

Successful term pregnancy in a patient with Wegener's granulomatosis complicated with renal failure: A Case report

Ibrahim Abdellahi, Ali Osman, Jaouad Kouach, Moussaoui Idrissi, and Mohammed Dehayni

Department of obstetrics & gynecology, Military Training Hospital Mohamed V, Rabat, Morocco

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ABSTRACT: Wegener granulomatosis is an autoimmune small vessel vasculitis highly associated with anti-neutrophil cytoplasmic antibodies and characterized by necrotizing granuloma of respiratory tract, disseminated vasculitis and glomerulonephritis. Its clinical manifestations and organ involvement are very widely. The etiology of Wegener granulomatosis is linked to environmental and infectious triggers inciting onset of disease in genetically predisposed individuals. Complications most often occur when the disease is not treated. Kidney disease causes glomerulonephritis with hematuria. It can quickly get worse with kidney failure, but can be prevented when the condition are controlled by medicines. We report a case of a pregnant woman with Wegener's granulomatosis whose disease involved the glomerulonephritis that lead the end stage renal failure and need the dialysis with Successful term pregnancy.

KEYWORDS: antineutrophil cytoplasmic antibodies (ANCA), Granulomatosis, Polyangiitis.

1 INTRODUCTION

Wegener's granulomatosis (WG) is a systemic necrotizing, small vessel vasculitis [1]. It was first described in the medical literature in the late 19th century and was formerly known by the eponymous name Wegener's Granulomatosis. The use of disease-descriptive, etiology based nomenclature is now recommended and preferable to the use of eponymous names, therefore since 2011 Wegener's Granulomatosis has been known as granulomatosis with polyangiitis (GPA) [2]. The disease peaks after the age of 40 years, thus, pregnancies in women with GPA are uncommonly observed [3]. Effects of WG on pregnancy are: Spontaneous abortion, Premature delivery, preeclampsia, Prelabor rupture of membranes, prepartum hemorrhage and retroplacental hematoma [4]. We report a case of the pregnant woman of 42 years old, known case of GP with a Kidney lesions cause hematuria, glomerulonephritis, , Kidney disease was quickly get worse. Kidney function may not improve even when the condition is controlled by medicines and finally she developed an end stage renal failure under renal dialysis and respiratory involvement (repetitive cough with hemoptysis with partially amelioration with treatments also affection neural as right facial paralysis and chronic sinusitis. The full multidisciplinary follow up with Successful term pregnancy with relapse during pregnancy.

2 CASE PRESENTATION

A 37-year-old Gravida 5, para 4, woman was referred to our department at 13 weeks' gestation owing to a 6-years diagnosis of WG, Her disease course initially included repetitive hemoptysis and at the same time she presented syndrome of the increase intracranial pressure, deviation of the mouth towards left and the examination finds an tetrapyramidal irritation and right facial paralysis which necessity the hospitalization in department of pulmonary at 2009. Antineutrophil cytoplasmic antibodies (ANCA) titer was positive she was put under bolus dose of corticosteroids: 500mg/day during 5 days then discharge oral under 30mg/day of corticoid and oral antibiotic amoxicillin 1g bid during 10days and asprine: 100mg/day, with regular follow-up.

4-years later she presented the dry cough associated hemoptysis and nocturnal dyspnea, lower limb oedema, she developed rapid progressive renal failure (40mg/dl to 70mg/dl within 5 days), urea 4g/l, normocytic normochromic anemia : 6mg /dl, massive proteinuria : 2g/24h, she was admitted the department of nephrology and was put under bolus dose of corticosteroid, blood transfusion. Further investigation was done as:

Endoscopy: found ulcerative esophagitis and pancreatitis. Sinus scanner show bilateral maxillary sinus. Echocardiography shows pericardial thickness.

The outcome was regression of respiratory symptoms and she developed end stage renal failure after a median of 15 months and she is on dialysis.

Laboratory investigations at 13 weeks' gestation showed normal liver function test, mild neutrophil leukocytosis and elevated C-reactive protein, worse renal function test (on dialysis). At the onset of pregnancy she was treated with 20 mg oral prednisolone, fetal growth assessment, congenital abnormalities screening test, and laboratory tests for gestational diabetes and preeclampsia were unremarkable during follow-up with absent ANCA positivity during first and second trimester. The rate of dialysis was two sessions per week in the first two trimesters passed to daily dialysis in third trimester.

