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Anti-RO/SS-A Antibody-Associated Autoimmune Vasculitis: Case Report

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***Abstract**: Anti-Ro/SSA antibodies are among the most frequently identified autoantibodies against extractable nuclear antigens and have been related with systemic lupus erythematosus (SLE) and Sjögren's syndrome (SS). However, these autoantibodies are also sometimes detected in other systemic autoimmune diseases. In the past, the knowledge of the prevalence of anti-Ro/SSA antibodies in various autoimmune diseases and symptoms has been expanded, and clinical importance of these autoantibodies is increasing. Their presence is associated with serologic hyperactivity, vasculitis and nervous system involvement. The aim of this study was to describe a case of anti-Ro/SSA antibodies positive autoimmune vasculitis, a rare and severe medical condition.*

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I. Background

Systemic autoimmune diseases are a category of medical disorders that affects various organs and are related to autoimmune responses. These are usually characterized by the development of autoantibodies against intracellular autoantigens. In fact, diagnosis, classification, and prognosis frequently rely on specificity and levels of the autoantibodies, in addition to clinical presentations and other laboratory evaluations. Among autoantigens, extractable nuclear antigens (ENA) are soluble cytoplasmic and nuclear components with more than 100 different antigens described [1]. Anti-Ro/SSA can be identified in 70-100% in SS [2] and 40-50% of patients with SLE [3]. These antibodies were originally described in 1961 as two precipitating antibodies reacting with antigens contained in extracts from lacrimal and salivary glands of patients with SS, termed SjT, and, SjDrespectively [4]. The authors named the antibody 'anti-Ro antibody' after the original patient in whom the antibodies were identified [5]. Anti-Ro antibodies bind to numerous intracellular proteins, predominantly the 52 and 60 kD Ro antigens, which varies in terms of their antigenicity and structure [6,7]. The mechanisms underlying both the induction and regulation of the immune response against the various Ro polypeptides are still uncertain [8]. Systemic vasculitis has been a diagnostic challenge in the areas of rheumatologyand clinical medicine for many years [9]. The diagnostic criteria for autoimmune vasculitis are a combination of radiologic, clinical, laboratory, and histopathologic findings.

II. Case Presentation

A 48-year-old male referred with a history of increasing joints pain, joint swelling and multiple small to large hemorrhagic bullae since 1 week prior. The patient had a history of alcohol consumption and smoking but did not have any history of drug abuse, previous liver and biliary or hemorrhagic diseases, transfusion, high risk sexual behavior, and recent travel to high risk regions for infectious diseases. The hemorrhagic bullae are mostly in lower and upper extremities, buttock regions and a few in head & neck and genital areas. It is associated with both knee and ankle joints swelling. There was no evidence of fever, diarrhea, constipation, ascites, hematemesis, lymphadenopathy, hepatosplenomegaly, or weight loss.

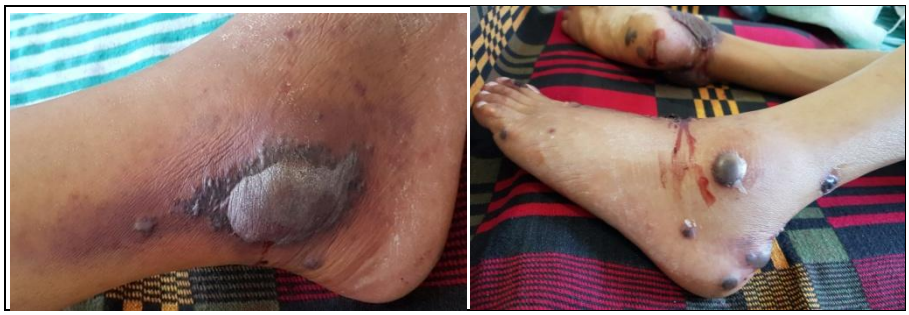

Fig. 1 Bullous vasculitis in lower limbs

III. Investigations

Patient's laboratory investigations and their results are shown in the Table 1. Skin biopsy result showed infected hemorrhagic bullous dermatitis.

Table 1. Laboratory data.

