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Challenges in The Diagnosis and Management of Idiopathic Inflammatory Myopathy: A Case Report

Alexander Tonny Limono[1*], Pande Ketut Kurniari[1], Gede Kambayana[2]

[1]Department of Internal Medicine, Faculty of Medicine,
Universitas Udayana / RSUP Prof IGNG Ngoerah, Bali, Indonesia

[2]Rheumatology Division, Department of Internal Medicine, Faculty of Medicine,
Universitas Udayana / RSUP Prof IGNG Ngoerah, Bali, Indonesia

*Corresponding author details: Alexander Tonny Limono; alexlimono@gmail.com

ABSTRACT

Background: Idiopathic inflammatory myopathy (IIM) is a group of heterogeneous disorders characterized by muscle weakness and muscle inflammation, elevated muscle enzyme, a sign of muscular inflammation, and other extra muscular manifestations. Due to how rare IIM is, as well as its lack of targeted treatments and the heterogeneity of the clinical presentation, significant challenges are posed to treatment deliveries. ***Case Illustration:*** A 30-year-old woman came with complaints of weakness in all four extremities 8 months ago accompanied by difficulty swallowing. The weakness worsened to the point where the patient had shortness of breath and a fever. Upon arrival at the hospital, the patient was in respiratory failure and intubated. Laboratory examination results showed an increase in serum creatine kinase (14.977 U/L), increased liver enzymes, and inflammatory biomarkers such as CRP and procalcitonin. Muscle biopsy results showed muscular dystrophy. Chest X-ray showed cardiomegaly with signs of pulmonary inflammation and pleural effusion. The patient received steroid therapy, cyclophosphamide, and antibiotic management for pneumonia. During treatment, the patient's condition worsened and she was declared dead due to sepsis. ***Conclusion:*** IIM is a rare disease and has various clinical variations. Extra muscular manifestations such as pulmonary disease complicate the management of IIM.

Keywords: idiopathic inflammatory myopathy; autoimmune disease; muscle inflammation.

INTRODUCTION

Idiopathic inflammatory myopathies (IIM) are a heterogeneous group of diseases characterized by muscle weakness, elevated serum creatine kinase (CK) levels, signs of inflammation on muscle biopsy, and extra muscular manifestations.[1] IIM is a rare condition, with an incidence of 1.16 to 19 cases/million/year and a prevalence of 2.4 to 33.8/100,000 people.3 Currently, several therapeutic modalities are available for IIM, but none are specific.6 Meanwhile, clinical trials related to appropriate management guidelines are also lacking due to the rarity of IIM.[2] The management of IIM is further complicated by extra muscular manifestations, such as interstitial lung disease and arthritis.[3] This report is intended to raise awareness of idiopathic inflammatory myopathies in adults.

Case Illustration

A 30-year-old woman presented with complaints of weakness in all four extremities since 8 months ago. The weakness started from the proximal upper limbs which gradually worsened and spread to other areas of the body until it was difficult to swallow food. In the past week, the patient complained of shortness of breath accompanied by coughing and fever. In the past 8 months, her weight has decreased by ±5 kg.

At the time of examination, the patient was intubated, vital signs showed blood pressure of 100/60 mmHg, pulse 145x/min, respiration 22x/min with ventilator, and temperature 37.7'C. Physical examination showed atrophy of the upper and lower extremity muscles. Complete blood count laboratory tests showed leucocytosis (11,290 µL) with dominant neutrophil cells (9.77), anaemia (Hb 8.2 g/dl), and thrombocytopenia (70,000/µL). Also found was an increase in creatine kinase (14977 U/L), an increase in SGOT (113.3 U/L) and SGPT (148.61 U/L), an increase in procalcitonin (152.85 ng/mL), LED (46 mm/hour), and CRP (228.07 mg/dL).

On examination of muscle biopsy on the left deltoid muscle and left gastrocnemius muscle, shows few muscle tissues. The muscle cells are of variable size. Fatty replacement and endomysial fibrosis are also seen. Histomorphology shows a muscular dystrophy. (Figure 1.). ANA IF examination was negative. Chest X-ray showed cardiomegaly, pneumonia, and bilateral pleural effusion. MRI head examination with contrast was within normal limits, to rule out brain abnormalities.

