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Resistant dermatomyositis as a manifestation of post COVID-19 infection: A case report

[1]Hugo D. Wiradarma, [1]Herlyani Khosama, [1]Arthur H.P. Mawuntu, [1]Seilly Y. Jehosua, [2]Aryani Adji, [3]Meilany F. Durry, [4]Luh A. Indrawati

[1]Department of Neurology, [2]Department of Dermatology and Venereology, [3]Department of Anatomical Pathology, University of Sam Ratulangi/ Prof. Dr. R.D. Kandou Hospital, Manado, North Sulawesi, Indonesia; [4]Department of Neurology, Universitas Indonesia/ Dr. Cipto Mangunkusumo Hospital, Jakarta, Indonesia

Abstract

Many autoimmune diseases were reported as a sequel of SARS CoV-2 infection. We report here a 51-year-old female who presented with myalgia, progressive quadriparesis, and skin rash 2 weeks after COVID-19 infection. She improved with high-dose methylprednisolone. She was given oral methotrexate and mycophenolic acid during follow up. However, her condition deteriorated until she passed away half a year after the initial illness. This case showed that dermatomyositis can be associated with COVID-19 infection.

Keywords: Dermatomyositis, Covid-19, autoimmune

INTRODUCTION

Autoimmune disease in Coronavirus disease 2019 (COVID-19) survivor is challenging, because not much is known of the autoimmune complication. The underlying pathophysiological mechanism is uncertain, whether it is a virus-induced or a virus-triggered inflammation. We present here a a patient of dermatomyositis who developed the illness after recovery from COVID-19 infection.[1-3]

CASE REPORT

A 51-year-old female presented with one week history of muscle weakness, myalgia on her shoulders and thighs, and myoglobinuria. She had rash in face and back one week prior to the muscle symptoms. There was no other symptoms, including weight loss. She did not have past history of cancer, and was taking metformin and allopurinol for diabetes mellitus and hyperuricemia. One month prior to hospital admission, she contracted COVID-19, having fever and hypoxia without muscular symptoms, and recovered after two weeks of treatment without further complication.

On presentation she was alert, had hypertension (157/85mmHg), tachycardia (122 times/min), was afebrile, and had normal oxygen saturation. She had skin rash on the face, shawl sign, proximal fingers joint, and muscle pain that was aggravated with pressure. There was also arm edema. Pulmonary and cardiac examination were unremarkable. (Figure 1)

Motor examination showed proximal predominant weakness, hypotonia, and hyporeflexia while other neurological examinations were unremarkable. Laboratory examinations showed leukocytosis (15.500/µL with predominant segmented neutrophil 80% and lymphocyte 11%), elevated AST and ALT (222 and 1383 U/L, respectively), creatinine kinase (8829 U/L), CKMB (1009 U/L), and CRP 48 mg/L. Electromyography examinations revealed myogenic MUAP, fibrillation, and positive sharp waves. Autoantibodies (anti RNP, Sm, SS-A, Ro-52, SS-B, PM/Scl100, Jo-1, Centromere B, PCNA, dsDNA, Nucleosomes, Histones, AMA-M2, Rib-P, DFS70) were negative. Muscle imaging was not available. Her skin biopsy showed sparse perivascular lymphocytic infiltration and vacuolar changes in the basal layer. Chest x-ray was unremarkable. Tumor markers panel (CEA, AFP, CA 125, CA 15.3, CA 19.9) showed only mild nonspecific elevation. (Figure 2)

Address correspondence to: Hugo D. Wiradarma, University of Sam Ratulangi/ Prof. Dr. R.D. Kandou Hospital, Manado, North Sulawesi, Indonesia. Email: amsahugo@yahoo.com

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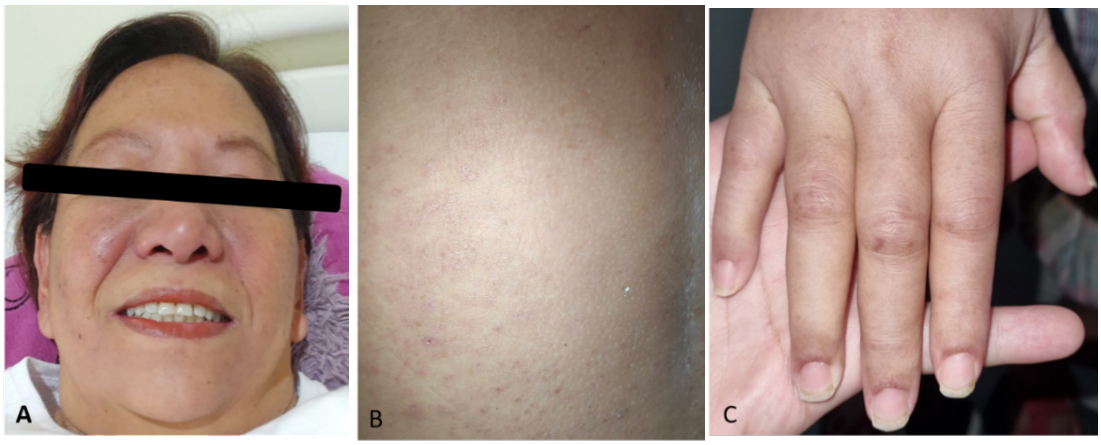

Figure 1. A. Rash on face, B. Rash on back, C. Hyperpigmentation on proximal interphalangeal joints resembling Gottron papule and periungual hyperpigmentation lesions.

