CASE REPORT

Postpartum onset and subsequent relapse of eosinophilic granulomatosis with polyangiitis

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SUMMARY

Eosinophilic granulomatosis with polyangiitis (EGPA) can affect women of childbearing age. However, reports of the disease in the postpartum period are limited. We present a case of postpartum-onset EGPA that went into clinical remission before relapsing in the subsequent postpartum period. Our patient presented with dyspnoea, arthralgia and rash, shown to be eosinophilic vasculitis, 3 days following the birth of her second child. CT of the thorax showed alveolar shadowing and mediastinal lymphadenopathy. She was treated successfully for EGPA with glucocorticoid therapy. She declined maintenance treatment during remission. Off treatment, she remained disease free throughout her next pregnancy. In the postpartum period she relapsed in an almost identical manner, requiring prolonged glucocorticoid therapy, cyclophosphamide and rituximab. This case highlights the importance of maintenance therapy around pregnancy in individuals with EGPA, and the need for careful monitoring of women with a history of EGPA in the postpartum period.

BACKGROUND

Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare eosinophil-rich and necrotising granulomatous inflammation often involving the respiratory tract, and necrotising vasculitis predominantly affecting small to medium vessels, and is associated with asthma and eosinophilia.¹ Although women of childbearing age can be affected by EGPA, reports of the disease around pregnancy, and especially in the postpartum period, are limited.² A case is presented of postpartum-onset EGPA that went into clinical remission before relapsing in the subsequent postpartum period.

CASE PRESENTATION

A 31-year-old woman with a history of asthma since childhood presented with dyspnoea, wheeze and productive cough 3 days following the birth of her second child. She also developed arthralgia and a nodular, pruritic, erythematous rash. Six weeks later, her breathlessness worsened and she became febrile with nausea, abdominal pain, rigors, peripheral numbness and a rash, shown on biopsy to be eosinophilic vasculitis.

INVESTIGATIONS

The patient was hypoxic (pO₂ 7.2 kPa on air) and antineutrophil cytoplasmic antibodies negative, with peripheral eosinophilia (13×10⁹/L), and CT of the thorax showed alveolar shadowing and mediastinal lymphadenopathy.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses, including infection and haematological malignancy, were explored by sputum cultures, blood cultures and a blood film. When these were negative, further investigation was not felt necessary given the patient's history, CT findings and positive skin biopsy.

TREATMENT

The patient was diagnosed with EGPA and successfully treated with intravenous followed by tapered oral glucocorticoids (18 months total). She was intolerant of azathioprine, and in remission she declined alternative maintenance therapy. Off treatment, she remained disease free for 6 months prior to and throughout her next pregnancy. In the postpartum period following the birth of her third child, her EGPA again became active with an almost identical presentation to that following her second pregnancy. She partially responded to glucocorticoids and cyclophosphamide, but required continued high-dose oral prednisolone to maintain disease control. Consequently, after 2 years, she was started on Rituximab and her early response has been promising.

DISCUSSION

On review of the literature, EGPA during pregnancy is rare and results in poor fetal outcome in approximately a third of cases.³ Such flares may occur due to immunological or hormonal changes, increased physiological stress reactivating latent disease, or may be picked up by more detailed medical surveillance.⁴ The estimated risk of a flare in pregnancy in an individual with a prior diagnosis of EGPA is reported to be between 25% and 50%. Obstetric risks of a flare of EGPA in pregnancy include pre-eclampsia (<5%), cardiac complications, flares of asthma, fetal loss (10–15%; around 50% if EGPA disease is active around conception), preterm birth (10–40%), intrauterine growth retardation (10–25%) and need for caesarean section (15–40%).⁵

Postpartum onset is less well documented. Four cases have been reported and three of these had been diagnosed with, or had symptoms of, asthma or allergic rhinitis prior to developing the disease. All four pregnancies had resulted in a live birth;^{3 6–8} however, severe cardiac involvement subsequently led to one maternal death. One of the surviving women completed a further pregnancy on low-dose (5 mg daily) prednisolone without recurrence of symptoms.⁶

Currently, triggers for EGPA onset and flares are not fully understood, and this case may therefore

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highlight a potential precipitating factor. This knowledge may aid in the understanding of the disease pathogenesis.

Learning points

- ▶ To the best of our knowledge, this is the first documented case of a woman initially presenting with eosinophilic granulomatosis with polyangiitis (EGPA) postpartum who had a further flare in the same period following a subsequent pregnancy. Temporal relationships and initial clinical features of both episodes were almost identical and thus a causal link might be inferred.
- ▶ This case also highlights the importance of maintenance therapy around pregnancy in individuals with EGPA.
- ▶ Physicians should be aware of the possibility that this condition might develop in pregnant or postpartum women, particularly those with asthma.

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Competing interests None declared.

Patient consent Obtained.

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