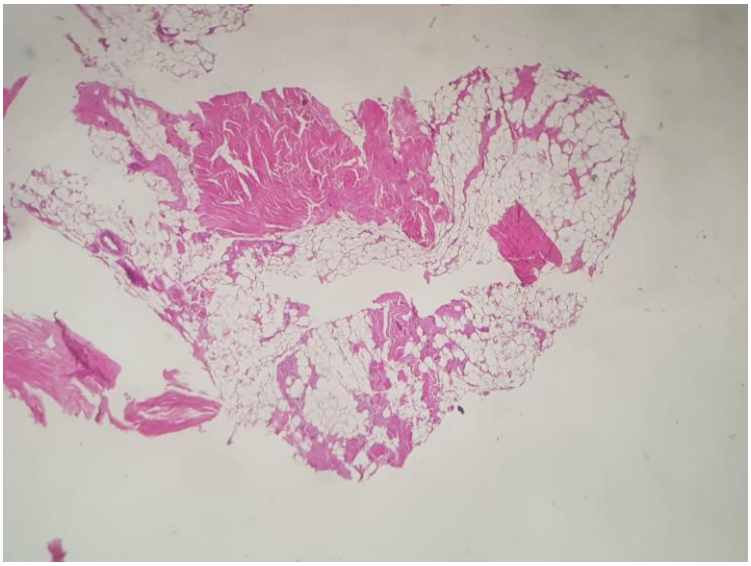

FIGURE 1: Histomorphology of the deltoid and gastrocnemius muscles shows a muscular dystrophy.

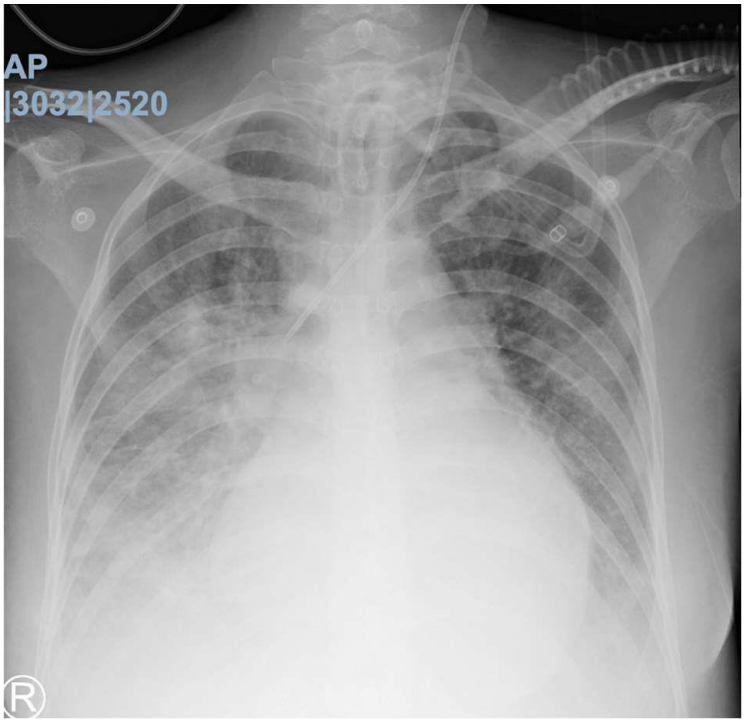

FIGURE 2: Chest X-ray showed cardiomegaly, pneumonia, and bilateral pleural effusion.

The patient was diagnosed with idiopathic inflammatory myopathy (IIM), The diagnosis of idiopathic inflammatory myopathy was established based on the ACR EULAR criteria, where in this case the following criteria were found: age >18 years and <40 years (score 1.5); objective symmetrical muscle weakness that is progressive and in the proximal upper extremities (score 0.7); symmetrical muscle weakness in the proximal lower extremities (score 0.5); neck flexors are weaker than neck extensors (score 1.6); proximal leg muscles are relatively weaker than distal muscles (score 1.2); dysphagia (score 0.6); increased serum CK levels (score 1.4); and perifascicular atrophy (score 1.9).

The aggregate/final score in the case was 9.4; so it is included in the IIM classification. Community-acquired pneumonia with pneumonia severity index (PSI) class 5 classification with sepsis and thrombocytopenia, hypokalaemia caused by inadequate intake, hypoalbuminemia caused by chronic inflammation, mild normochromic normocytic anaemia caused by anaemia in chronic diseases, and immobilization with a high risk of venous thromboembolism.

During treatment, the patient was given fluid therapy and supportive nutrition. Also given Methylprednisolone 125 mg every 12 hours IV and Cyclophosphamide 1 gram in 250 cc of 0.9% NaCl for 3 days, paracetamol 1 gram every 8 hours, drip KCl 50 mEq every 12 hours, post-transfusion 2 bags of packed red cells, post-transfusion Albumin 20% 3 flasks, Amikacin 1.5 grams every 8 hours IV until day-10 then continued with Meropenem 1 gram every 8 hours IV and compression stockings. During treatment, pneumonia worsened and finally, the patient died due to sepsis.

