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Case report on chronic autoimmune hemolytic anemia in mixed connective tissue disease

[1]Dr.K.Shivaraju, Department of internal medicine, Krishna institute of Medical Sciences, Secunderabad -500003, Telangana, India

[2]Mandhala Saikrishna, Pharm. D, Bharat Institute of Technology, JNTUH, Telangana, India.

Corresponding Author: Mandhala Saikrishna, Pharm. D, Bharat Institute of Technology, JNTUH, Telangana, India.

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Abstract

Mixed connective tissue disease (MCTD) generally presents as mild anaemia and other systemic organ involvement but, in this case, chronic anaemia is a prominent feature of haemolysis and systemic abnormalities were not seen in this case significantly.

Mixed connective tissue is an overlapping disease (MCTD), that was first identified in the year of 1972 recognized as an entity disease with a combination of some clinical features. Autoimmune haemolytic anaemia (AIHA) is characterized by the producing of antibodies against self-red blood cells. autoimmune haemolytic anaemia is a rare disorder with the estimate of incidence 1-3 cases 100,000 per year. Here we present a case of 75 years old female patient who was admitted to the tertiary care hospital with complaints of chronic lower back pain from a few weeks and complaints of SOB on exertion and abdominal fullness after eating. She had a past history of haemolytic anaemia and low haemoglobin from few months and multiple blood transfusions were done and usage of steroids presently. After clinical evaluation, she was finally diagnosed with mixed connective tissue disease-induced chronic autoimmune haemolytic anaemia. After that, she had been treated with appropriate medications and two units of least incompatible blood transfusions were done. Finally, her condition was clinically improved and symptoms were subsided gradually.

keywords: Mixed Connective Tissue Disease, Autoimmune Haemolytic Anaemia, Haemolysis, Auto Antibodies.

Introduction

Mixed connective tissue is an overlapping disease (MCTD), has been recognized as an entity disease with a combination of some clinical features from systemic lupus erythematosus, polymyositis or dermatomyositis, scleroderma, systemic sclerosis, rheumatoid arthritis, Sjogren's syndrome. The prevalence of MCTD in the Indian for Alaska America is 6.4 per 100000 population in Japan 2.7 patients per million population are reported (1). Autoimmune haemolytic anaemia (AIHA) is characterized by producing antibodies against red blood cells. autoimmune haemolytic anaemia is a fairly uncommon disorder with an estimated of incidence 1-3 cases per 100,000 per year. AIHA is often associated with extra haematological features as severe renal and

CNS involvement which may require corticosteroid and immunosuppression treatment (2).

Case presentation

75 years old female patient was admitted to the tertiary care hospital with complaints of chronic lower back pain from few weeks and complaints of SOB on exertion and abdominal fullness after eating. She was a past history of haemolytic anaemia and low haemoglobin from few months and multiple blood transfusions were done and usage of steroids presently. She was no history of bleeding manifestations, no chest pain, no palpitation, no decreased urine output, or burning micturition. After clinical evaluation, routine investigations were sent which showed severe anaemia (Hb 5gm/dl), Direct Coombs test 4+& indirect coombs test 2+, and low complement levels (C3&C4). In the ANA profile, ANA-IF showed positive and strongly positive for SSA and SSB and RO52 positive, increased reticulocyte count. Based on these findings she was diagnosed with mixed connective tissue disease-induced chronic autoimmune haemolytic anaemia. based on physical examination laboratory investigations she was found to have osteoporosis of the lumbar spine.US Whole abdomen showed hepatosplenomegaly. 2D Echo report was showed dilated right atrium, papillary muscle calcification, no pulmonary artery hypertension (PAH), no pulmonary embolism (PE)or clots.

In view of severe anaemia patient planned for a blood transfusion but because of high incompatibility discuss it with blood bank officials and transfused the least incompatible blood with precautions 2 units of least incompatible blood transfusion and no side effects were observed. After transfusion medical oncologist review was taken and started and steroids [IV initially later oral and MMF (mycophenolate mofetil)]. The patient clinically improved, haemoglobin (8.2) increased, advised discharge, and to follow up on an OPD basis. Discharge medications were a combination of tolperisone and paracetamol, a combination of pregabalin and nortriptyline, rabeprazole with domperidone, prednisolone 50mg for 5 days and 40mg next 5 days, and tablet MMF 500 mg.

