Case Report

Dermatomyositis and autoimmune hepatitis

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Association of dermatomyositis and autoimmune hepatitis: A case report

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Abstract

The association of dermatomyositis (DM) and autoimmune hepatitis (AIH) is rare and presents a diagnostic and therapeutic challenge. We describe the case of a 36-year-old man with DM diagnosed in 2012 and treated with corticosteroid and methotrexate. The patient achieved total remission 18 months later. In 2022, an AIH was diagnosed (cytolysis, cholestasis, anti-LC1, and anti-SLA antibodies) while DM was in remission. Liver function normalized after two months of treatment with mycophenolate mofetil and corticosteroids. Liver damage in systemic autoimmune diseases can result from viral, iatrogenic, or autoimmune processes. The association between DM and AIH is exceptional and has only been documented in one previous observation. Autoantibodies are essential for diagnosing and managing patients with inflammatory myopathy and AIH. In conclusion, this exceptional association of AIH and DM raises many questions regarding the presence of etiopathogenic links, such as genetic predisposition, autoimmunity disorders, viral infection triggers, or simply a happenstance.

Keywords: Autoimmune; hepatitis; myositis.

Introduction

The association of dermatomyositis (DM) and autoimmune hepatitis (AIH) is exceptional and presents a diagnostic and therapeutic challenge.[1] We report a new observation that underscores the utility of autoantibodies in the screening of autoimmune diseases associated with DM.

Case Report

A 36-year-old male was diagnosed with DM in 2012 based on the association of periorbital erythroderma, banded erythema of the hands, bilateral and symmetrical proximal muscle deficit, inflammatory polyarthralgia, Raynaud's syndrome, chronic cough and dyspnea with a chronic cough. Mega capillary-specific microangiopathy was identified using capillaroscopy.

Laboratory investigations showed an inflammatory syndrome and myolysis at 1.5 times the usual range. Nerve conduction studies showed marked myopathic motor units consistent with myopathy. Chest X-rays revealed diffuse reticulonodular opacities. Interstitial lung disease (ILD) with fibrosis was assessed by chest computed tomography (CT). The spirometry was normal. Anti PM/Scl 75 and anti-SSA-Ro52 antibodies were positive in the immunological blood tests with no positivity of myositis-specific antibodies. No underlying neoplasia was detected. The diagnosis of DM was established. The patient had no other signs of another connective tissue disease (especially systemic scleroderma). The patient was treated with oral route corticosteroid at 1 mg/kg/day and six-monthly pulses of cyclophosphamide at 0.7 g/m^2 of body surface area. Methotrexate (10 mg per week) was afterward prescribed as a maintenance therapy. The clinical outcome was favorable with the disappearance of the cough and an improvement in dyspnea (grade I). The different controls of chest CT showed stability of the ILD and all along the follow-up, the respiratory function tests were normal. Methotrexate was withdrawn in 2017. In December 2022, (the patient was 46 years old), the routine blood analysis revealed cytolysis (Aspartate aminotransferase (AST)=259UI/l [normal laboratory range <34 IU/L] and alanine aminotransferase (ALT) = 419 IU/L [<55 IU/L]) and elevated gamma-glutamyl transferase (GGT)=118UI/L [9–36])). There was no sign of relapses of the DM (creatine kinase (CK) and lactate dehydrogenase (LDH) levels were normal). Abdominal ultrasound was normal. Anti-HBc antibody and anti-HBs antibodies (201 IU/ml) were both positive, while the HBs antigen was negative. The polymerase chain reaction for hepatitis B virus was negative (<49 copies/ml, <6 IU/ml, <0.78 logUI/ml). Hepatitis C virus serology was negative. Anti-SSA Ro52, anti-KU, and anti-OJ antibodies were positive. Anti-liver cytosol type 1 (LC1) and anti-soluble liver antigen (SLA) antibodies were positive in the liver dot (Euroimmun Kits). The immunoglobulin test showed a rise in IgG (21.6 g/l [normal range 8–12]). There were no abnormalities on the fibro scan. The diagnosis of AIH associated with DM was established. Mycophenolate mofetil (2g/d) and general corticosteroids (1 mg/kg/d) were administered. Liver function normalized after two months of treatment (ALT= 9 IU/l, AST=16 IU/l, GGT=49 IU/l). A liver biopsy was not considered necessary by gastrologists since the liver enzyme levels returned to normal rapidly after the treatment initiation. After a continued follow-up of 18 months, the patient is in complete remission with no relapse of either AIH or DM.