DISCUSSION

IIM is classified based on clinical features, age of onset, immunohistopathology, and response to therapy. Some of the most common types include dermatomyositis (DM), polymyositis (PM), necrotizing autoimmune myopathy (NAM), and sporadic inclusion body myositis (IBM).[1,2,] The prevalence of IIM ranges from 2.4 to 33.8 per 100,000 people, with the female gender being more dominant, especially in the DM type. However, IBM is said to be more common in men, and the age predilection for over 50 years is more dominant.[1,3]

Autoimmune mechanisms appear to underlie the process of inflammatory myopathy.[4] The pathogenic mechanisms of most immune-mediated diseases are associated with chronic organ inflammation that can be caused by specific interactions between genetic and environmental risk factors.[5] Activation of the immune system in these diseases often involves innate and adaptive mechanisms, as well as non-immune mechanisms. The inherited HLA 8.1 haplotype is a major risk factor for the common IIM phenotype in some populations. Meanwhile, environmental risk factors can include viruses, bacteria, ultraviolet radiation, smoking behaviour, occupational and perinatal exposure, drugs (including biological agents), and dietary supplements.[4,6] In this case, a 30-year-old woman complained of weakness throughout the body since 8 months earlier. Weakness began to be felt in the upper limbs, when lifting both shoulders, accompanied by difficulty swallowing.

In a complete blood laboratory examination, leucocytosis was found, as an increase in creatine kinase (CK) of 14977 U/L, and an increase in procalcitonin, LED, and CRP.

A muscle wall biopsy examination found muscle lesions/myopathy. The European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) developed a new classification system for IIM based on a probability score model. These criteria use clinical findings (muscle weakness, extra muscular manifestations, and skin rashes), laboratory findings (Anti-Jo1 and muscle enzymes), and muscle biopsy findings to produce a final score. In this patient, it was 9.4; so it is included in the IIM classification.

The main goal of IIM therapy is to restore muscle strength, reduce inflammation, and prevent damage to other organs.[7] Management of this condition requires a multidisciplinary approach including neurology, rheumatology, dermatology, pulmonary, and physiotherapy. [6,8] Management consists of non-pharmacological and pharmacological. Non-pharmacological management in the acute phase includes bed rest and passive movement to prevent contractures. After the acute phase is resolved, patients should do active exercises to prevent muscle atrophy and contractures. Prospective, double-blind, placebo-controlled studies published for the treatment of IIM are still very limited.[9] Current treatment recommendations are presented in Figure 3. However, these recommendations are still considered inadequate, because the response to treatment continues to vary among IIM patients. Treatment of IIM includes immunosuppression such as corticosteroids and high-dose intravenous immunoglobulin/IVIG. Several therapies that are still in phase 3 research include T-cell modulators (abatacept) and cannabinoid receptor type 2 agonists (lenabasum) for DM. [6,9,10]

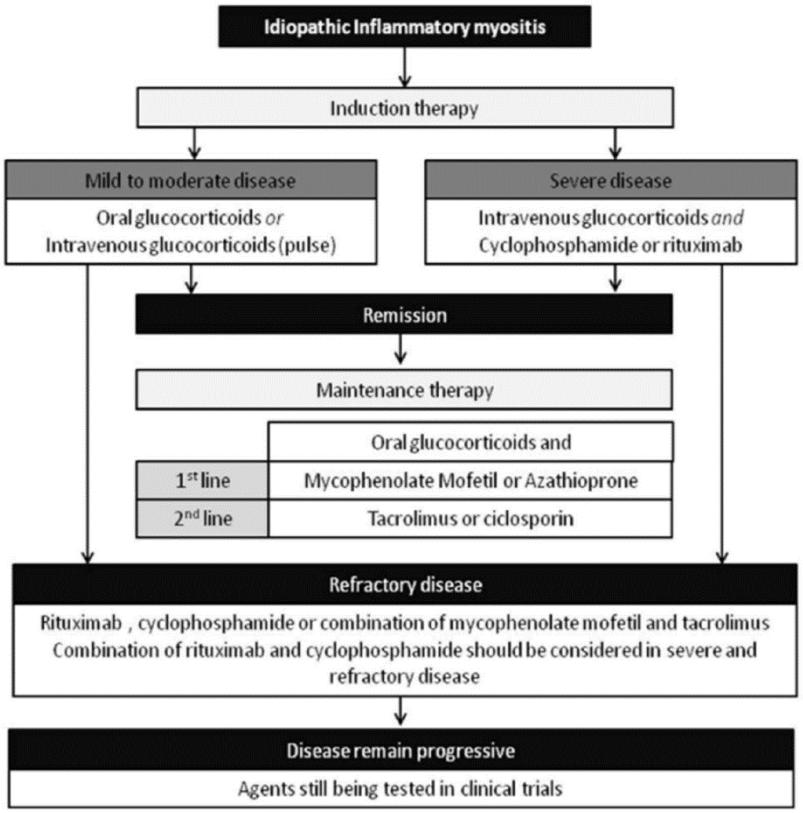

FIGURE 3: IIM therapy recommendations.

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