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## RESEARCH ARTICLE

### A RARE CASE OF OVERLAP SYNDROME.

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#### Abstract

A 20 year female presented with features of overlap syndrome constituting SLE and Wegener's Granulomatosis (Granulomatosis with polyangitis). She was admitted with findings of SLE including skin lesions, joint pains, renal manifestations and laboratory evidence. On further suspicion and features of sinusitis, lower respiratory tract involvement, ear involvement, we further investigated which pointed towards WG. Together we finally arrived at a diagnosis of Overlap syndrome.

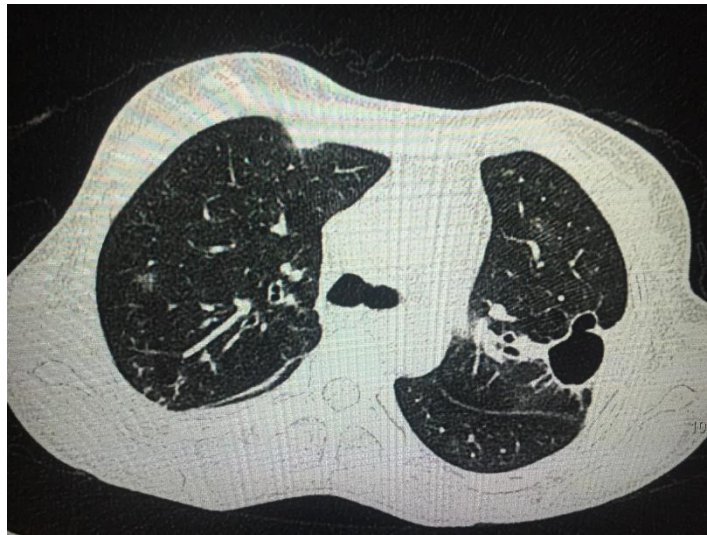
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#### Case report:-

A 20 year old girl presented to Emergency with complaints of fever and joint pains since 15 days and rash since 1 week. History revealed low grade continuous fever since 15 days without chills and rigors, which was associated with pain, redness and swelling of small and large joints. After 1 week of fever there appeared rash over the body and then ulcers appeared over the buccal mucosa and over the lips with difficulty in opening the mouth and difficulty in swallowing and ulcers in the nasal cavity. Severe headache and facial pain. Cough since 5 days with scanty mucoid to mucopurulent sputum without blood. On the day of admission she also had complaints of hearing loss and pain in both ears without any ear discharge. Weight loss, malaise and hair loss since the onset of illness. No significant menstrual, past, family and personal history.



On examination she was ill built and nourished, temperature-100F, PR-120/min, BP-100/70mmhg, RR-20/min. Pallor present. Multiple diffuse hyperkeratotic hemorrhagic crusting with few erosions distributed in reticular pattern on sun exposed areas of face and trunk and also unexposed areas of trunk. Erythematous lesions distributed in livedoid pattern are seen over both thighs. Multiple well defined bleeding ulcers are seen over lips and covered with purulent discharge are seen over buccal mucosa. Hypertrophic and bleeding gums are seen. Tongue showed whitish coating. Nasal cavity examination showed ulcers over the septum and lateral wall. Fundus examination showed optic disc vasculitis. Ear examination showed bilateral normal EOC and tympanic membrane intact. All major and minor joints appear to be mildly swollen and are tender on palpation. RS-NVBS with occasional rhonchi and crepts. Rest of the systemic examination was normal.



Blood picture showed anemia with neutrophilic leukocytosis, ESR-high, serum creatinine and blood urea increased. Urine routine examination showed albumin and granular casts, 24 hour urinary protein showed significant proteinuria. Chest X ray- evidence of round to oval well defined thin walled cavity noted in the left midzone with tiny cystic lesions lucencies. Sputum AFB and Bronchial washing AFB are negative. CRP reactive, RA factor nonreactive, ANA strongly positive, Anti ds DNA strongly positive, C ANCA strongly positive with high titers, P ANCA slightly above normal. CT chest – evidence of well defined round to oval cavity with thick wall noted involving apical segment of right upper lobe and apico-posterior segment of left upper lobe suggestive of Wegener's granulomatosis. PTA-bilateral mixed moderate hearing loss. Renal biopsy showed normal study with no immune deposits suggesting pauci immune glomerulonephritis.

All these together satisfy SLICC 2012 criteria for SLE and ACR 1990 criteria for GPA so diagnosed as Overlap syndrome. She was given glucocorticoids with which she showed some signs of improvement. Cyclophosphamide was given in pulse doses. There was good recovery.

### **Discussion:-**

SLE is a heterogeneous group of autoimmune diseases with a diverse array of clinical manifestations ranging from localized skin lesions to life threatening systemic manifestations associated with antibodies directed against cell nuclei. Diagnosis of SLE is by any 4 (1 in each category) of systemic lupus international collaborative criteria 2012.

GPA(Wegener's) is a autoimmune systemic necrotizing vasculitis of small arteries and veins with granulomas. It is characterized by granulomatous vasculitis of upper and lower respiratory tracts together with glomerulonephritis. Antibodies are against antiprotein-3 in some against antimyeloeroxidase-3. Diagnosis of GPA is by atleast 2 of 4 ACR 1990 criteria.

This case satisfies criteria for both SLE and GPA. Very few cases exist in combination in literature. It can be termed as overlap syndrome.

### **Conclusion:-**

Though SLE is the most common autoimmune disease, careful clinical examination, suspicion and proper laboratory investigations will guide us to detect other associated autoimmune diseases. Overlap of SLE and GPA is rare and this is one of such rare cases.

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