Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Dermatology

Anti- Synthetase Syndrome with Kaposi Sarcoma: An Unusual Association

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DOI: 10.36347/sjmcr.2024.v12i03.012 | **Received:** 06.02.2023 | **Accepted:** 13.03.2024 | **Published:** 17.03.2024

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Abstract **Case Report**

Kaposi sarcoma (KS) is a rare angioproliferative tumor whose etiology is associated with human herpesvirus 8 and it is widely known as a complication of acquired immunodeficiency syndrome and, the assocciation with anti synthetase syndrome (ASS) have rarely been reported. Herein we present a case of a 64-year-old woman diagnosed with ASS and Kaposi's disease as a rare association.

Keywords: Anti-synthetase syndrome. Kaposi sarcoma, paraneoplastic.

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 [1-4].

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 [5, 6].

Only tow of association of ASSD with Kaposi sarcoma have been reported in the literature [7], 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 eryhtemato-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) Figure 1 & 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.

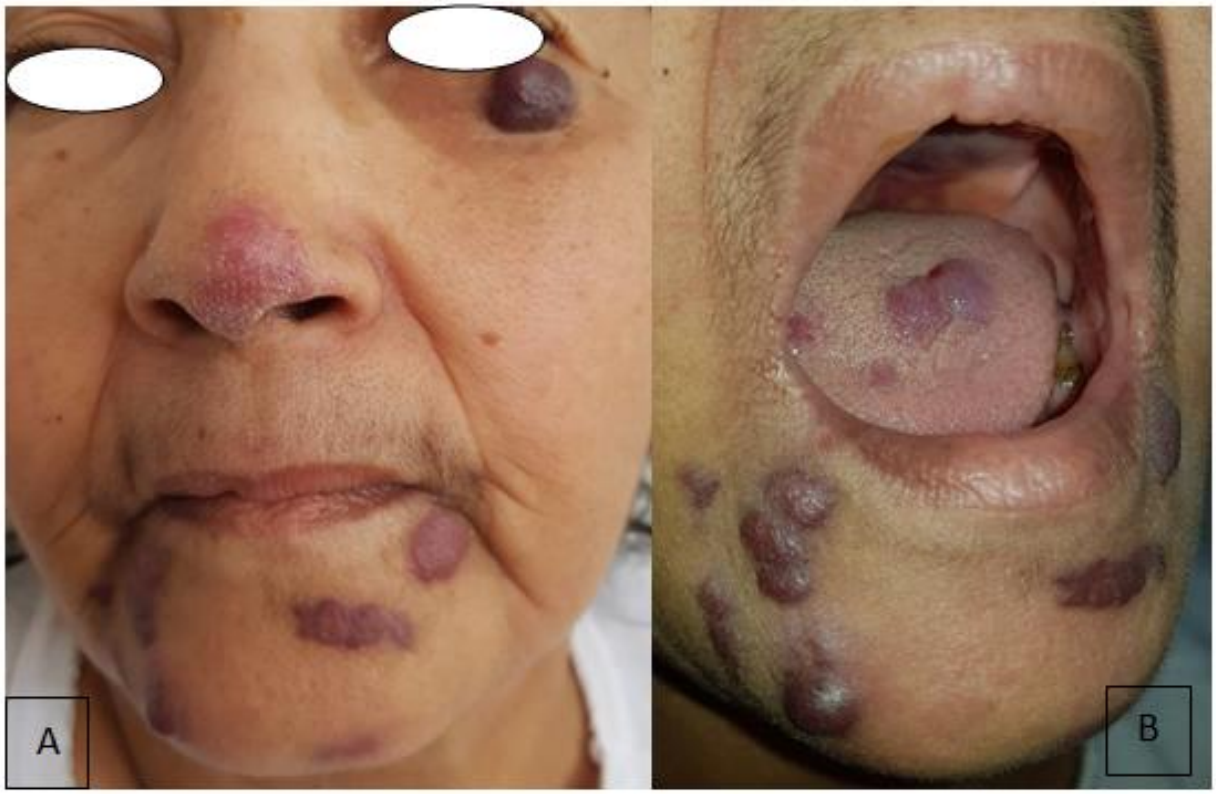

Figure 1: Kaposi's lesions on the face (A) and in intraoral (B)

