Atypical Whipple’s disease

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We report an unusual case of Whipple’s disease diagnosed with help from the patient himself. The patient presented with rash resembling dermatitis herpetiformis, constipation, and intermittent diarrhea. A suspicion of celiac disease led to duodenal biopsy, which showed features of Whipple’s disease on histology and electron microscopy. [Indian J Gastroenterol 2005;24:31]

Initially considered to be an intestinal disorder, Whipple’s disease is a multisystem disease. Virtually any organ may be infected by *Tropheryma whippelii*. The disorder has been described most frequently in rural regions and Caucasian farmers of mid Europe.2 A few cases have been reported in Asian populations.

A 54-year-old man complained of itchy, erythematous, vesicular rash, symmetrically distributed over the extensor surfaces, for nine months. He was clinically diagnosed as having dermatitis herpetiformis. However, histological examination and immunofluorescence of the skin biopsy proved negative. In the meantime, the patient searched the internet and found an association of dermatitis herpetiformis with celiac disease. He had a history of 3-5 episodes of foul-smelling diarrhea every year, each lasting for less than 7 days, for the last 25 years. These were followed by constipation, difficult defecation and a sense of incomplete evacuation. One episode lasted for approximately 2 months in 1990. In 1998 he lost 5 Kg weight with change in bowel habits and occasional diarrhea. The patient was diagnosed two months ago to have pulmonary tuberculosis on the basis of reticular opacities in the right upper zone on X-ray and CECT, and was being treated for the same.

Associating his intestinal symptoms with celiac disease, he reported back for gastroenterology consultation. On examination, other than the rash, systemic examination was noncontributory. Upper GI endoscopy and colonoscopy were normal. MRI of the brain showed focal ischemic gliosis of the cortex and subcortical white matter of the posterior parietal lobe. Duodenal biopsy showed widened villi with dilated lacteals with normal crypt-to-villus ratio. The lamina propria was expanded by numerous histiocytes with granular cytoplasm. There was PAS-positive material (rods, granules and sicks) in the histiocytes. Ziehl-Nielsen and mucicarmine stains were negative. Electron microscopy revealed rod-shaped bacilli morphologically consistent with bacilli of Whipple’s disease (Fig). The patient was put on co-trimoxazole 480 mg twice daily; 6 months later his diarrhea has resolved and constipation has improved.

Weight loss and chronic diarrhea are amongst the commonest symptoms of Whipple’s disease, seen in two-thirds of patients.1 Arthritis too is very common. Gastrointestinal symptoms, which generally begin later and ultimately lead to the diagnosis, consist of episodic and watery diarrhea or steatorrhea, in many cases accompanied by colicky abdominal pain and, in 20%-30% of patients, by occult blood in the stool.1,2

On endoscopy, Whipple’s disease is associated with pale yellow shaggy mucosa alternating with erythematous, erosive, or mildly friable mucosa in the post bulbar region of the duodenum or in the jejunum; alternatively, whitish-yellow plaques can be seen in a patchy distribution.3 Our patient did not have any endoscopic findings possibly due to suppression by preceding antituberculous treatment inclusive of rifampicin, which has bactericidal effect on *Tropheryma whippelii*.

Histology shows granular foamy macrophages stained purple with PAS; in addition, diastase-resistant and silver-positive inclusions representing more or less intact remnants of ingested bacteria might be visible.4 Skin hyperpigmentation, seen in 40%-60% of cases, particularly of light-exposed areas, is not unusual and is often erroneously diagnosed as Addison’s disease.2,5 Skin lesions resembling dermatitis herpetiformis have not been described, and make this case unusual. Lymphadenopathy, which is seen in 40%-60% of cases, was conspicuous by its absence. A neuroradiological lesion was evident but was clinically silent.

References


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