She was diagnosed to have dermatomyositis in association with post COVID-19 infection. High dose methylprednisolone (60 mg/day) and methotrexate (5-7.5 mg/day weekly) were administered. Although the myalgia gradually resolved and laboratory value normalized, muscle strength did not recover and she developed dysphagia. Fiberoptic endoscopic evaluation of swallowing displayed no significant abnormality. On six months follow-up, methotrexate was switched into mycophenolic acid 720 mg/day as there was no clinical improvement. However, not long after switching, her condition deteriorated further resulting in worsening muscle weakness, dyspnoea, and dysphagia until she eventually died because of aspiration.

DISCUSSION

Dermatomyositis is a rare disease which is more prevalent in females, associated with type 1 interferonopathy, and characterized by features of skin manifestation, muscle inflammation, and can be associated with other organ manifestations such as interstitial lung disease, arthritis, and malignancy. We made the diagnosis of dermatomyositis according to ENMC criteria from 2018 workshop based on the presence of lesions resembling Gottron's papule, interface dermatitis, proximal muscle weakness, and elevated muscle enzyme in addition to other skin manifestations, although not supported by the important dermatomyositis-specific autoantibodies panel.

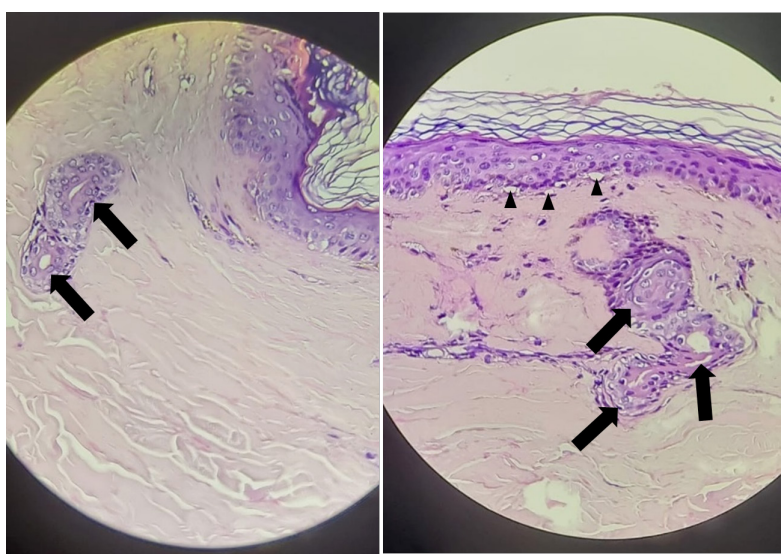

Figure 2. Skin biopsies revealed sparse perivascular lymphocytic infiltration by white blood cells (arrow) and vacuolar changes in the basal layer (arrow head).

Dermatomyositis has a characteristic of vascular inflammation which causes perifascicular atrophy in pathologic examination, suggesting that dermatomyositis might be a vasculitis autoimmune process. Skin biopsy displayed perivascular infiltration and vacuolar changes in basement membrane suggesting interface dermatitis which often seen in dermatomyositis.[4-6]

COVID-19 is a global pandemic which had been previously reported to have an association with autoimmune diseases, including dermatomyositis. In our patient, the close temporal relationship to COVID-19 suggested the possibility of association of dermatomyositis to post COVID-19. The mechanism of COVID-19 induced-dermatomyositis is uncertain but there were reports of the association. About 10% of COVID-19 patients developed muscle symptoms and elevated CK. It is believed that RNA virus infection along with other environmental factors and genetic susceptibility culminates in type 1 interferonopathy causing dermatomyositis. Furthermore, dermatomyositis patients have three linear epitopes of immunogenicity having high sequence identity with SARS-CoV-2 protein. Dermatomyositis with seronegative antibodies to SAE, MDA5, NXP2, and Mi2 has been reported in association with COVID-19. It is believed that in dermatomyositis and COVID-19 interferon-1 (IFN1) is upregulated. A persistent IFN response promotes antigen presentation resulting in T and B cells activation which may be responsible for production of autoantibodies. Myxovirus resistance protein A, the signature of interferon activation, is observed in dermatomyositis. Janus kinase type 1 and 2 inhibitors were reported to be efficacious in severe COVID-19, indicating overlapping disease pathways in interferon. Regarding the presence of myoglobinuria and prominent CK elevation resembling CK level in immune-mediated necrotizing myopathy, we suspected that our patient had Mi-2. Study showed that among dermatomyositis subtypes, the highest CK elevation is identified in Mi-2. [3-7]

Although our autoantibodies findings was limited to negative anti-tRNA synthetase panel, we could not exclude the possibility of post COVID-19 associated dermatomyositis because of our patient's clinical manifestations were closely associated with COVID-19 infection, and no other causes could be found, with no signs and symptoms of malignancy.

Dermatomyositis is generally treated with glucocorticoid plus azathioprine or methotrexate as a first line treatment. Nevertheless, our patient did not respond well to the treatment which suggested that she has resistant type dermatomyositis. Intravenous immunoglobulin or plasmapheresis could be considered, however these were not given due to financial consideration.[8]

In conclusion, this case add to the case of DM in association with COVID-19 reported in the literature. Dermatomyositis can occur after COVID-19 infection because of the similarity in the regulation of IFN1 in both conditions.

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DISCLOSURE

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Conflict of interest: None

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