Discussion

This case report highlights a rare association between DM and AIH. Our patient was first diagnosed with DM. During the disease, increased transaminase levels can be related to flares of muscle inflammation.

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However, an increase in AST and ALT more than CK or associated cholestasis should point to liver damage. There has been evidence of a correlation between DM and viral hepatitis, particularly hepatitis B, but no clear proof of a cause-and-effect relationship has been found. Nevertheless, the theory that a viral component is involved in the etiology of DM remains viable.[2] In our case, the hepatitis B infection was resolved and could not account for the cytolysis and the increased level of GGT. Moreover, screening for other causes of liver damage was negative.

In systemic autoimmune diseases, liver damage can be viral, iatrogenic, or, less commonly, autoimmune. AIH is a rare auto-immune liver disease mainly affecting women around 40 years.[3,4] Its association with rheumatic diseases is frequently reported during systemic lupus erythematosus (0.7 to 2.8%), Sjogren's syndrome (1.4 to 35%), and rheumatoid arthritis (1.6 to 5.4%) (3). The coexistence of AIH and polymyositis was reported in five cases between 1985 and 2011.[5–9] They were all women, aged between 20 and 48 years at the time of diagnosis.

However, the association of DM and AIH seems to be exceptional. It was only reported in a single case of a 58-year-old woman diagnosed with simultaneous DM and AIH with good outcomes after corticosteroids and azathioprine.[1] Cytolysis with positive smooth muscle antibodies during DM was also reported in another case of a 19-year-old woman but the diagnosis of AIH was not formally maintained.[10]

In our case, AIH was confirmed due to liver test abnormalities and the positivity of specific antibodies and there was no need for liver biopsy.[11]

When it comes to the diagnosis and management of patients with inflammatory myopathy and AIH, autoantibodies are essential. To handle these complicated disorders, an integrated clinical approach is necessary. The preferred indicators of type 1 AIH are anti-SLA antibodies. These antibodies have a low prevalence but a high specificity for AIH.[12] They have been discovered more recently but are less common (15–20% of cases) and seem to be linked to a poor prognosis that includes more severe histology, a longer time to reach disease remission, a higher likelihood of relapse, the need for a liver transplant, and death.[13] These findings were not observed in a recent study where the positivity of anti-SLA antibodies was not correlated with distinct or severe clinical features in AIH. However, in this same study patients who tested positive for anti-SLA exhibited a faster response to immunosuppressive treatment.[14]

The positivity of Anti-LC1 antibodies is associated with AIH type II (30 and 50%). They are correlated with AIH activity and associated with adverse clinical outcomes and faster disease progression.[13,15] Autoantibodies have been reported more often during viral hepatitis C infection and less frequently during viral hepatitis B.[16]

Finally, our observation has multiple unique features: the patient's male gender, the development of AIH ten years after the diagnosis of DM and while it was in remission, the rarity of such a relationship involving nosological challenges, and lastly the coexistence of anti-SLA and anti LC1 antibodies. Furthermore, our patient had a long-lasting remission despite these low prognostic markers

Conclusion

We report an exceptional association of AIH to a DM. The rarity of this association implies questions as to possible links: genetic predisposition, autoimmunity disorders, viral infection triggers, or happenstance.

To shed light on these issues, it is interesting to report these uncommon correlations.

