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CUTANEOUS MANIFESTATIONS AS THE KEY TO DIAGNOSIS OF CHURG STRAUSS SYNDROME: A CASE REPORT

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Abstract
Churg-Strauss syndrome is a rare systemic vasculitis characterized by asthma and other allergy symptoms as well as eosinophilia and necrotizing vasculitis involving small and medium-sized vessels. Its prevalence in the general population ranges from 1-3 cases per million a year, varying according to the population studied. The authors describe a case of a male patient affected by the disease with important systemic manifestations and not very florid skin lesions.
Keywords: Skin manifestations; Churg-Strauss Syndrome; Vasculitis

Introduction

Churg-Strauss syndrome (CSS), or eosinophilic granulomatosis with polyangiitis, is a rare systemic vasculitis characterized by asthma and other allergic symptoms, besides eosinophilia and necrotizing vasculitis of small and medium vessels.[1]The prevalence in the general population is of 1-3 cases/million, and varies according to the population studied. The condition affects both sexes with the same frequency, in the third and fifth decades of life.[2]

Case Report:

68 yr male resident of panipat, ex handloom worker by occupation, came to general medicine opd. Patient was apparently well 5 months back when he developed c/o fever which was of low grade ,intermittent in nature, no specific diurnal variation relieved on taking antipyretic medication, associated, with sweating, not associated with chills and rigors. He had also c/o post nasal drip, increased by coldweather, dust, perfumes etc., a/w headache. He had c/o breathlessness ,mMRCdysnea scale grade I,relieved on nebuliser, injhydrocort and tbderiphylline.He had also c/o multiple red spots on bilateral upper and lower limbsfrom last 3 months, which wereinsidious in onset, progressive in nature, occurred after every episode of fever, red bluish in colored, nonitching, non ulcerative, nodular and self resolved within 3-5 days.No history of similar episode in the past. Known case of allergic rhinitis.Known case of Asthma on MDI with spacer budacort, salbutamol and tbderiphyllin, Known case of treated old pulmonary kochs , ATT taken for 6 months , 25 years back, Not a known case of diabetes and hypertension..

On general physical examination- patient was conciuos oriented to time place person and hemodynamical stable. On local examination-multiple erythmatous plaques on bilateral upper and lower limbs, 10-12 mm in size, non tender, palpable, non blanchable, non ulcerative, reddish, blue colored. Pallor icterus cyanosis clubbing and lymphadenopathy were absent. Bilateral intermittent rhonchi present. Rest systemic examination was normal.

Comletehemogram total eosinophil count was 1700 per cubic mm, 20%. Renal function test, liver function test, lipid profile werewith in normal limit. ESR was 96mmin first hour. Mountox test test was 18 mm induration positive. CRP POSITIVE. RA factor was negative. Stool for ova cyst negative. Viral marker negative. Urine complete examination was normal.ANA by ELISA –negative. C-ANCA-Neg, P-ANCA-Neg, Serum filarial antigen-Not detected, S. Total IgE-1653 IU/ml (0-100 IU/ml), S. Aspergillusfumigatus antibody-0.19 kua/l (<0.35), PBF for microfilaria-neg, widal test-neg, IgM/IgG TYPHI dot-neg, PBF for MP-Neg, Blood c/s-sterile, Urine c/s-sterile. ECG, 2-D Echo and ultrasound abdomen and pelvis were normal.

On Skin biopsy- Dermis shows diffuse dense ,periadnexal andperivascular inflammatory infiltrates composedof eosinophils admixed with few neutrophils andlymphocytes. Although no granuloma are seen,Histopathological features are closer to the clincaldiagnosis of EGPA(figure 1).

X ray PNS VIEW- maxillary sinusitis and CT PNS-right frontal and left maxillary mucosal thickening suggestive

sinusitis (figure 2). CECT CHEST – brochiectasis changes and bullae in left lobe.

CECT ABDOMEN –Revealed no significant abnormalities. Pulmonary function test suggestive severe obstructive airway disease.

Nerve conduction velocity suggested pure motor neuropathy involving bilateral lower limb and upper limb (with possibility demyelinating disease).

Points favour to churgstrauss syndrome are significant eosinophilia, allergic rhinitis, sinusitis, k/c/o bronchial asthma, severe obstructive airway disease, neuropathy and skin biopsy.

Patient was treated with inj methyl prednisolone 500mg iv od for 3 days and tbprednisolone 50mg od 1month and tapered slowly and Tb azathioprine 50mg on follow up. Meter dose inhaler was used for sever obstructive disease.

On follow up fever, rashes and eosinophilia were improved within a week.

