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Cardiology

Segmental Hypokinetic Cardiomyopathy in Churg Strauss Syndrome: A Case Report and Review of a Spectacular Evolution

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Abstract **Case Report**

Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome) is a necrotizing systemic vasculitis of small vessels, associated with late-onset asthma, blood and tissue hypereosinophilia. Cardiac involvement is a major concern and has been described as the leading cause of mortality. In this context, we report the case of a 48-year-old woman with Churg Strauss syndrome, suffering from late-onset asthma, pansinusitis, hypereosinophilia, and cardiac damage, consisting of segmental hypokinetic cardiomyopathy with an altered ejection fraction and healthy coronary artery, complicated by a flutter treated by electric cardioversion. The combination of bolus corticosteroid therapy with Azathioprine, as well as the reduction of the rhythm disorder, led to an improvement of the contractile function and the ejection fraction in this patient's case. This observation illustrates a particular and exceptional form of myocardial damage due to Churg-Strauss syndrome and its spectacular evolution under background treatment and rhythm control.

Keywords: Eosinophilic granulomatosis, hypereosinophilia, Cardiac involvement, Azathioprine.

INTRODUCTION

Churg–Strauss syndrome (CSS) is vasculitis associated with pANCA (anti-neutrophil cytoplasmic antibodies) and is very often complicated by heart damage [1].

Guillevin *et al,* [2] identified 5 factors associated with poor prognosis (the "Five Factor Score" [FFS]). Among these factors, they pointed out cardiomyopathy, which requires intensive immunosuppressive therapy.
However, the type of cardiac disease may range from mild to severe and can cause substantial morbidity and mortality.

PATIENT AND OBSERVATION

The patient was 48 years old and had a history of asthma, pansinusitis related to Churg Strauss syndrome, and adrenal insufficiency due to corticosteroid use. She also has a history of electrically reduced common flutter complicated by an ischemic stroke of the basilar trunk and posterior cerebral artery, despite prior anticoagulation, successfully removed by thrombectomy.

The patient described stage II NYHA dyspnea, paresthesias in both hands, and altered general condition.

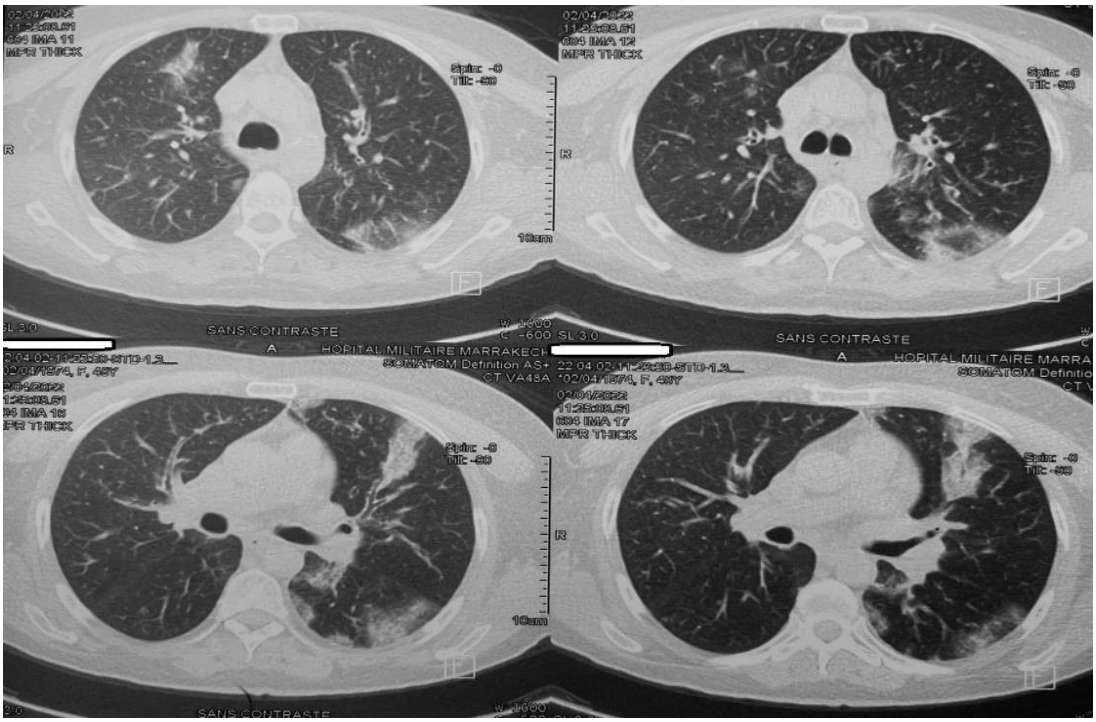
In addition, the number of eosinophilic polymorphs on the blood count was 6420 per µliter.

The CT scan showed bilateral ground-glass and crazy paving of the lungs and chronic maxillo-ethmoidal sinusitis. The pulmonary function tests showed a reversible obstructive syndrome after the application of bronchodilators. The electro-myography showed a sensory carpal tunnel on the right arm.

