



# Myorhythmia unmasks Whipple's disease– A rare case

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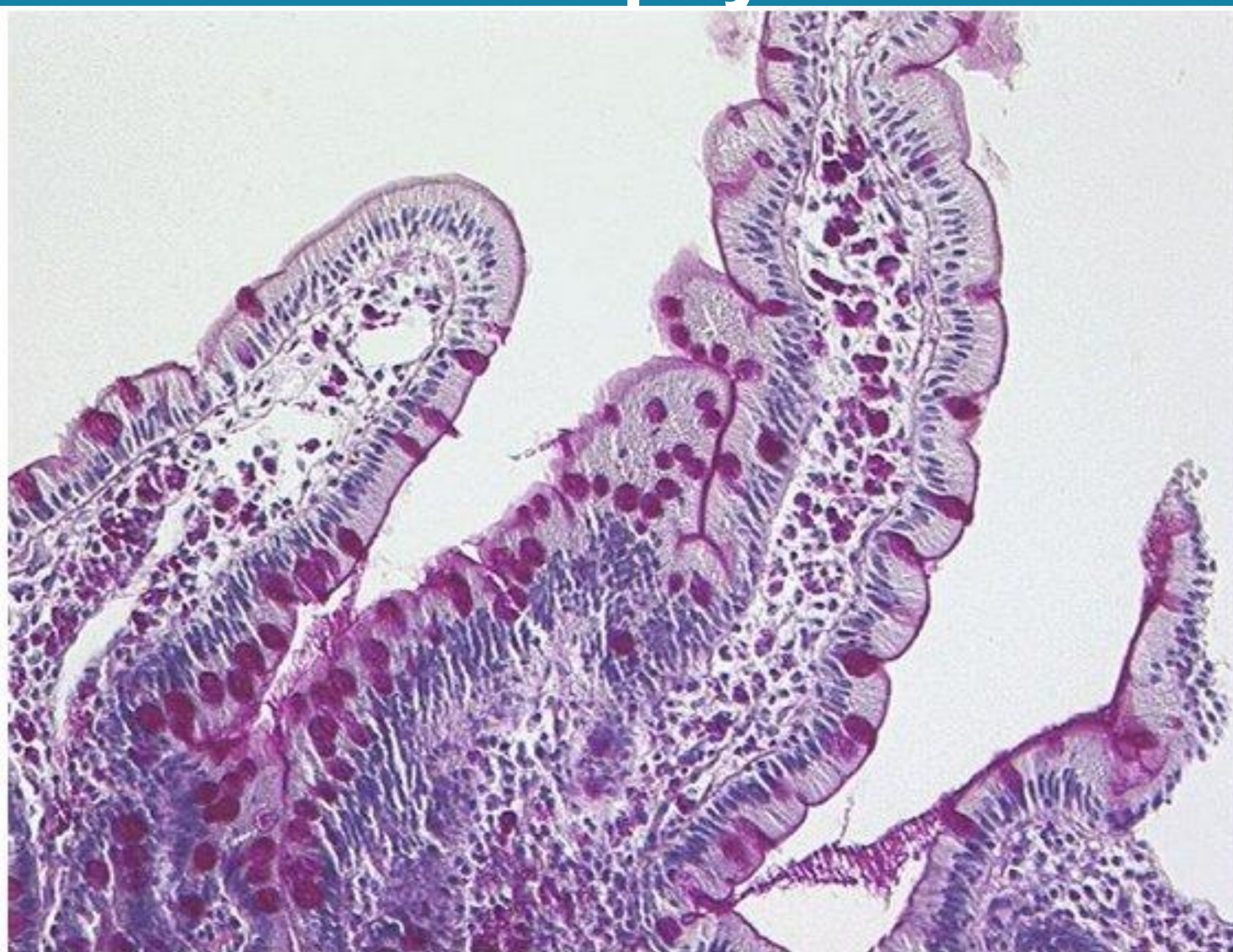
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## Introduction

- ❖ Whipple's disease: Rare systemic illness
- ❖ Bacterium *Tropheryma whippelii*- a gram positive bacillus related to Actinomycetes
- ❖ Symptoms often include gastrointestinal disturbances, weight loss, joint pain.
- ❖ Neurological manifestations such as oculo masticatory myorhythmia, a condition characterized by rhythmic eye and jaw movements.
- ❖ Recognizing oculo masticatory myorhythmia (OMM) is crucial in diagnosing central nervous system manifestations of Whipple's disease, as it serves as a pathognomonic indicator.

## Biopsy



- He was started on IV Ceftriaxone 2gms OD for 2 weeks, followed by oral antibiotic Cotrimoxazole DS BD for 1 year. Patient is under regular follow up.

## Case details

### History:

Involuntary movements Oro-facial region 6 months.  
No h/o limb weakness/ cranial nerve dysfunction/ incoordination/seizures.  
Abdominal pain with loose stools, loss of appetite 1 year

### Examination:

Rhythmic myoclonus or spasm occurring in synchronous bursts involving several adjacent regions mainly jaw and face