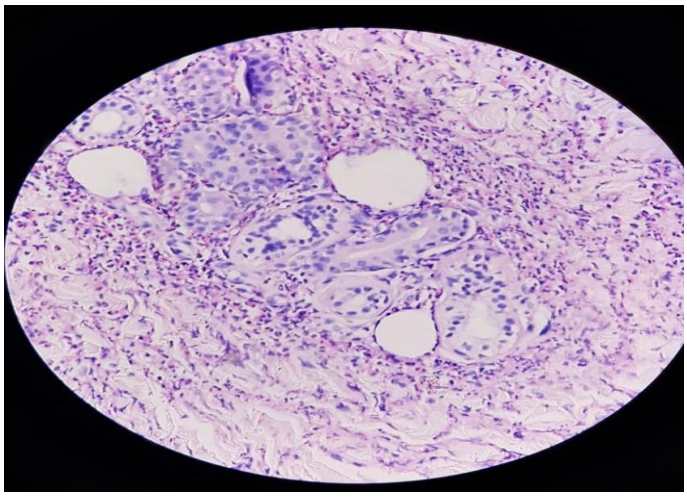

Figure 1: On Skin biopsy- Dermis shows diffuse dense, periadnexal and perivascular inflammatory infiltrates composed of eosinophils admixed with few neutrophils and lymphocytes. Although no granuloma are seen, Histopathological features are closer to the clincal diagnosis of EGPA

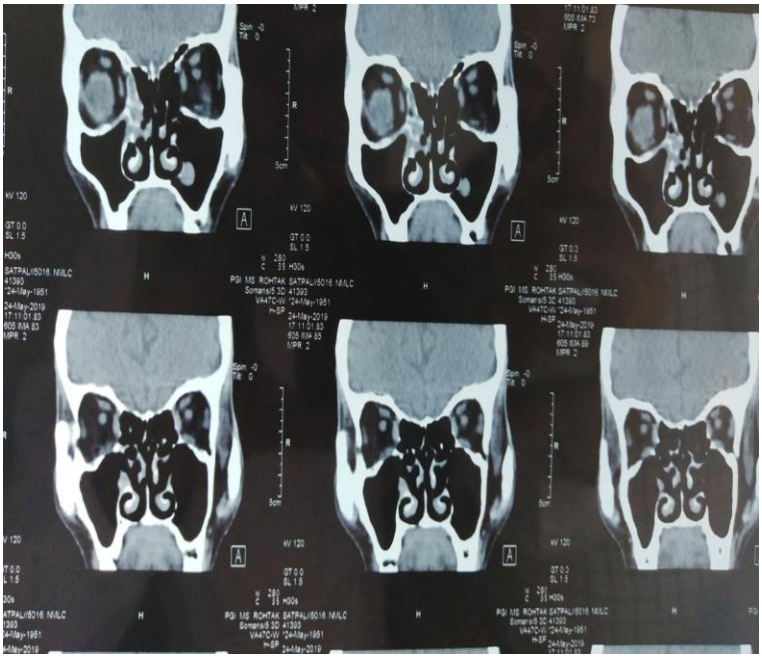

Figure 2: CT PNS-right frontal and left maxillary mucosal thickening suggestive sinusitis

Discussion

CSS is characterized by the onset of vasculitis, that manifests as multiple mononeuropathy, purpura, general symptoms and eosinophilia in a patient with previous asthma. However, some patients can develop asthma or eosinophilia and vasculitis simultaneously.[1]The initial and most common manifestations of CSS include the involvement of the respiratory tract with asthma and allergic rhinosinusitis, with possible nasal polyps, pulmonary infiltrates and pleural effusion. Peripheral neuropathy, manifested by multiple mononeuropathy, occurs in a third of the cases. Renal involvement affects about 30% of patients and can cause urinary abnormalities, including rapidly progressive renal failure. There is gastrointestinal involvement in a third of patients, that can present with abdominal pain, nausea, vomits, diarrhea, intestinal bleeding and acute abdomen.[3] Cardiovascular system is affected in about 10% of cases and this involvement is the primary cause of death in patients with CSS. Among the most important manifestations are coronary arteritis and myocarditis.[4]Although rare, central nervous system involvement is the major cause of mortality among patients.[3] About 50% of patients will present with skin changes. These include: 1) hemorrhagic lesions, such as palpable purpuras, petechiae, ecchymosis and hemorrhagic bullae, and 2) dermal or subcutaneous papules and nodules, frequently located on the scalp or distributed bilaterally over the extensor surfaces of the extremities. Other changes that can also be observed less frequently are urticaria, erythematousmacules and livedo reticularis.[5] CSS has three evolutionary phases. In the first phase, that can last years or decades, only respiratory manifestations such as asthma, rhinitis and nasal polyps occur. In the second phase, there is peripheral and tissue eosinophilia, affecting primarily the lungs, intestines and myocardium. In the third phase, there is systemic vasculitis affecting nerves, kidneys and skin.[3]Despite the mild cutaneous manifestations of the patient, there were four undisputable diagnostic criteria present: history of asthma, eosinophilia, neuropathy and vascular eosinophilic infiltrate on the histology. Besides, p-ANCA was positive, making it possible to diagnose the third phase of CSS. CSS prognosis is variable and depends on the initial extension of the disease and the organs affected. Five factors were described that are associated to a higher mortality among patients; which are: creatinine higher than 1.58mg/dL, proteinuria higher than 1 g/day, gastrointestinal involvement, central nervous system involvement and cardiomyopathy.[6] These criteria help identify the patients that need a more aggressive immunosuppressive therapy, due to the higher chance of mortality and higher frequency of relapse. Conventional treatment includes the use of corticosteroids. Cutaneous manifestations of CSS are one type of clinical presentation for this condition. Recognition of these lesions by the dermatologist was essential for the clinical suspicion and confirmation of diagnosis, that allowed adequate treatment, reducing morbidity and contributing for the prevention of irreversible lesions in vital organs.[7]

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