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Letter to the Editor

Anti-mitochondrial antibody-associated myositis with eosinophilia and dropped head

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Dear Editor,

Idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of diseases with the following recognized main entities: dermatomyositis; polymyositis; necrotizing autoimmune myopathy; sporadic inclusion body myositis; and non-specific myositis [1]. Recently, IIM associated with anti-mitochondrial M2 antibodies (AMAs), sometimes called as AMA-associated myositis, has been identified as a new clinical entity [2]. AMA-associated myositis is clinically characterized by a chronic disease course, muscle atrophy, and cardiac involvement [2,3]. Pathologically, granuloma formation is reported as a characteristic pathological finding of AMA-associated myositis. Nevertheless, it is seen only in 25% of the patients [2]. In contrast, necrotic and regenerating fibers are seen in 96% of the patients, indicating that majority of the patients show myofiber necrosis and regeneration without granulomatous change, which may well be diagnosed as immune-mediated necrotizing myopathy (IMNM) [2]. We, herein, report a case of AMA-associated myositis accompanied with eosinophilia presenting with dropped head that developed rapidly, but without muscle atrophy.

A 60-year-old Japanese man with diabetes mellitus and chronic kidney disease had undergone hemodialysis for 9 years. He had been taking antihypertensive and antidiabetic drugs. He was referred to the hospital due to a 2-week history of elevated eosinophil counts and serum creatine kinase (CK) level at regular blood examinations. He was asymptomatic. Laboratory tests showed a white blood cell (WBC) count of 19,000/μL, with 9025/μL eosinophils, hemoglobin level of 11.3 g/dL, platelet count of 10.9×10^4/μL, CK of 2245 IU/L, asparate transaminase of 70 IU/L, alanine aminotransferase of 125 IU/L, alkaline phosphatase of 315 IU/L, and γ-glutamyltranspeptidase of 11 IU/L. Antinuclear, anti-neutrophil cytoplasmic, and anti-aminoacyl-tRNA synthetase antibodies were all negative. Chest X-ray and transthoracic echocardiography did not show any abnormal findings. Abdominal computed tomography did not show signs of cirrhosis. Bone marrow biopsy did not show myeloid and lymphoid neoplasms.

Three weeks later, he began to feel posterior cervical pain and showed a dropped head. He visited another orthopedic clinic and received intramuscular injection of betamethasone (4 mg) into his neck muscle twice, but without noticeable effect. The strength of his neck extensor muscles was weak, with manual muscle testing grade of 3/5. However, the other muscle strength was preserved, with a grade of 5/5. Muscle atrophy was not observed. His WBC count was 14,300/μL, with 2717/μL eosinophils, and CK level was 2373 IU/L. Positron-emission tomography-computed tomography showed an increased uptake of 18F-fluorodeoxyglucose in the cervical muscles (Fig. 1A), and the short-tau inversion recovery magnetic resonance imaging showed a high intensity in the obliquus capitis inferior, semispinalis cervicis, and levator scapulae muscles without atrophy (Fig. 1B), indicating the presence of active inflammatory process in the muscles. Electromyography in these muscles showed a myogenic change. Muscle biopsy of the left paraspinal muscle at levels C5-C6 provided evidence of active necrotic and regenerating process in the muscle in addition to moderate variation in muscle fiber size but without apparent mononuclear cell infiltration including eosinophils. No nemaline body was observed. On immunohistochemistry, histocompatibility complex class 1 was expressed, and complement C5b-9 membrane attack complex was deposited on the sarcolemma of the non-necrotic muscle fibers. Histologically, an IMNM diagnosis was made. Anti-signal recognition particles and anti-3-hydroxyl-3-methylglutaryl-coenzyme A reductase antibodies were both negative; however, anti-mitochondrial M2 autoantibodies were positive. He was, therefore, diagnosed as having AMA-associated myositis accompanied with eosinophilia. Treatment with oral prednisolone 60 mg/day was initiated immediately after the muscle biopsy, and then his symptoms began to improve. The eosinophil count rapidly became normal, and CK level also began to decrease. One and a half months later, he became asymptomatic and CK returned to normal level. Prednisolone was tapered without recurrence and terminated 13 months after the initiation.