75 years male

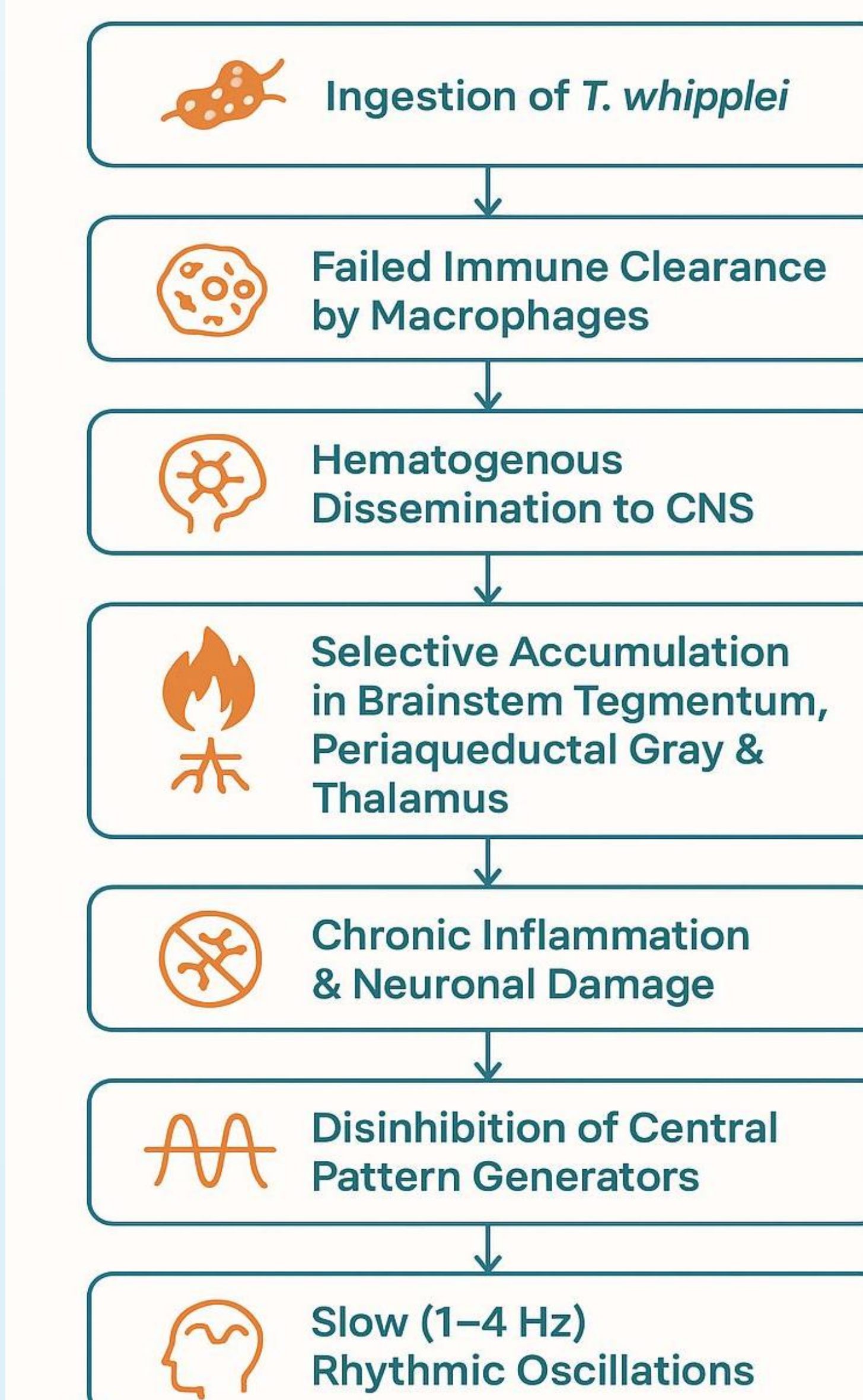
### Past history:

Coronary artery disease S/P CAG on anti-platelets and statins.  
Type 2 diabetes mellitus, systemic hypertension and chronic kidney disease, on regular medical management.

### Investigations:

Upper gastrointestinal endoscopy with D2 biopsy was done.  
Biopsy with PAS staining showed chronic duodenitis with features suggestive of Whipple's disease.

## MECHANISM OF MYORHYTHMIA IN CNS WHIPPLE'S DISEASE



## TREATMENT FOR CNS INVOLVEMENT

### INITIAL PHASE

- Inj Ceftriaxone 2gm IV once daily for 2-4 weeks
- Inj Penicillin G 4 million units IV every 4 hours
- or Inj Meropenam 1gm IV every 8 hours for 2-4 weeks

### MAINTENANCE PHASE

- TMP-SMX (one double-strength tablet (160 mg TMP/800 mg SMX)) twice a day) for one year

## Discussion

- ❑ Isolated involvement- central nervous system and heart valves, can occur in the absence of the classic findings of whipple's disease.
- ❑ Neurologic findings 10 to 40 percent of patients with classic Whipple's disease. Isolated CNS infection rarely occur. The incidence of CNS disease increases over time in an infected patient
- ❑ Most commonly, CNS involvement is asymptomatic and only diagnosed by PCR detection of *T. whippelii* in the CSF. Among patients with symptomatic CNS involvement, cognitive dysfunction, including dementia, other memory impairment and confusion, is the most common abnormality.
- ❑ Testing of small bowel biopsy: Upper GI endoscopy with biopsies of the small intestine is the diagnostic test of choice.
- ❑ CNS involvement is very difficult to manage. For those who have severe CNS symptoms or brain lesions, adjunctive corticosteroids may be beneficial.

## Conclusion

- The presence of myorhythmia necessitates aggressive and prolonged antibiotic therapy capable of crossing the blood-brain barrier (e.g., ceftriaxone followed by long-term trimethoprim-sulfamethoxazole). Delaying treatment can lead to further, irreversible neurological decline.

- Bally JF, Méneret A, Roze E, et al. Systematic review of movement disorders and oculomotor abnormalities in Whipple's disease. *Mov Disord* 2018; 33:1700.
- Louis ED, Lynch T, Kaufmann P, et al. Diagnostic guidelines in central nervous system Whipple's disease. *Ann Neurol* 1996; 40:561.