Case Report

Exacerbation of hypereosinophilic syndrome with pulmonary involvement in two consecutive pregnancies: a case report and review of the literature

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Received: 5 October 2014
Accepted: 26 October 2014

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ABSTRACT

Hypereosinophilic syndrome represents a heterogeneous group of disorders with the common feature of prolonged eosinophilia of unknown cause and organ system dysfunction including the pulmonary system. Hypereosinophilic syndrome, with and without pulmonary involvement, in association with pregnancy is very rare, and to the best of our knowledge only one case of hypereosinophilic syndrome with pulmonary involvement during pregnancy has been previously reported in the medical literature. We describe a case of a patient with previously symptomatic hypereosinophilic syndrome with pulmonary involvement who experienced exacerbations of her disease during two consecutive pregnancies. To the best of our knowledge this is the first report which demonstrates a worsening effect of pregnancy on both eosinophil count and end organ involvement in a patient with previous diagnosis of hypereosinophilic syndrome.

Keywords: Hypereosinophilic syndrome, Pulmonary involvement, Pregnancy, Exacerbation

INTRODUCTION

Hypereosinophilic syndrome (HES) is a rare disease which is classically defined as: 1) peripheral blood eosinophil count greater than \(1.5 \times 10^9/L\) for 6 months or longer, 2) evidence of organ involvement, and 3) absence of secondary causes of eosinophilia, such as allergic and parasitic disorders, solid and hematologic malignancies, Churg-Strauss disease, and HTLV infection.\(^1\) This classic definition was recently extended to include patients with new onset clear-cut idiopathic hypereosinophilia causing end-organ involvement in whom treatment is begun rapidly to prevent further damage, as these patients do not fulfil the duration criterion.\(^2\) Proposed new definition of HES includes eosinophil levels higher than \(1.5 \times 10^9/L\) on at least two occasions separated by at least 4 weeks (excluding cases where urgent therapy is required) and evidence of eosinophil-related organ damage.\(^3\) Symptoms of this disease vary, as different organs may be involved. The lungs are commonly involved in HES. Pulmonary involvement can vary widely, ranging from chronic dry cough or bronchial hyperreactivity, often without radiological abnormalities, to restrictive disease with pulmonary infiltrates. Prolonged disease may result in pulmonary fibrosis.\(^2\)

HES, with and without pulmonary involvement, in association with pregnancy is very rare, and to the best of our knowledge only one case of HES with pulmonary involvement during pregnancy has been previously reported in the medical literature.\(^4\) We describe a case of a patient with previously symptomatic HES with pulmonary involvement who experienced exacerbations of her disease during two consecutive pregnancies. To the best of our knowledge this is the first report which demonstrates a worsening effect of pregnancy on both eosinophil count and end organ involvement in a patient with previous diagnosis of HES.
CASE REPORT

A 32-year-old previously healthy female underwent workup on an outpatient basis due to episodic cough and dyspnea. Repeated laboratory tests demonstrated significant persistent peripheral blood eosinophilia (>8.0 x 10^9/L). Chest x-ray film showed bilateral pulmonary infiltrates, and chest computerized tomography (CT) scan revealed multiple patchy infiltrates in both lungs with areas of ground glass opacities and bilateral hilar and mediastinal lymphadenopathy. Pulmonary function testing revealed a moderate obstructive disease with air trapping. Workup for secondary causes of eosinophilia was negative and echocardiogram was normal. She was diagnosed as suffering from HES with pulmonary involvement, and was begun on daily prednisone therapy of 60 mg, which was later tapered to 5 mg/day. The patient did well under this regimen, with resolution of cough and normalization of peripheral blood counts. Repeated chest x-ray film and pulmonary function testing, performed 3 months later, were normal.

A year later she was admitted to the OBGYN department due to the onset of labor at 40 weeks gestation. During her stay at the OBGYN department she complained of dyspnea and persistent productive cough with clear mucus without fever for the past several weeks. Physical examination was notable for prolonged expirium with expiratory wheezing. Her oropharynx was clear, and cardiac examination was normal. Other physical findings were unremarkable. Routine laboratory tests revealed marked eosinophilia (10.4 x 10^9/L), without other abnormalities. Chest x-ray film showed again bilateral pulmonary infiltrates, and pulmonary function testing revealed moderate obstructive disease with air trapping. The respiratory complaints were attributed to exacerbation of pulmonary disease due to HES, and prednisone dosage was increased to 60 mg/day, with subsequent dosage tapering down to 5 mg/day. Her response to treatment was favorable, with resolution of symptoms and normalization of peripheral blood counts. She gave birth to a healthy male infant through vaginal delivery, with the infant not showing hypereosinophilia.

Two years later, the patient became pregnant again, while still on the same prednisone regimen which was started three years earlier. She was admitted to the internal medicine department for worsening dyspnea and dry cough when she was at 29 weeks of gestation. Physical examination revealed marked expiratory wheezing and prolonged expirium. Laboratory tests revealed eosinophilia of 2.8 x 10^9/L, and chest x-ray film was normal. Once again a diagnosis of exacerbation of pulmonary disease due to HES was made, and prednisone dosage was increased to 20 mg/day. The patient responded well to treatment, with resolution of respiratory complaints and peripheral eosinophilia. She later delivered a healthy male infant through vaginal delivery, with the infant not showing hypereosinophilia.

DISCUSSION

Studies performed in healthy subjects found pregnancy to be accompanied by leukocytosis and decrease in circulating eosinophil counts. Subsequently, much later, a case report was described showing this phenomenon in a patient with HES and pulmonary involvement, who experienced complete resolution of pulmonary complaints and normalization of peripheral blood count during pregnancy. Following delivery her eosinophil counts rapidly increased to pre-pregnancy levels. Very few reports, however, have described the opposite phenomenon: development or exacerbation of HES during pregnancy, with involvement of the lungs, and the heart, and also without end organ damage.

A variety of pulmonary complications have been reported in pregnant women with HES, or other types of eosinophilic lung disease. One case report described a patient who developed new onset HES with pulmonary involvement during pregnancy characterized by cough, dyspnea, and pulmonary infiltrates. Interestingly, the patient's newborn also developed a transient elevation of blood eosinophil count, which resolved at the age of 8 months, and has remained normal since. Another report described a case of acute eosinophilic pneumonia during pregnancy, which led to emergent cesarean section due to fetal distress as a result of maternal hypoxemia. Relapses of chronic eosinophilic pneumonia during pregnancy and in the post-partum period have also been reported.

Corticosteroids have been the cornerstone of management in HES since the syndrome was defined, and are the first-line therapy for most patients. Our patient was on corticosteroids since her diagnosis of HES, with good initial clinical response. She required an increase in corticosteroid dosage during both pregnancies, in order to control her respiratory symptoms and rising eosinophil counts.

The present case describes a patient with previously symptomatic HES with pulmonary involvement which exacerbated during pregnancy, and was well controlled with prednisone treatment afterwards, only to exacerbate again during her next pregnancy. We believe this is the first report which demonstrates a worsening effect of pregnancy on both eosinophil count and end organ damage in a patient with previously diagnosed HES.

In summary, although eosinophil count may decrease during pregnancy, the opposite may also occur with exacerbation of HES. More research and clinical experience is needed in order to determine the exact effect of pregnancy on eosinophils and on the clinical course of HES.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required
REFERENCES


DOI: 10.5455/2320-6012.ijrms201411110