AMA-associated myositis usually shows a chronic disease course and muscle atrophy [2,3]. Muscle involvement appeared subtle initially, sometimes detected solely by asymptomatic CK elevations, occurring over the course of months to years [2,3], and this disease course can be confused with muscle dystrophy. However, in the present case, muscle symptoms developed rapidly approximately one month after the CK elevations, which was coincidentally found at regular blood examinations.

Often IIM involves the neck flexor muscles more predominantly than the extensors, which is included in the classification criteria on IIM of the European Neuromuscular Centre [4]. In fact, only a few IIM cases

Abbreviations: IIM, idiopathic inflammatory myopathy; AMA, anti-mitochondrial antibody; IMNM, immune-mediated necrotizing myopathy; CK, creatine kinase; WBC, white blood cell

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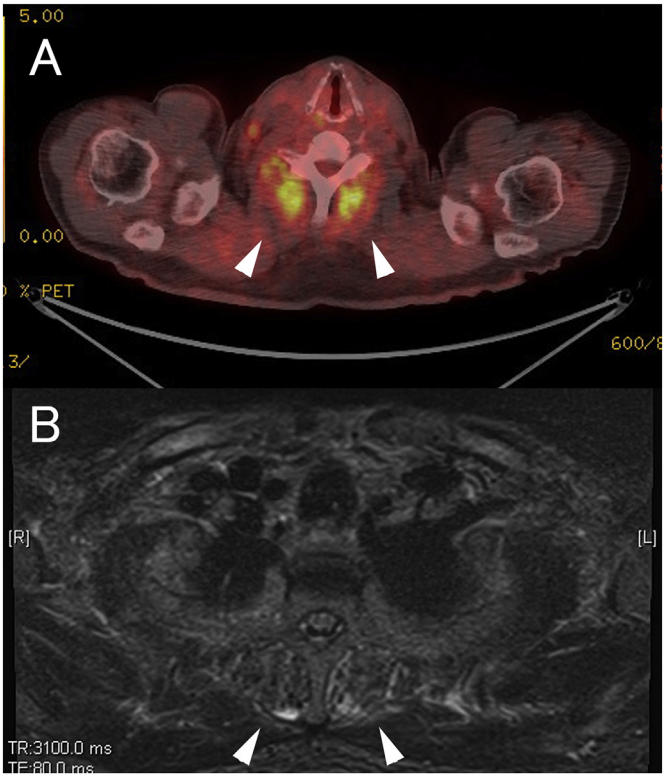

Fig. 1. (A) Positron-emission tomography-computed tomography shows an increased uptake of 18F-fluorodeoxyglucose in cervical muscles (arrow heads). (B) The short-tau inversion recovery magnetic resonance imaging demonstrates high-intensity in semispinalis cervicis without atrophy (arrow heads).

with neck extensor weakness or dropped head have been reported [5,6]. On the other hand, in AMA-associated myositis, paraspinal muscles are sometimes affected and become atrophic, leading to lordotic posture or dropped head [2,3]. Maeda et al. reported that muscle CT images were assessed in 13 of 24 patients of AMA-associated myositis, and 10 patients showed muscle atrophy with proximal dominance. Three patients were reported to show lordotic posture and have muscle atrophy with fatty changes in paravertebral muscles, and the disease duration before diagnosis was more than 2 years [2]. In the present case, dropped head developed rapidly with muscle inflammation, but without muscle atrophy.

IIM accompanied with eosinophilia is rare. Only a few cases have been reported, but the cause of eosinophilia was unknown [7]. The etiological relationship between AMA-associated myositis and eosinophilia is not known in our patient; however, both serum CK elevations and eosinophilia appeared at the same time and resolved in response to corticosteroid therapy without recurrence after cessation of the therapy. Our patient may be the first case of AMA-associated myositis showing eosinophilia without primary biliary cirrhosis.

In conclusion, AMA-associated myositis can follow a relative acute disease course. Physician should be aware that acute development of dropped head may be one of the symptoms of AMA-associated myositis, even without muscle atrophy.

Declarations of interest

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