The patient presented at 32 weeks mild chest pain, cough and hemoptysis, mild preterm labor, the chest radiography displays bilateral parenchymal infiltrates, with ANCA positive.

Patient received IV corticosteroid and oral calcium channel blockers with good improvement.

At 36 weeks of gestation patient presented spontaneous labor pain and she delivered 2900g healthy baby with apgar 10/10 at first minute and five *minutes*. In postpartum, the patient continued to receive corticosteroids and dialysis with remission.

3 DISCUSSION

WG is a systemic, necrotizing, small-vessel vasculitis. Vascular inflammation and occlusion leading to tissue ischemia [1]. Its annual incidence is 5 to 10 cases per million populations with equal frequency in males and females [2]. Patients with active disease may have several contraindications or risk factors for complicated pregnancy: renal impairment, cardiac insufficiency, uncontrolled asthma in eosinophilic granulomatosis with WG, so that the pregnancy should be planned when the vasculitis shows minimal activity on remission, on pregnancy-safe medication for at least 6 months [4, 5, 6].

The aetiology of WG may originate from infectious include: bacterial mycobacterial, fungal or viral infections, and environmental, chemical, toxic or pharmacological triggers in people who are genetically predisposed to this autoimmune disease [2].

Untreated WG disease has been reported to be fatal within a year of diagnosis in more than 80% of patients [1, 7].

The early symptoms of WG: include the signs like: arthralgia, fever or localized symptoms such as sinusitis, cough, or hemoptysis, anemia, skin ulcers, neural paresis, and others. However they are non-specific manifestations of systemic vasculitis which contribute to a clinical diagnosis of WG and are not pathognomonic [7]. In general for diagnosis of ANCA, there has been no consensus on definitive diagnostic criteria. Both the BSR and EULAR guidelines insist on the necessity of symptoms and signs of vasculitis in the presence of direct histological or serological evidence (positive ANCA) or specific indirect evidence such as imaging [8].

Different study reports that WG is a frequently relapsing disease, and in patients with renal involvement the mortality rate is high and chronic renal failure develops in a considerable fraction of the patients [9]. Because of the high specificity of ANCA in Wegener's granulomatosis and microscopic polyangiitis, ANCA evaluation is indicated in all cases of acute renal failure in patients of any age. However, renal failure is a late clinical manifestation of these vasculitic disorders [7, 2].

In our patient the renal involvement of the disease was very early with development of end stage renal failure that leads on hemodialysis even during pregnancy. The respiratory symptoms were observed at third trimester, with ANCA positivity while it was negative at beginning of the pregnancy and relapse of disease at 32 weeks of gestation occurred [1].

Cyclophosphamide, in combination with steroids, has long been accepted as the gold standard for the management of patients with systemic necrotizing vasculitis. It is very teratogenic and should not be used in pregnancy [10, 5]. Also it has been associated with spontaneous abortions and various birth deformities when it is used during early pregnancy, and so its administration is indicated in life-threatening forms of the disease at the third and late second trimester of pregnancy [8, 11, 12].

The majority of prednisolone taken by the mother is metabolized and very little crosses the placenta unless the dose is greater than 20 mg daily. Nevertheless, corticosteroids increase the risk of maternal hypertension, pre-eclampsia, gestational diabetes and infection [5]. Alternative therapeutic options include cyclosporine and intravenous immunoglobulin that can be used during early pregnancy [1, 3].

In the present case, patient was treated by corticosteroid (bolus IV dose relay by oral way) before and during pregnancy with good result.

4 CONCLUSION

Pregnancy in a patient with WG should be considered as high risk. It is related with various complications and diverse outcome. In this fact, Pregnancies should be planned when the disease is in remission since this reduces maternal complications, increasing the chances of a successful pregnancy. Prematurity is the most common obstetrical complication of WG.

The mother needs to be followed up regularly after delivery because of the high risk of post-partum flare.

CONFLICT OF INTEREST

The authors no conflict to declare.

AUTHORS CONTRIBUTION

All authors contributed to the manuscript's preparation in writing and literature review. All authors approved the final manuscript.

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