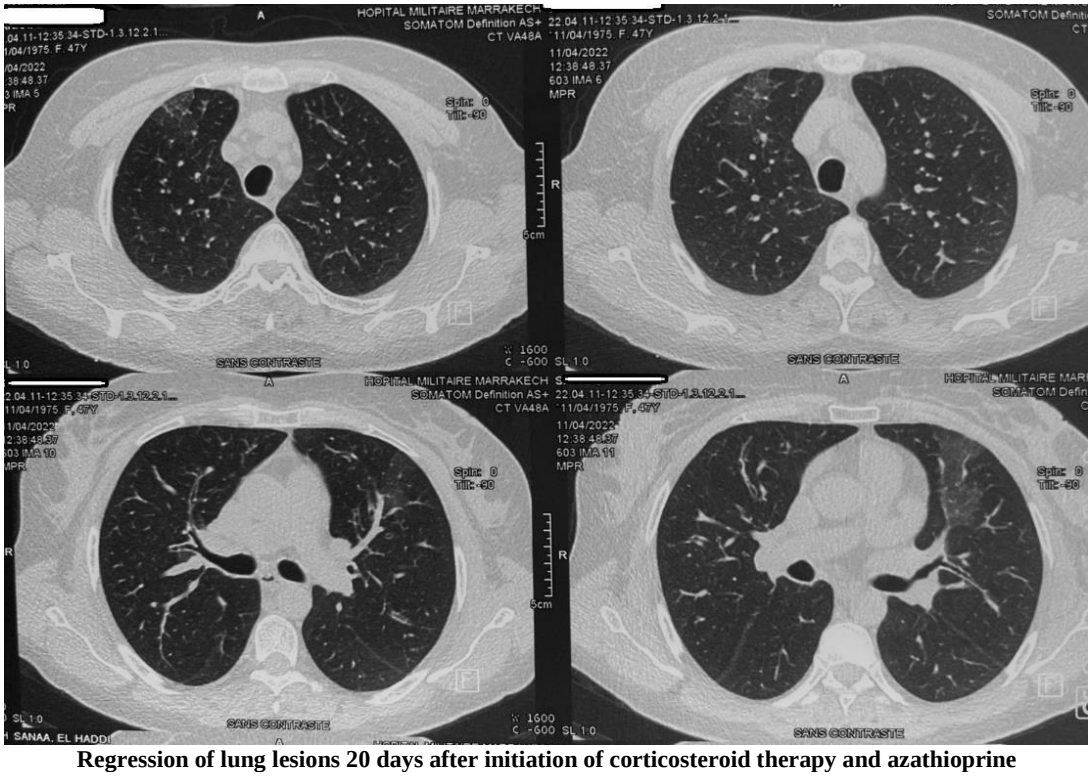
The chest ultrasound showed segmental hypokinesia at the level of the septum at the basal and medial levels, with a moderately altered ejection fraction estimated at 45%. Subsequently, coronary angiography was performed and no coronary artery involvement was identified.

In order to initiate the treatment, we first administered bolus solumedrol and then oral corticosteroid therapy in combination with azathioprine. 20 days after the beginning of treatment, the dyspnea

was no longer reported, and the CT scan showed a clear improvement of the lung damage. The 3-month follow-up showed a spectacular improvement of the ejection fraction to 51%, and the correction of the contractile function of the septum.



Pre-treatment scan showing multiple ground glass foci and crazy paving



Regression of lung lesions 20 days after initiation of corticosteroid therapy and azathioprine

DISCUSSION

In their series of autopsies, Churg and Strauss found more than 50% of cadavers with heart lesions [3] affecting the three layers of the heart.

Clinical data from large cohorts revealed pericardial effusion and the clinical pattern of cardiomyopathy as CSS's most common cardiac features [4, 5]. Cardiac valve abnormalities, granulomatous myocarditis, coronary vasculitis, and

other clinical features have all been described but are mainly restricted to single case reports.

Apart from clinical assessment, screening for EGPA-related cardiac involvement should include electrocardiography, echocardiography, and plasma troponin measurement to detect myocardial ischemia. Echocardiography may show wall-motion abnormalities, signs of pericarditis, or intra ventricular thrombus. Coronary abnormalities can be ruled out by left-heart catheterization. Cardiac MRI provides a detailed anatomic description of the lesions; both first-pass deficits, and late gadolinium enhancements, are suitable to detect myocarditis and myocardial fibrosis. Therefore, active inflammation and fibrous changes remain difficult to differentiate [6, 7].

In our case, we observed an isolated involvement of the inter ventricular septum. In CSS cases, the myocardial damage is related to the activation and proliferation of PNEs [8]. Therefore, we can suggest that the localized decrease in myocardial strain reflects regional myocardial infiltration.

The therapeutic management of heart lesions for CSS patients consists mainly of corticoids in the case of pericarditis [9], and the addition of cyclophosphamide in the case of myocardial involvement [2]. Management is empirical and is based on the same immunosuppressive therapies as those administrated for severe visceral involvement, which themselves arise from the therapeutic approaches used in other forms of ANCA-related vasculitis. The preponderant role of eosinophils may encourage the use of targeted molecules such as anti-interleukin 5 monoclonal antibodies, which are currently being tested in phase II/III in CSS [10], or omalizumab (an anti-IgE antibody) used in essential hyper eosinophil syndrome [11]. We also mention that omalizumab may be associated with the onset of CSS in few cases [12].

However, for patients requiring high glucocorticoid exposure to achieve durable disease control, many experts routinely add medium-potency immunosuppressants, such as azathioprine or methotrexate, as a glucocorticoid-sparing measure [13].

CONCLUSION

Churg Strauss syndrome is one of the most puzzling primary systemic vasculitides. In CSS cases, cardiac involvement is frequent, underestimated, and therefore the diagnosis is often late and retrospective leading to a poor prognosis. However, our case shows an amazing recovery under corticosteroids and immunotherapy. As a result, we can conclude that the detection of endomyocarditis in CSS patients requires early and aggressive treatment.

DECLARATION OF INTERESTS

The authors declare that they have no conflicts of interest in relation to this article.

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