopes as gliadin proteins. Role of nutritional deficiencies especially of mineral and vitamins has also been postulated but this does not seem to have a strong basis since correction does not improve the clinical picture [4]. Similarly, the response of neurological manifestations to a gluten free diet is controversial with variable results obtained in different clinical trials [5]. More evidence is needed for establishing the efficacy of gluten free diet, but the experience with this patient highlights the utility of gluten-free diet.

CONCLUSION

Chorea is a known but rare manifestation of coeliac disease and can occur even in the absence of gut involvement. Coeliac disease should always be kept in the differential diagnosis of chorea, especially in cases where the etiology is not obvious.

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