Discussion

MCTD which was first identified in 1972 is a syndrome with overlapping clinical features of systemic sclerosis and polymyositis/dermatomyositis along with a positive anti-U1 RNP Antibody. The characteristic presenting symptoms include Raynaud’s phenomenon, interstitial lung disease, pulmonary artery hypertension, arthritis, swollen hands, and proximal muscles weakness. In contrast to SLE renal and CNS involvement is not seen in MCTD. A population-based study done in Norway revealed that MCTD is a rare occurrence at a rate of 0.21 per 100000 adults and more common in females than in males approximately 75% of patients have low-grade anaemia while some patients have leukopenia but thrombocytopenia is seen very rarely and cases of MCTD presenting with bleeding manifestations are rare (3). Some features are similar and some features contrast in this case because the patient had chronic anaemia& no bleeding manifestations were seen.in contrast, thrombocytopenia was seen in our case.

The patient's preliminary presentation usually includes non-unique signs such as arthralgia, myalgia or muscle weakness, swollen digits, dysphagia or acid reflux, Raynaud’s phenomenon, SOB on activity, general malaise, and fatigue. Some features were seen in our case those were myalgia and shortness of breath on exertion. Over some time, signs and symptoms are

dominated by symptoms of any one of the three diseases along with high titres of anti-U1 RNP antibody. The etiology of MCTD is unknown because it is an autoimmune disease MCTD can run in families and is known to affect women are more often than men. And in this case patient with no family history of MCTD and the gender is female. The lack of causal relationship and a variety of manifestations make it difficult to diagnose this real condition (4).

The appearance of swollen hands or fingers associated with the high levels of ANA should require careful monitoring and observation for the possible appearance of overlapping features. High concentrations of anti-RNP antibodies are a strong predictor of the development of MCTD in these patients. usually, MCTD will affect any organ system. MCTD over different connective tissue diseases Raynaud's phenomenon and or swollen hands/puffy fingers, no extreme renal & CNS affection, greater extreme arthritis than the typical insidious onset of pulmonary artery hypertension not associated with fibrosis of the lung. the primary complaint of MCTD is pyrexia of unknown origin. Tracing fever to a coexisting serositis myositis aseptic meningitis, lymphadenopathy, or intercurrent infection may occur. Most MCTD patients have initial involvement of the skin. Severe joint involvement may see in MCTD patients. In contrast to our case, the patient had no features of severe joint involvement. About three-quarters of patients with MCTD exhibit pulmonary involvement. But in this case, no lung involvement was seen. 30% of patients with MCTD have symptoms associated with cardiac involvement and up to 40% have the subclinical disease, all three layers of the heart can be affected. Cardiac involvement alone is responsible for one-fifth of MCTD related deaths. the myocardial disease can occur and if so, it could be secondary to pulmonary artery hypertension, which is often asymptomatic in the early stages. In this case, is 2D echo report was showed that grade 1 left ventricular diastolic dysfunction and papillary muscle calcification and dilated right atrium. The absence of chronic kidney disease is typical in MCTD. In this case, also no abnormal renal function was seen. Patients with MCTD measure unspecific laboratory and haematological abnormalities. the only universal finding is ANA positive. other common abnormalities are mild anaemia leukopenia which mainly affects lymphocytes, rheumatoid factor positive and anti-cyclic CCP antibodies, antibodies against heterogeneous nuclear ribonucleoprotein antiphospholipid antibodies which are less common than in SLE. Less common and laboratory abnormalities include thrombocytopenia, thrombotic thrombocytopenic purpura, coombs-positive haemolytic anaemia, and red blood cell aplasia. (5). In this case, the patient presented with severe anaemia thrombocytopenia Congress positive haemolytic anaemia.

Mote S et al, reported one case of mixed (cold& warm) autoimmune haemolytic anaemia in SLE. But in our case patient had a warm type of autoimmune haemolytic anaemia (2).

Katewa R et al, reported one case of mixed connective tissue disease with a late presentation of pulmonary artery hypertension (4). But in our no pulmonary artery hypertension was seen.

David S et al reported a case of coombs positive haemolytic anaemia after the clinical onset of scleroderma. But in our case homes, positive

haemolytic anaemia is present but no signs of scleroderma were observed (6).

There is no particular therapy for MCTD and interventions are directed at alleviating the symptoms. Dealing with pain and fatigue is a major challenge, as pain is still under-treated and fatigue is still ineffectively assessed. non-pharmacological strategies and specialized programs are needed, such as exercise patient education, supportive measure for physiological and emotional burden management are needed. The treatment choices are restricted to NSAIDs, corticosteroids, and hydroxychloroquine and their combinations, immunosuppressive drugs such as azathioprine, methotrexate, MMF, cyclosporine, and biologics such as infliximab, etanercept, rituximab. Therefore, there is a need for alternative therapies with fewer side effects (7&8).

Conclusion

In MCTD, normally mild anaemia is present and other organ abnormalities were seen extensively. But here chronic anaemia is a prominent feature of significant haemolysis required blood transfusions and other systemic dysfunctions were not seen in this case significantly. so we are concluding that clinicians should suspect MCTD when chronic anaemia is present and even absence of other organ abnormalities.

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