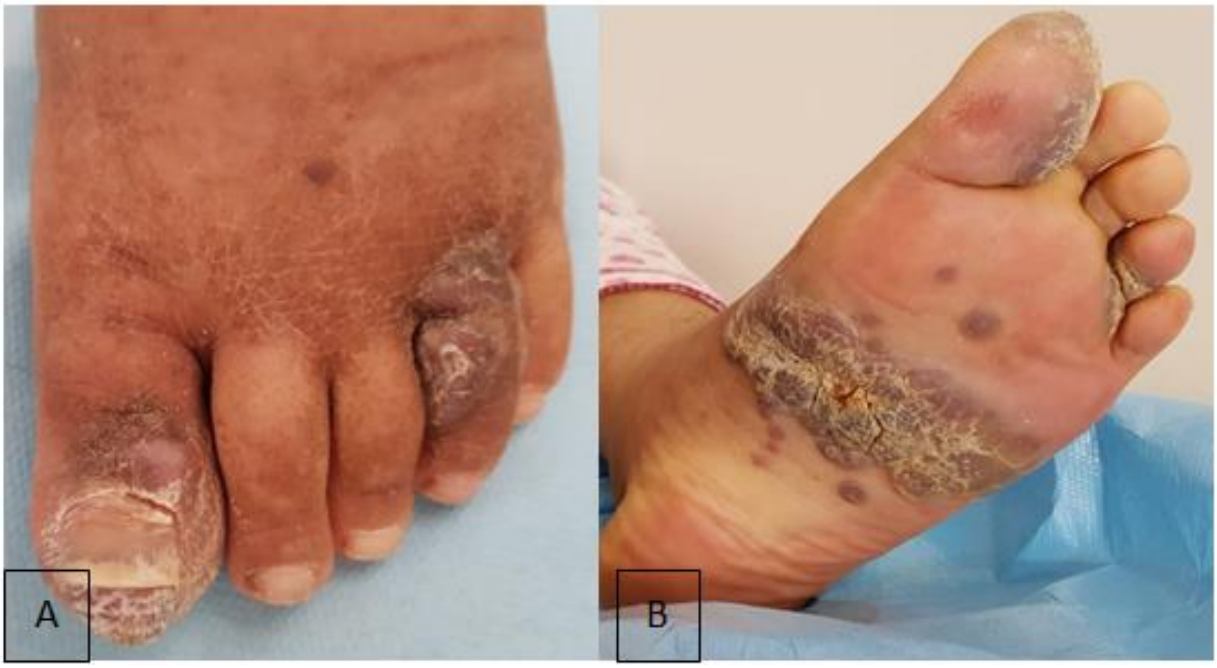

Figure 2: Kaposi's lesions on feet

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 [8].

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 [9]. 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 [10].

ASS has an annual incidence of approximately 0.6/100 000 [11]. 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 [12].

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 treatement by glucocorticoid Therefore, glucocorticoid-induced KS could not be ruled out [14].

Sellitto *et al*., reported a case of KS with ASS paraneoplastic syndrome in which ASS was stable during chemotherapy and relapsed after chemotherapy discontinuation [7].

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 [15].

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

Funding: None

Conflicts of Interest: The authors declare no conflicts of interest.

