A 29-year-old Chinese woman complained of recurring dizziness for 3 months and gait disturbance for 2 months. On admission (April 2016), she was found to have a wide-based gait, horizontal nystagmus and positive heel-knee-shin test. Initial hematological, biochemical, microbiological, hormonal investigations and tumor serum markers were unremarkable. Oligoclonal bands were observed in cerebrospinal fluid (CSF) without other pathological changes, while magnetic resonance imaging (MRI) of cervical spine and brain were normal. An anterior mediastinal mass was noted by computed tomography (CT) [Figure 1A]. Despite a lack of paraneoplastic antibodies in CSF or serum, such as anti-neuronal nuclear autoantibody type 1 (ANNA-1, also known as “anti-Hu”), ANNA-2 (also known as “anti-Ri”), Purkinje cell antibody type 1 (PCA-1, also known as “anti-Yo”), anti-CV2 (also known as “collapsin response mediator protein 5 [CRMP5]”), anti-Ma, anti-Amphiphysin, anti-Tr (also known as “delta/notch-like epidermal growth factor-related receptor [DNER]”) and anti-glutamic acid decarboxylase (GAD), her neurological manifestation still pointed toward one of the paraneoplastic neurological syndromes (PNSs).

The anterior mediastinal mass was removed surgically and pathologically confirmed as extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma), characterized by lympho-epithelial lesions. The immunohistochemical analysis showed that the neoplastic cells were CD20 (+), BCL2 (+), CD3 (-), CD5 (-), CD10 (+), CD23 (+), Cyclin D1 (+), SOX-11 (+), BCL6 (-), CD117 (-) and demonstrated λ light chain restriction. Particularly, her rheumatological tests showed positivity for antinuclear antibody, antibodies to Sjögren’s syndrome-related antigen A and rheumatoid factor, along with mildly decreased C3 level. The diagnosis of primary Sjögren’s syndrome (pSS) was considered despite the lack of characteristic dryness. The patient had a painless left parotid mass removed in 2012 with a pathology report of “parotid adenoma with reactive lymphoid hyperplasia”. Given the seemingly unusual association between the previous parotid mass, current MALT lymphoma and possible pSS, previous paraffin sections of parotid mass tissue were used to recheck pathological phenotype of the mass. Significantly, the parotid mass was shown to be MALT lymphoma.

Her neurological symptoms had no changes after removal of the mediastinal mass. Subsequently, she received rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone (R-CHOP) chemotherapy. However, she progressively presented with dysarthria and dyskinesia, and became confined to a wheelchair. Ten months after diagnosis, dryness of eyes and mouth appeared. In April 2017, she underwent a new evaluation. The sicca symptoms, abnormal Schirmer’s test, and multifocal lymphocytic sialadenitis of the labial glands further verified the establishment of pSS. There was no evidence of hereditary ataxia. Repeat positron emission tomography-CT and detection of paraneoplastic antibodies were negative. Brain MRI exhibited cerebellar atrophy [Figure 1B]. After tear substitutes, hydroxychloroquine, and prednisone were given, dryness symptoms were improved. Mycophenolate mofetil and rehabilitation training were prescribed for later treatment. During the 2-year follow-up, her cerebellar ataxia stabilized, and she could stand up and slowly walk on her own, with cerebellar imaging showing no obvious changes.

The patient experienced MALT lymphomas arising in distinct sites, both antecedent to the diagnosis of pSS. MALT lymphoma is an indolent lymphoma, closely associated with chronic inflammation resulting from autoimmune disorders or infection. The therapeutic choice of MALT lymphoma is heterogeneous, mainly depending

