A 39-year-old man was admitted with confusion, hypersomnia, loss of memory and a 3-year history of weakness, recurrent arthralgia, fever and weight loss. Physical examination showed peripheral adenopathy, facial myoclonus and external ophthalmoplegia. Cerebral magnetic resonance imaging demonstrated diffusely increased thickness of the meninges. The cerebrospinal fluid was clear with increased protein concentration and contained few lymphocytes and monocytes. Some monocytes stained strongly with diastase-PAS reaction, with coarse bodies (A) or bacillary structures (B) in the cytoplasm. Peripheral blood monocytes (C) and bone marrow macrophages also contained PAS-positive diastase-resistant material. These findings were suggestive of Whipple’s disease, even in the absence of gastrointestinal symptoms. PCR positivity for Tropheryma whippelii in peripheral blood and cerebrospinal fluid, as well as typical histological findings on duodenal biopsy, confirmed the diagnosis. The patient was treated with trimethoprim-sulphamethoxazole and ceftriaxone with significant improvement of his condition.

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Figure 1.