

Anti-synthetase syndrome with Kaposi sarcoma: An unusual association

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INTRODUCTION

Anti-synthetase syndrome (ASS) is an autoimmune condition characterized by the presence of autoantibodies directed against an aminoacyl transfer RNA synthetase (aaRS) including anti-Jo-1, anti-PL-7, anti-PL1-2, anti-EJ, anti-OJ, anti-KS, anti-Zo, anti-YRS/HA, along with clinical features that include interstitial lung disease (ILD), myositis, Raynaud's phenomenon, fever, mechanic's hands, and arthritis.

Unlike adult-onset polymyositis and dermatomyositis, which may represent paraneoplastic manifestations of cancer, ASS is generally deemed not to be associated with malignancy in adults.

only tow of association of ASSD with Kaposi sarcoma have been reported in the literature , herein we report a rare case of this rare association.

Herein we present a case of a 64-year-old woman diagnosed with ASSD and who was found to have Kaposi's disease as a rare association.

CASE REPORT

A 64-year-old woman with a history of diabetes and ischemic heart disease for 3 years presented since 6 months an anti-synthetase syndrome revealed by fever, arthritis, polymyositis, diffuse interstitial lung disease found in Computed tomography scan with Laboratory investigations revealed elevated creatine phosphokinase (CPK) and the presence of auto-antibodies anti Jo1 and anti R052. No treatment has been taken by the patient in recent months.

The patient consulted in our dermatology department for recent-onset painful and itchy skin lesions

The physical examination found angiomatous nodules in the left sub-palpebral region, on the toes, with erythematous-violet macules on the face, trunk and lower limbs, as well as a few purple patches on the chin, ears, palmar-plantar region and in the intergluteal fold, associated with involvement of the oral mucosa (intraoral purple macules and nodules) Figure1-2

Histological assessment of skin biopsy of a sole lesion was indicative of Kaposi sarcoma.

Human immunodeficiency virus 1/2 antibodies were negative.

Investigations in search of other organ involvement showed hematological involvement in the form of pancytopenia, neurological involvement in the form of axonal sensory neuropathy, and digestive involvement in the form of gastric kaposi nodules, as assessed by endoscopy-guided biopsies. A taxane-based chemotherapy was indicated, and the patient died one week after her first session.



Figure 2: Kaposi's lesions on feet

DISCUSSION

Kaposi sarcoma (KS) is a rare angioproliferative tumor whose etiology is associated with human herpesvirus 8 (HHV 8). KS lesions typically involve the skin or mucosal surfaces and are characterized by purplish, red-blue, or brown-black macules, papules, and nodules which are prone to bleeding and ulceration. Definitive diagnosis requires biopsy revealing characteristic angioproliferative features. There are five widely recognized types of KS, which are histologically indistinguishable but differ in epidemiology and prognosis. These include classic, endemic, iatrogenic, epidemic and nonepidemic KS was rarely reported in association with ASS how was defined as a distinct clinical entity for the first time in 1990 by Marguerie et al, as the presence of idiopathic inflammatory myositis, interstitial lung disease (ILD) and other clinical manifestations such as arthritis, Raynaud phenomenon, keratoconjunctivitis sicca, and subcutaneous calcinosis in patients with anti-ARS autoantibodies . Since then, ASS has been considered a subtype of idiopathic inflammatory myopathy (IIM) due to being closely associated with myositis and myositis associated ILD, arthritis, and other clinical features Unlike the other types of idiopathic inflammatory myopathy (IIM) which may represent paraneoplastic manifestations of cancer, ASS is generally deemed not to be associated with malignancy in adults.

The presence of anti-Jo1 antibodies was associated with a higher risk of cancer and ASS patients with both anti-Jo1 and anti-Ro52 antibodies are more prone to cancer than those with anti-Jo1 antibodies alone highlighting the significance of anti-Ro52 antibodies in ASS patients with cancer.(13) our patient presented ASS with both anti jo1 and anti R052 antibodies

The first case of KS occurred in an ASS was reported by Laura .B at 2013 , after 2 months of treatment by glucocorticoid Therefore, glucocorticoid-induced KS could not be ruled out. Sellitto et al reported a case of KS with ASS paraneoplastic syndrome in which ASS was stable during chemotherapy and relapsed after chemotherapy discontinuation. Nan He et al reported a case ASS) complicated with KS 2 months after being diagnosed with ASS. Our patient diagnosed with KS 6 months after ASSD and she didn't receive any immunosuppressive treatment before skin lesions appearance. Sufficient understanding of the pathophysiological relationship between ASS and KS is lacking. Further studies and cases series are needed to fully understand this association.

CONCLUSION

The association between KS and ASS is rarely reported. A preventive cancer screening and follow-up including dermatological examination should be recommended for ASS patients especially those treated with immunosuppressant drugs



Figure 1: Kaposi's lesions on the face (A) and in intraoral (B)