

Churg Strauss Syndrome with Migratory Polyarthrititis

Arnon Blum* and Amal Arabi

Department of Medicine; Baruch Padeh Poria Hospital Bar Ilan University, Lower Galilee 15208, Israel

Abstract

Churg Strauss Syndrome (CSS) is a rare systemic vasculitis associated with asthma, eosinophilia, sinusitis and pulmonary infiltrates. A 49 years old woman was admitted with migratory polyarthrititis and Churg Strauss Syndrome. We will discuss the findings that lead to the diagnosis of CSS and we will raise the possibility that she might have a new syndrome that includes CSS with migratory polyarthrititis. Until now there have no reports describing patients with CSS and arthritis. There have been some cases with arthralgia, but our case represents the first case that was admitted with arthritis. This case is a novel case describing a new combination of migratory arthritis in a patient suffering from CSS.

Case Report

A 49 years old woman has severe asthma for the last 30 years with daily asthmatic attacks. Two years ago she started treatment with Omalizumab (bi-monthly) due to the severe asthmatic attacks with a significant clinical improvement. She was admitted with a history of 3 weeks' migratory arthrititis involving the wrists in both arms. Every few days a new joint became painful, red and swollen. The migratory polyarthrititis was located in both wrists (each arm separately, never at the same time in both arms and legs) and ankles with sensory loss in both feet. The involved joints were swollen, edematous, erythematous, and painful. Her past history-she suffered from recurrent sinusitis and was diagnosed with nasal polyps. No painful joints or edematous joints were recorded over the years.

On admission she had bilateral diffuse inspiratory hales and expiratory wheezes. The right ankle was hot, swollen and erythematous. The heart rate was regular without murmurs. The abdomen was soft, non-tender, without hepato-splenomegaly. Neurological examination was normal without pathological reflexes or motor deficit, but she had a sensory deficit in both legs and palms.

ESR was 60 mm/1st hour, hs-CRP was >100 mg/L. Hemoglobin 11.6 gr% (MCV-79, MCH-26.2, Hct 35%, RDW 13.3%). Leukocytes 11,000/ml³ (eosinophils 33%, neutrophils 50%, basophils 0.5%, lymphocytes 11.9%), platelets 357,000. Iron level was 60 microgr/dL. Free T4 was 1.06 ng/dl and TSH was 2.18 IU/ml. Vitamin B12 and folic acid were normal (335 pg/ml and 4.69 ng/ml). Biochemistry was normal with normal kidney (creatinine 0.6) and liver function (AST 39 u/l, ALT 36 u/l, ALP 104 u/l, bilirubin total 0.3 mg/dl). Immunoglobulin E (IgE) was high (3160 IU/ml), IgG 1755 mg/dl, IgA 187 mg/dl and IgM was 164 mg/dl. ANA was positive-speckled, c-anti nuclear cytoplasmic antibodies (c-ANCA) and rheumatoid factor was positive. P-ANCA and anti-cyclic citrullinated peptide (anti-CCP) were negative. Complement levels were normal (C3 165 mg/dl, C4 39.6 mg/dl). ASLO was negative. Blood, urine and stool cultures were all negative. Serological tests for Brucella, Rickettsia, and Q fever were negative. Urine analysis presented proteinuria 75 mg/dl (0.32 g/24 h). Urine microscopy was normal without casts.

ECG showed a normal sinus rhythm, an axis of 30° without conduction delay or bundle branch block, without QT or PR prolongation. Chest X-ray showed bilateral infiltrates that were resolved within 24 hours (Figures 1a and 1b). An echocardiogram was normal. During hospitalization she was treated with systemic corticosteroids (prednisone 1 mg/kg) with a significant clinical improvement.

Discussion

We suspected that she might have Churg Strauss Syndrome

because she had severe asthma with hypereosinophilia and extremely high levels of IgE. Besides, she also had neuropathy and paranasal sinus abnormalities, and the temporary self-resolving pulmonary infiltrates [1]. Asthma is present in 96-100% of patients and usually precedes the phase of vasculitis by a period of weeks to years [1,2] Arthralgia may be present but arthritis is rare. Renal involvement is typical and is usually mild [1,2] Anti-nuclear cytoplasmic antibodies (ANCA) are

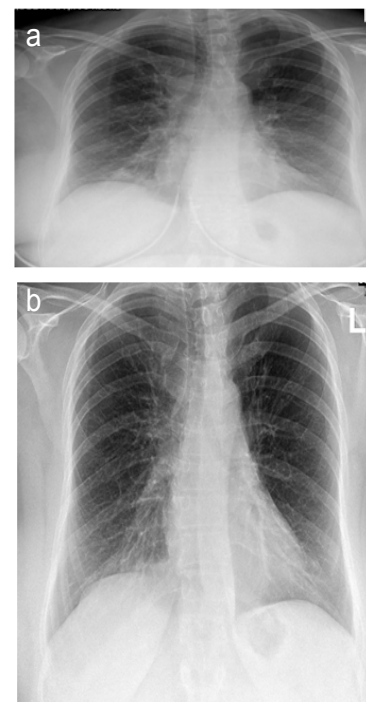


Figure 1: a and b: Chest X rays showing disappearance of the infiltrates spontaneously.

*Corresponding author: Arnon Blum, Department of Medicine; Baruch Padeh Poria Hospital Bar Ilan University, Lower Galilee 15208, Israel, Tel: 97246652687; E-mail: ablum@poria.health.gov.il

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demonstrated in 48-66% of patients [2] There are differences in organ involvement according to ANCA status: ear, nose and throat signs, peripheral neuropathy and renal involvement are more frequent in ANCA (+) than in ANCA (-) patients [3] Omalizumab may trigger CSS, but it is more acceptable that corticosteroids tapering down (allowed by these drugs) and not Omalizumab that causes CSS flares [4]. However, in our case the severe asthmatic attacks and recurrent sinusitis with nasal polyposis were present for many years and preceded treatment with Omalizumab.

This is the first patient that has been described in the literature who had suffered from Churg Strauss Syndrome and migratory polyarthritis.

References

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