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on involved organ and the extension of the disease. pSS is a systemic autoimmune disease, characterized by lymphocytic infiltration in exocrine glands and leading to impaired secretory function, with B cell hyperactivity vital in its pathogenesis. While 30-40% of pSS patients are affected with systemic involvement, the diverse extraglandular complications can be the primary manifestations, without sicca symptoms, contributing to the difficulty in accurate diagnosis. Patients with pSS have 15-fold higher risk of B cell lymphomas, which are predominantly MALT lymphomas, that often develop in the organs where pSS is active, such as salivary glands. Indeed, most salivary MALT lymphomas are associated with pSS. We speculate that her previous parotid MALT lymphoma is related to pSS. Though the parotid MALT lymphoma was pathologically misdiagnosed, the diagnostic excisional biopsy exerted a partially therapeutic role. Nevertheless, in regard to her mediastinal MALT lymphoma, we cannot distinguish between a distant relapse or the expansion of primary lymphoma lesion initially coexisting with the parotid lesion. Importantly, the progression of lymphoma in pSS can be heralded by high pSS disease activity. The process of autoimmunity likely started very early before the diagnosis of pSS. Probably due to the individual’s highly compensatory gland function or elevated perception threshold, lack of sicca symptoms concealed the underlying disorder. The “silent” autoimmunity and chronic B cell activation may account for the occurrence and progression of MALT lymphoma in the case.

Based on subacute cerebellar dysfunction at onset, cerebellar atrophy in later imaging studies, and the underlying lymphoma, the diagnosis of paraneoplastic cerebellar degeneration (PCD) in this case was definite. PCD is one of classical PNSs, rarely described in MALT lymphoma. PNSs are probably immune-mediated based on the evidence that tumors ectopically express substances mimicking antigens that normally present in the nervous system and onconural antibodies can be identified both in serum and CSF in some patients. While the presence of onconural antibodies is useful in defining a neurological syndrome as paraneoplastic, less than 50% patients with PNSs have known onconural antibodies detectable either in CSF or in serum. Multiple sclerosis (MS) could be an important differential diagnosis, which can be one of neurological complications related to pSS. In this case, MRI results ruled out MS.

Management of the underlying tumor and immunosuppressive therapy are two approaches for PNSs treatment. Syndromes such as PCD are clinically occult and subacute, due to the delayed treatment and irreversible pathological changes leading to severe loss of Purkinje cells of cerebellum, so the treatment often results in disease stability rather than recovery. Considering the immune-mediated pathogenic mechanism shared by PNSs and pSS in this case, immunomodulation is the fundamental treatment. Meanwhile, the intensive management of pSS could decrease the risk of progression of MALT lymphoma. After tumor resection, chemotherapy, and the treatment of immunosuppressants, the patient had no sign of exacerbation over two years.

Here, we presented cerebellar ataxia occurring in a patient with evolutionary pSS. Her cerebellar ataxia was deemed as paraneoplastic and associated with MALT lymphoma. MALT lymphoma was the initial and primary presentation of pSS, which could be regarded as a special extraglandular manifestation of pSS. Encountering patients with MALT lymphoma, especially located in salivary glands, it is necessary to investigate for pSS.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s)/patient’s guardians has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the article. The patients/patient’s guardians understand that their names and initials will not be published and due efforts will be made to conceal the identity of the patient, although anonymity cannot be guaranteed.
Corrigendum: Long non-coding RNA HOTAIRM1 promotes proliferation and inhibits apoptosis of glioma cells by regulating the miR-873-5p/ZEB2 axis

In the article entitled “Long non-coding RNA HOTAIRM1 promotes proliferation and inhibits apoptosis of glioma cells by regulating the miR-873-5p/ZEB2 axis” published on issue 2, pages 174-182, volume 133 of Chinese Medical Journal,[1] the author and affiliation section should be corrected as follows:

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Reference
1. Lin YH, Guo L, Yan F, Dou ZQ, Yu Q, Chen G. Long non-coding RNA HOTAIRM1 promotes proliferation and inhibits apoptosis of glioma cells by regulating the miR-873-5p/ZEB2 axis. Chin Med J 2020;133:174-182. doi:10.1097/Cm9.000000000000915