Severe relapse of Wegener’s granulomatosis during the early postpartum period

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Pregnancy in Wegener’s granulomatosis (WG) has been rare, but increasing numbers of pregnancies are now being reported, not least because of the use of less toxic drug regimens. New onset disease during or after pregnancy has been noted previously, but postpartum relapse has not been reported so far. Here, we report a severe postpartum relapse of WG after longstanding remission.

A 27 year old woman was first diagnosed with WG of the upper and lower respiratory tract, central nervous system, eye, and skin in 1995. Treatment with methylprednisolone (500 mg/day for 5 days), oral cyclophosphamide (3 mg/kg), intravenous immunoglobulins, and co-trimoxazole induced remission. Cyclophosphamide was stopped in 1996. Prednisolone and co-trimoxazole were stopped in August 1997 and June 1999, respectively, while the patient continued to be in full remission.

The patient subsequently became pregnant, with an uneventful delivery in September 2000 and a normal postpartum period. In 2004, the patient became pregnant again after she had been counselled about the risk of relapse. When seen in our clinic in August 2004, she was in good health; the C reactive protein (CRP) was normal and antinuclear cytoplasmic antibodies (ANCA) were negative.

At that time, proteinuria was 0.29 g/day and urine examination showed no dysmorphic erythrocytes. The patient subsequently had another uneventful delivery on 20 September 2004.

On 27 September, the patient presented with urinary tract infection, which was treated appropriately. On 7 October, she developed an abscess of the left mamma requiring incision and drainage. In mid-October 2004, the patient developed maxillary pain, dry cough, and fever that did not respond to antibiotic treatment. On 18 October, she was first seen in our clinic after she had received treatment elsewhere during pregnancy.

Computed tomography showed sinusitis and pulmonary infiltrates as well as nodules, and bronchoscopy demonstrated bronchitis. Bronchoalveolar lavage showed neutrophil and eosinophil alveolitis with no growth in culture. The CRP peaked at 439 mg/l with normal procalcitonin. Urine examination disclosed dysmorphic erythrocytes, and proteinuria increased to 0.85 g/day. Serum creatinine level and clearance were normal. The ANCA became positive at a titre of 1/32. A relapse was diagnosed with involvement of upper respiratory tract, lung, and kidney. Methylprednisolone (500 mg/day) and pulsed intravenous cyclophosphamide were started, with good response, and CRP values declined.

As of January 2005, the patient is well with steroids and monthly cyclophosphamide.

The number of pregnancies in patients with WG is reviewed elsewhere. Some 26 pregnancies in patients with WG have been reported, and we describe the first in which relapse occurred post partum. Relapse during pregnancy has been noted previously and it has been estimated that among women with WG who conceive in remission, about one in four relapse. Lima and colleagues described two relapses during pregnancy. Active disease at the onset of pregnancy appears to be correlated with poor outcome, and maternal mortality has been reported. It has been noted that other small vessel vasculitides occur in association with pregnancy as well.

We report the first case of relapse during the early postpartum period. We were surprised to see widespread disease develop so quickly after pregnancy, especially after 7 years of remission. The preceding infections may also have triggered the relapse. Our patient underlines the observation that length of remission does not predict an uncomplicated course during and after pregnancy. Moreover, our patient confirms the impression that uneventful previous pregnancies do not exclude a relapse with subsequent pregnancies.

Finally, our report refutes the assumption that persistently negative ANCA titre indicate a low likelihood of peripartum relapse. Treatment of active WG in pregnancy has been reviewed elsewhere.

In conclusion, we report the first case of relapse of WG post partum. We fear that vasculitis in conjunction with pregnancy may occur more often than expected and propose meticulous reporting of such cases. We speculate that immune events associated with pregnancy may trigger disease, as in lupus. We emphasise the need for pregnancy counselling in female patients of childbearing age with vasculitis and recommend close surveillance during pregnancy and post partum.

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