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References

1. de Souza FHC, Barros TBM, de Moraes MT, Missumi LS, Lima FR, Levy-Neto M, et al. [Autoimmune hepatitis and dermatomyositis: a rare association]. Acta Reumatol Port 2012;37(3):264-267. [Portuguese]
2. Zhang J, Wen XY, Gao RP. Hepatitis B virus-related liver cirrhosis complicated with dermatomyositis: A case report. World J Clin Cases 2019;7(10):1206-1212. [CrossRef]
3. Wang CR, Tsai HW. Autoimmune liver diseases in systemic rheumatic diseases. World J Gastroenterol 2022;28(23):2527-2545. [CrossRef]
4. Özaslan E, Günşar F, Çiftçibaşı Örmeci A, Hatemi İ, Efe C, Akyol G, et al. Diagnosis and treatment of autoimmune hepatitis: Questions, Answers, and illustrative cases: Endorsed by Autoimmune Liver Diseases Special Interest Group, Turkish Association for the Study of Liver. Turk J Gastroenterol Off J Turk Soc Gastroenterol 2023;34(Suppl 2):S1-S33. [CrossRef]
5. Bradley JD, Pinals RS, Gupta RC. Chronic active hepatitis associated with polymyositis. Association with precipitating mitochondrial M-B antibody. J Rheumatol 1985;12(2):368-371.
6. Ko KF, Ho T, Chan KW. Autoimmune chronic active hepatitis and polymyositis in a patient with myasthenia gravis and thymoma. J Neurol Neurosurg Psychiatry 1995;59(5):558-559. [CrossRef]
7. Marie I, Levesque H, Courtois H, François A, Riachi G. Polymyositis, cranial neuropathy, autoimmune hepatitis, and hepatitis C. Ann Rheum Dis 2000;59(10):839-840. [CrossRef]
8. Stefanidis I, Giannopoulou M, Liakopoulos V, Dovas S, Karasavvidou F, Zachou K, et al. A case of membranous nephropathy associated with Sjögren syndrome, polymyositis and autoimmune hepatitis. Clin Nephrol 2008;70(3):245-250. [CrossRef]
9. Kurihara Y, Shishido T, Oku K, Takamatsu M, Ishiguro H, Suzuki A, et al. Polymyositis associated with autoimmune hepatitis, primary biliary cirrhosis, and autoimmune thrombocytopenic purpura. Mod Rheumatol 2011;21(3):325-329. [CrossRef]
10. Quincey VA, Solanki KK, Lamont D, Rademaker M. Rare coexistence of dermatomyositis and smooth muscle antibodies, with abnormal liver function tests. N Z Med J 2016;129(1442):86-88.
11. Björnsson E, Talwalkar J, Treeprasertsuk S, Neuhauser M, Lindor K. Patients with typical laboratory features of autoimmune hepatitis rarely need a liver biopsy for diagnosis. Clin Gastroenterol Hepatol 2011;9(1):57-63. [CrossRef]
12. Efe C, Ozaslan E, Wahlin S, Purnak T, Muratori L, Quarneti C, et al. Antibodies to soluble liver antigen in patients with various liver diseases: A multicentre study. Liver Int 2013;33(2):190-196. [CrossRef]
13. Toh BH. Diagnostic autoantibodies for autoimmune liver diseases. Clin Transl Immunol 2017;6(5):e139. [CrossRef]
14. Yüksekyayla O, Kina N, Ulaba A, Emin Ergün M, Batibay E, Şimşek C, et al. The frequency and clinical significance of antibodies to soluble liver antigen/liver pancreas in autoimmune hepatitis: a prospective single-center study. Eur J Gastroenterol Hepatol 2024;36(5):652-656. [CrossRef]
15. Terziroli Beretta-Piccoli B, Mieli-Vergani G, Vergani D. Autoimmune Hepatitis: Serum Autoantibodies in Clinical Practice. Clin Rev Allergy Immunol 2022;63(2):124-137. [CrossRef]
16. Rabie RA, Hadhoud A, Abdelazim S, Radwan MI, Attia MH, Hasuna MA. Autoantibodies profile in autoimmune liver diseases and chronic viral hepatitis. Egypt J Immunol 2024;31(1):58-66. [CrossRef]