CASE REPORT

Successful Pregnancy in Untreated Limited Wegener's Granulomatosis

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Summary

A thirty four year old female presented with upper and lower respiratory symptoms in the third trimester of pregnancy. After the delivery of a healthy baby, the symptoms progressed to involve multiple organ systems and eventually a diagnosis of limited Wegener's Granulomatosis (Carrington-Liebow syndrome) was made. The extremely rare combination of WG and pregnancy, especially the onset of disease in late pregnancy is discussed. The successful outcome of pregnancy even without treatment of WG is the highlight of the case.

Key Words: Limited Wegener's Granulomatosis, Carrington-Liebow syndrome, Pregnancy

Introduction

Wegener's granulomatosis (WG) is a systemic granulomatous vasculitis that primarily involves the upper and lower respiratory tracts and kidneys. It belongs to primary systemic vasculitides of unknown etiology associated with antineutrophilic cytoplasmic antibodies (ANCA). Klinger first described this disease as a special form of polyarteritis nodosa in 1931. WG was established as a distinct clinicopathologic entity later in that decade by Wegener. The disease exists in two forms: an indolent form called limited, initial, or locoregional WG, and a fulminant form called active or generalized WG. The generalized form is diagnosed by the American College of Rheumatology diagnostic criteria that include nasal or oral inflammation, nodular infiltrates on chest imaging, nephritis, granulomatous inflammation on biopsy. The presence or absence of renal disease separates the generalized and limited WG respectively. Carrington and Liebow in 1966[1] first described 16 patients of WG without renal involvement. Since then various reports of Limited WG (Carrington-Liebow syndrome) with varied manifestations have been published. We report a case of limited Wegener's that presented during pregnancy and then progressed systemically after the successful completion of pregnancy.

Case Report

A thirty four year old female presented with dry cough, low-grade fever, headache and watery nasal discharge of 20 days duration in the 32nd week of pregnancy to a rural hospital. There was no history of similar complaints in the past or during the patient's previous two successful pregnancies. On examination the patient was febrile, nasal mucosa was erythematous. Other system examination was unremarkable. The initial laboratory investigations showed leukocytosis with an ESR of 110 mm/hour. All other biochemical parameters including kidney and liver function tests were normal with a normal urine analysis. A chest x-ray done with an abdominal shield revealed multiple nodular parenchymal opacities in the left upper and

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right middle zones. The patient was not investigated further and was started on antitubercular therapy despite a negative sputum report for acid-fast bacilli.

While there was little improvement in the symptoms, the patient delivered a healthy male baby weighing 2.6 kilograms in the 38th week through the vaginal route. However, she remained symptomatic despite taking antitubercular drugs, which she discontinued after 5 weeks. Over the next two months she developed redness of eyes, numbness over the medial aspect of the left hand and over the left calf. She continued to take treatment in the local hospital where she was even given a short course of oral steroids with marked improvement in symptoms.

Before presenting to us, she developed a nonpruritic erythematous rash over the dorsum of both feet (Figure 1) without photosensitivity or rash over any other part of the body. There was no history of peripheral edema, oliguria or haematuria. No past history of allergic disorders, blood dyscrasias, tuberculosis or of any other systemic or metabolic disorder. The patient had two live issues with no history of menstrual disturbance, abortion or foetal loss. No family history of a similar disorder was found.

On examination she was normotensive with an erythematous maculopapular rash over feet, episcleritis and conjunctivitis of both eyes and weakness of small muscles of the left hand. Investigations showed haemoglobin of 11gm/dl, leukocyte count of 8000 cells/cu.mm, ESR of 96 mm/hour, blood urea of 28mg/dl, creatinine of 1.1 mg/dl. All other biochemical investigations including urine analysis were normal with no evidence of red cells in the urine. A Mantoux test was 2mm. Sputum for acid-fast bacilli was negative. A repeat chest x-ray showed similar findings.

An HRCT chest revealed multiple scattered nodular lesions with partial consolidation of the posterior segment of the left upper lobe and lateral segment of the right middle lobe (Figure 2). CT scan of paranasal sinuses was suggestive of maxillary sinusitis. ANA and RA factor were negative. Serum levels for Angiotensin Converting Enzyme (ACE) was 22U/L (normal <40U/L), c-ANCA was highly positive with a level of 36.4 U/ml (normal- 0.00 to 7.00U/ml). The tissue obtained by a transbronchial biopsy of the lung revealed necrotizing granulomatous vasculitis consistent with Wegener's Granulomatosis. A nerve conduction study revealed axonal neuropathy of the left ulnar and left sural nerves.