Blood index	**Case**	**Normal range**	**Blood index**	**Case**	**Normal range**	**Blood index**	**Case**	**Normal range**
Anti-CCP	<7	<17 U/ml	ANA	Negative		Anti-HBc	Non-reactive	
RA (Quantitative)	10	≤14 IU/ml	ANCA	Negative		Potassium	4.9	3.6-5 mmol/L
Tacrolimus	0.81	3-20 ng/ml	Anti-AMA-M2	Negative		Creatinine	1.5	0.7-1.2 mg/dL
Anti-dsDNA	Negative		Anti-M2-3E	Negative		Alkaline phosphatase	229	38-126 U/L
Anti-Nucleosome	Negative		Anti-LKM	Negative		eGFR	55	>60 ml/min
Anti-Histone	Negative		Anti-LC	Negative		ALT/SGPT	452	<41 U/L
Anti-SS-A	Positive		Anti-SLA/LP	Negative		AST/SGOT	203	<40 U/L
Anti-Ro52	Positive		Anti-PGDH	Negative		Bilirubin (Total)	1.1	0.1-1.4 mg/dL
Anti-RNP/Sm	Negative		Anti-RP11	Negative		ESR	63	<15 mm/1sthr
Anti-Sm	Negative		Anti-RP155	Negative		WBC	16.74x10^{3}	4-11x10^{3}/uL
Anti-Mi-2 alpha	Negative		Anti-gp210	Negative		RBC	4.30x10^{6}	3.8-5.8x10^{6}/uL
Anti-Mi-2 beta	Negative		PCNA	Negative		HGB	11.1	12-17g/dL
Anti-Ku	Negative		Anti-DFS70	Negative		PLT	446	150-400x10^{3}/uL
Anti-Centromere A	Negative		HIV	Non-reactive		Blood culture	Sterile	
Anti-Centromere B	Negative		Anti-HCV	Non-reactive		PT	11.0	10-14 sec
Anti-Sp100	Negative		HBsAg	Non-reactive		INR	0.87	
Anti-PML	Negative		Anti-HBs	Non-reactive		Procalcitonin	0.116	<0.05 ng/ml
Anti-Scl-70	Negative		HBeAg	Non-reactive		CRP	29.02	<5 mg/L
Anti-PM-Scl-100	Negative		Anti-HBe	Non-reactive				
Anti-PM-Scl-75	Negative		Serum protein electrophoresis	Decreased albumin Increased α2, β2 & r globulins				
Urine index	**Case**	**Normal range**	**Urine index**	**Case**	**Normal range**	**Urine index**	**Case**	**Normal range**
SG	1.025	1.003-1.030	Glucose	Normal		Bil	17	umol/L
pH	5.0	4.5-8.0	Ketone	0.5 mmol/L		Random urine protein	739	<150 mg/L
Leu	25	0-4/ul	UBG	17 umol/L		Creatinine	378.46	40-287 mg/dL
Protein	0.25 g/L		Ery	0-4/ul		Urine Protein:Creat-inine	0.19	<0.2 g/g

Anti-CCP=Anti-cyclic citrullinated peptide; RA=Rheumatoid arthritis; Anti-dsDNA=Anti double-stranded DNA; Anti-RNP/Sm=anti-small nuclear ribonucleoproteins; Anti-Sm=Anti Smith antibody; ANA=Antinuclear antibody; ANCA=Anti-neutrophilic cytoplasmic antibody; Anti-AMA-M2=Anti Mitochondrial M2 antibody; Anti-LKM=Antibody to Liver Kidney Microsome; Anti-LC=Antibody to Liver Cytosol; Anti-SLA/LP=Anti-soluble liver antigens/liver-pancreas antibodies; Anti-PGDH=Anti-Prostaglandin Dehydrogenase; Anti-RP11=Anti-rabbit Prestige 11; Anti-RP155=Anti-rabbit Prestige 155; Anti-gp210=Anti-Glycoprotein 210; PCNA=Polyclonal antibody; Anti-DFS70=Anti-dense fine speckled 70; HIV=Human immunodeficiency virus; Anti-HCV=Antibody against hepatitis C virus; HBsAg=Hepatitis B surface antigen; Anti-HBs=Antibody against hepatitis B surface antigen; HBeAg=Hepatitis B e-antigen; Anti-HBe=Antibody against hepatitis B e-antigen; Anti-HBc=Antibody against hepatitis B core-antigen; eGFR=Estimated Glomerular Filtration Rate; ALT=Alanine aminotransferase, AST=Aspartate aminotransferase; ESR=Erythrocyte sedimentation rate; WBC=White blood cell; RBC=Red blood cell; HGB=Haemoglobin; PLT=Platelet; PT=Prothrombin time; INR=International Normalised Ratio; CRP=C-Reactive Protein; SG=Specific Gravity; Leu=Leukocyte; UBG=Urobilinogen, Ery=Erythrocytes; Bil=Bilirubin

IV. Discussion

The clinical, serologic, andhematologic features associated with autoantibodies to the small molecular weight ribonucleoproteins Ro (SS-A) were discovered. A prominent clinical relationship of anti-Ro (SS-A) antibodies with extraglandular disease (purpura, vasculitis, and lymphadenopathy) was seen. hematologic abnormalities (anemia, leukopenia, and thrombocytopenia) were also associated with the presence of anti-Ro (SS-A) antibodies. Furthermore, anti-Ro (SS-A) antibody was associated with increased serologic reactivity in terms of rheumatoid and antinuclear factors, hyperglobulinemia, cryoglobulinemia, and hypocomplementemia. The presence of anti-Ro (SS-A) antibodies defines a subset of patients with Sjogren's syndrome who have systemic clinical manifestations comprising vasculitis, hematologic abnormalities, and serologic hyperreactivity [6]. Anti-Ro antibodies have been used as a valuable diagnostic marker for SLE and SS, they are the most prevalent autoantibodies among various autoimmune diseases [1]. Inflammatory vascular disease also is a progressively recognized complication occurring in nearly one-third of the SS patients [6]. Although the pathogenic role of autoantibodies in autoimmune disease has not yet been clarified, hypotheses have been put forward showing that anti-Ro antibodies might have a direct role in damaging tissues. Anti-Ro52 antibodies may have pathological roles not only by damaging tissues directly but also by inhibiting the activity of Ro52 antigens. Further investigations into the Ro autoantigen-autoantibody system may offer a new policy for treating autoimmune diseases [1].

V. Conclusion

Despite being a rare condition, it is important to be aware of the development of autoimmune vasculitis in the clinical course so the appropriate treatment can be promptly instituted, thus avoiding the progression of lesions and possible associated complications.

Author's contribution

SS was principal author who conceptualized the manuscript, wrote background and methods sections, results and discussion sections of the manuscript, interpretation of the results, and revised the final drafts of the manuscript. **OHK** assisted in conceptualization of the manuscript.

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