A unique case of hemi-tongue pseudohypertrophy, necrotizing myopathy, and erythema nodosum

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Abstract

A 46-year-old woman developed slowly progressive tongue weakness with a pseudohypertrophic change on the right side of her tongue. She subsequently developed weakness in her proximal lower extremities, skin erythema and a sustained increase of muscle enzymes at 11 M after the onset. A biopsy of the quadriceps muscle showed necrotizing myopathy and a skin biopsy showed erythema nodosum. The present case showed characteristic clinical manifestations that may represent a rare variant of sarcoidosis.

Introduction

Herein, we report a 46-year-old woman who developed slowly progressive tongue weakness with a pseudohypertrophic change, and consecutively displayed an increase in muscle enzymes, weakness in her proximal lower extremities, and skin erythema at 11 M after the onset.

Case Report

A woman began to feel a swelling of the tongue at the age of 46. Within 3 months (M), she had developed dysarthria, and dysphagia by 4 M, and because her symptoms gradually worsened, she was admitted to our hospital by 6 M.

Upon initial admission to our hospital, the right side of her tongue was mildly swollen, and upward tongue movement was primarily disturbed (Figure 1A-D). She showed mild weakness in her neck flexion (MMT 4), but did not show any other neurological deficits. Laboratory data revealed a normal ESR (11/24 mm) and WBC (4,620 /µL), but serum CK was markedly elevated at 2,940 IU/L with a mild increase in aldolase (ALD, 23.1 U/L), AST (68 IU/L), ALT (72 IU/L) and LDH (696 U/L). Serum calcium (9.1 mg/dL), angiotensin converting enzyme (ACE, 7.0 U/L), and soluble IL-2 receptor (sIL-2R, 417 U/mL) were normal. Serum autoantibodies such as ANA, SS-A, SS-B, ds-DNA antibody, MPO-ANCA, and PR3-ANCA were all negative. The thyroid hormone was normal. Several infection tests for syphilis, hepatitis B, hepatitis C and HIV all tested negative. Analysis of cerebrospinal fluid (CSF) revealed a normal cell count (1/µL), protein (38 mg/dL).

A whole-body CT showed neither lymphadenopathy (BHL) nor tumors. A brain MRI showed no lesion in the medulla oblongata or hypoglossal canal (Figure 1E). A tongue MRI showed hyperintense on the right side of her tongue, both in the T1 weighted image (T1WI) and the T2 weight-ed image (T2WI) (Figure 1F and G, arrowheads). A muscle biopsy of the quadriceps muscle showed necrotizing myopathy and a skin biopsy showed erythema nodosum. The present case showed characteristic clinical manifestations that may represent a rare variant of sarcoidosis.

Discussion and Conclusions

The present case initially showed weakness on the right side of the tongue with pseudohypertrophy (Figure 1A-D) and fatty degeneration (Figure 1F-H). Tongue weakness and atrophy are usually caused by hypoglossal nerve palsy, but tongue pseudohypertrophy is also caused by idiopathic hypoglossal nerve palsy.1 As for the present case, a tongue MRI and tongue biopsy showed similar finding to the previous case.1 In addition, a unilateral limited tongue lesion suggests hypoglossal nerve palsy (Figure 1F-H).

Lobular panniculitis of skin erythema is a typical pathological change to erythema nodosum, which is an important physical finding in systemic diseases such as sarcoidosis, Behçet’s disease, infection, and cancer.2 In the present case, both infection and malignancy were ruled out by her clin-
ical course, blood tests, and whole-body CT. A quadriceps muscle biopsy revealed mild necrotizing myopathy (Figure 1O-P; arrows). Sarcoidosis can present both myopathy without typical lymphadenopathy,3 and hypoglossal nerve palsy.4,5 Although sarcoidosis-specific findings such as elevated serum marker (ACE, lysosome, and Ca), lymphadenopathy, the accumulation of $^{67}$Ga, and noncaseating granuloma were not shown in the present case, sarcoidosis is more likely than other etiologies. A successive and careful follow-up will hopefully uncover the exact diagnosis in the future.

References
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