REFERENCES

1. Mirrakhimov, A. E. (2015). Antisynthetase syndrome: a review of etiopathogenesis, diagnosis and management. *Current medicinal chemistry*, *22*(16), 1963-1975.
2. Connors, G. R., Christopher-Stine, L., Oddis, C. V., & Danoff, S. K. (2010). Interstitial lung disease associated with the idiopathic inflammatory myopathies: what progress has been made in the past 35 years?. *Chest*, *138*(6), 1464-1474.
3. Lega, J. C., Fabien, N., Reynaud, Q., Durieu, I., Durupt, S., Dutertre, M., ... & Cottin, V. (2014). The clinical phenotype associated with myositis-specific and associated autoantibodies: a meta-analysis revisiting the so-called antisynthetase syndrome. *Autoimmunity reviews*, *13*(9), 883-891.
4. Witt, L. J., Curran, J. J., & Strek, M. E. (2016). The diagnosis and treatment of antisynthetase syndrome. *Clinical pulmonary medicine*, *23*(5), 218-226. doi: 10.1097/CPM.0000000000000171
5. Galindo-Feria, A. S., Notarnicola, A., Lundberg, I. E., & Horuluoglu, B. (2022). Aminoacyl-tRNA synthetases: on anti-synthetase syndrome and beyond. *Frontiers in Immunology*, *13*, 866087. doi: 10.3389/fimmu.2022.866087. PMID: 35634293; PMCID: PMC9136399.
6. Zanframundo, G., Faghihi-Kashani, S., Scirè, C. A., Bonella, F., Corte, T. J., Doyle, T. J., ... & Cavagna, L. (2022). Defining anti-synthetase syndrome: a systematic literature review. *Clinical and experimental rheumatology*, *40*(2), 309-319. doi: 10.55563/clinexprheumatol/8xj0b9. Epub 2022 Feb 25. PMID: 35225224; PMCID: PMC10411341
7. Sellitto, A., Adinolfi, L. E., Romano, C., Iovino, F., Auriemma, P. P., Russo, D., & de Vita, F. (2018). Plantar Kaposi sarcoma revealed by antisynthetase syndrome. *JCR: Journal of Clinical Rheumatology*, *24*(5), 281-285. doi: 10.1097/RHU.0000000000000650. PMID: 29239932.
8. Vangipuram, R., & Tyring, S. K. (2019). Epidemiology of Kaposi sarcoma: review and description of the nonepidemic variant. *International Journal of Dermatology*, *58*(5), 538-542. doi: 10.1111/ijd.14080. Epub 2018 Jun 11. PMID: 29888407.
9. Marguerie, C., Bunn, C. C., Beynon, H. L. C., Bernstein, R. M., Hughes, J. M. B., So, A. K., & Walport, M. J. (1990). Polymyositis, pulmonary fibrosis and autoantibodies to aminoacyl-tRNA synthetase enzymes. *QJM: An International Journal of Medicine*, *77*(1), 1019-1038.
10. Dalakas, M. C., & Hohlfeld, R. (2003). Polymyositis and dermatomyositis. *The Lancet*, *362*(9388), 971-982.
11. Mirrakhimov, A. E. (2015). Antisynthetase syndrome: a review of etiopathogenesis, diagnosis and management. *Current medicinal chemistry*, *22*(16), 1963-1975.

12. Olazagasti, J. M., Baez, P. J., Wetter, D. A., & Ernste, F. C. (2015). Cancer risk in dermatomyositis: a meta-analysis of cohort studies. *American journal of clinical dermatology*, *16*, 89-98.
13. Marie, I., Hatron, P. Y., Dominique, S., Cherin, P., Mouthon, L., Menard, J. F., ... & Jouen, F. (2012, June). Short-term and long-term outcome of anti-Jo1-positive patients with anti-Ro52 antibody. In *Seminars in arthritis and rheumatism* (Vol. 41, No. 6, pp. 890-899). WB Saunders.
14. Bragado, L., Gutierrez, L. R., Cuende, E., & Gonzalez, J. L. L. (2013). Kaposi's Sarcoma in a Steroid-treated Antisynthetase Antibody Syndrome Patient. *Reumatología Clínica (English Edition)*, *9*(4), 243-245.
15. He, N., Lu, D., Xue, J., & Wang, X. (2020). Anti-alanyl tRNA positive antisynthase syndrome with Kaposi sarcoma. *International Journal of Rheumatic Diseases*, *23*(6), 828-832. https://doi.org/10.1111/1756-185X.13861