Thus, a diagnosis of limited WG with mononeuritis multiplex was made and the patient started on oral prednisolone (50mg/d) and cyclophosphamide (100mg/d). The patient has subsequently improved with no recurrence of ocular or skin manifestations. Her cough had also decreased. However, the numbness over the ulnar distribution of the left hand had increased leading to severe incapacitation.

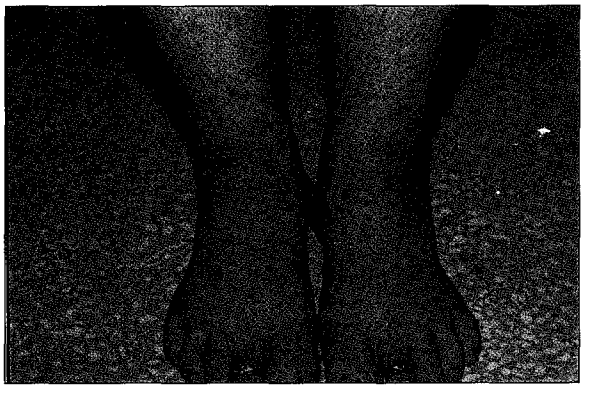

Fig. 1: Erythematous maculopapular vasculitic rash over the feet

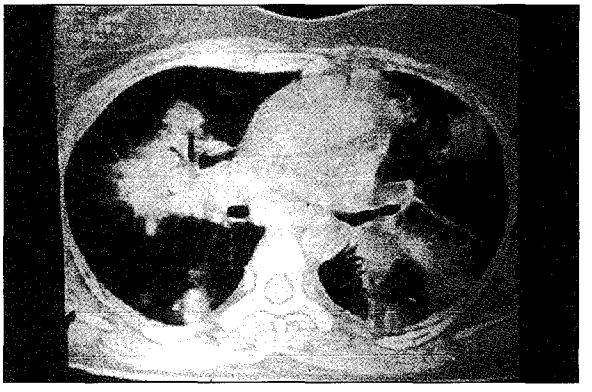

Fig. 2: High resolution contrast enhanced CT scan of the chest showing multiple scattered nodular lesions with partial consolidation of the posterior segment of the left upper lobe and lateral segment of the right middle lobe

Discussion

Reports of pregnancy in WG are rare. Fewer still are reports of WG presenting for the first time in pregnancy. As far as could be determined WG has been described with 26 pregnancies in 20 women till date. Two out of these were cases of limited Wegener's[2,3]. Out of these WG was diagnosed for the first time during pregnancy in 6 cases only three of which culminated in a successful outcome[2], the rest being cases of pregnancy in patients already diagnosed to be having WG.

In the present report, the patient presented for the first time in the 32nd week of pregnancy. The disease later flared up with manifestations of skin vasculitis, upper and lower respiratory involvement but without evidence of nephropathy as evidenced by a normal urine analysis and KFTs with no significant proteinuria. The HRCT chest, lung biopsy and c-ANCA prove the diagnosis of limited WG beyond doubt.

The effect of pregnancy on the course of WG is not known with certainty. In a series of 5 pregnancies in three patients by Auzary[3] et al, no flare up occurred during pregnancy. But flare-ups of the disease during pregnancy are described. In contrast some other reports do not show any increase in symptoms during pregnancy. The outcomes of pregnancies have also been variable. Though some cases of abortion and foetal death are reported, most pregnancies have resulted in a favourable outcome. Prematurity[3] however is a common complication with a mean duration of gestation of 36 weeks.

In the present case, the pregnancy was uneventful with no pre-eclampsia, prematurity, increased maternal morbidity or foetal adverse effects. Although most immunosuppressants are avoided during pregnancy, there are reports of successful treatment of WG in pregnancy with corticosteroids, azathioprine, and even cyclophosphamide. In a case reported by Dayoan[2] et al a healthy male baby was born to a patient suffering from limited Wegener's being treated with cyclophosphamide.

So to conclude, limited Wegener's Granulomatosis may rarely present for the first time in the third trimester of pregnancy. The highlight of this case is the successful outcome even without treatment, which is rare.

References

1. Carrington CB, Liebow AA. Limited forms of angiitis and granulomatosis of Wegener's type. Am J Med 1966; 41: 497-527.
2. Dayoan ES, Dimen LL, Boylen C. Successful treatment of Wegener's granulomatosis during pregnancy. Chest 1998; 113: 836-38.
3. Auzary C, Thi Huong D, Wechsler B, Vauthier-Brouzes D, Piette JC. Pregnancy in patients with Wegener's granulomatosis: report of five cases in three women. Ann Rheum Dis 2000; 59: 800-04.