A rare case of early onset nephrotic syndrome in pregnancy

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Summary

Nephrotic syndrome occurs very rarely, about 0.012-0.025% of all pregnancies.

Here, we report a rare case of early onset nephrotic syndrome developing de novo in the 17th week of pregnancy. A renal biopsy was done and the specimens revealed typical features of focal segmental glomerulosclerosis. The patient had a progressive clinical course of disease despite steroid treatment. Suffering from severe intrauterine growth restriction, the fetus died in utero. After delivery, steroid treatment was continued. The patient had normal renal function with a decrease in proteinuria in the second and fifth month postpartum. This report points out the poor fetal prognosis associated with an early onset nephrotic syndrome. Pregnant patients with early onset nephrotic syndrome should be carefully evaluated for the presence of chronic renal disease, and primary renal pathology should be included in the differential diagnosis of massive proteinuria in early pregnancy.

Key words: Nephrotic syndrome, Pregnancy, Fetal outcome.

Introduction

Nephrotic syndrome occurs very rarely, about 0.012-0.025% of all pregnancies [1, 2].

Increased proteinuria is a common finding during pregnancy as increased filtered load normally seen in pregnancy results in increased urinary protein excretion, glucosuria, and aminoaciduria. However nephrotic amounts are always abnormal. Preeclampsia can be complicated by nephrotic syndrome. On the other hand, in early pregnancy, nephrotic proteinuria with hypertension is considered suggestive of native renal disease rather than preeclampsia [1, 3]. Early diagnosis and proper management are very important for maternal and fetal health [2].

A case of a pregnant woman with early onset nephrotic syndrome with hypertension developing de novo during pregnancy is presented. Renal biopsy specimens revealed focal segmental glomerulosclerosis and despite treatment of the mother, prognosis of the fetus was poor.

Case

A 21-year-old primigravid woman with persistent edema, hypertension, and nephrotic range of proteinuria at the 19th week of gestation was referred to our hospital. Her past medical history was unremarkable. She had no urinary abnormalities or hypertension before pregnancy. She had first noticed facial and periorbital edema at 17 weeks of gestation. On examination her blood pressure was noted to be 180/100 mmHg. She had lower limb and sacral edema. Laboratory investigation showed no abnormalities except proteinuria of 9.3 g/24 hours. The ultrasound scan of her kidneys showed grade 1 paranchymal pathology.

An ultrasound scan showed that the fetus was growing on the fifth percentile, and the amniotic fluid volume appeared normal. The patient was treated with oral α-methyldopa (1500 mg daily), a low-salt diet was and fluid restriction. As there was a strong suspicion of renal disease, a renal biopsy was performed after her blood pressure had stabilized. Renal biopsy specimens revealed typical features of focal segmental glomerulosclerosis (Figure 1). At immunohistochemical examination there was no accumulation of IgG, IgM, IgA and C3. We started her on 1 mg/kg prednisolone for massive proteinuria. Over subsequent days the patient had a progressive clinical course and despite the medical interventions with steroids she developed uncontrolled hypertension and edema. Suffering from intrauterine growth restriction (IUGR), the fetus was lost in utero in the 27th week of gestation. After termination of pregnancy, the treatment was maintained with angiotensin-converting enzyme inhibitor and prednisolone (40 mg/day). Two months post-termination she was seen in the renal clinic. Her 24-hour urinary protein excretion was down to 2.8 g and renal function was normal. Her blood pressure was 120/80 mmHg with antihypertensive treatment. After a 5-month follow-up, she was observed as having normal albumin levels and stable proteinuria (1 g/day) and her prednisolone dose was gradually tapered to 8 mg/day.

Discussion

Pregnant patients with nephrotic syndrome should be carefully evaluated for the presence of chronic renal disease [4]. Women who have gestational nephrotic range proteinuria or preeclampsia before 30 weeks' gestation are more likely to have underlying renal disease [1]. It is necessary to find out whether the nephrotic syndrome is evidence of an underlying nephropathy or just due to the preeclampsia. Sometimes it is necessary to treat the nephrotic syndrome with steroids [2]. In pregnancy steroid treatment has additional maternal and fetal problems. Thus, it is important to know the histology before starting the treatment [3, 4].

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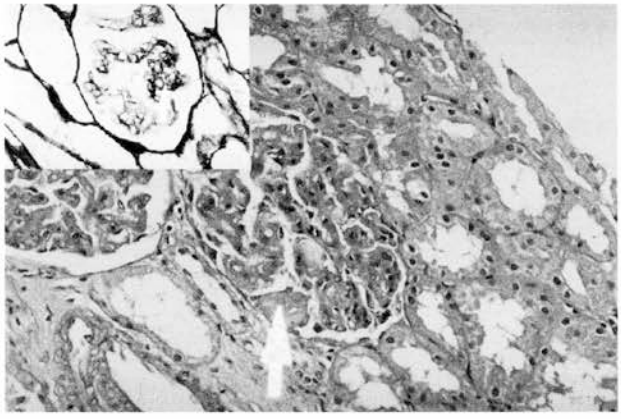

Figure 1. — Segmantal sclerosis area in renal biopsy specimens (100 x; H&E - 200 x; PAMS).

Renal disease may develop de novo during pregnancy. Investigation of the cause of renal disease during pregnancy can be conducted with serologic, functional, and ultrasonographic testing [4]. Renal biopsy for renal dysfunction of unknown cause or symptomatic nephrotic syndrome performed during pregnancy is not contraindicated. The results of histopathological studies are extremely useful in counseling regarding continuation or termination of pregnancy, potential maternal and fetal outcome, and recommending specific therapeutic modalities. On the other hand renal biopsy in pregnancy is a morbid procedure and should be considered only if it offers the opportunity to make a diagnosis other than severe preeclampsia especially in cases developing early in pregnancy [2, 3]. Severe preeclampsia is sometimes associated with nephrotic syndrome [5]. In most of these patients clinical signs first develop in the third trimester. Renal-biopsy specimens taken from preeclamptic patients are sometimes associated with focal-segmental glomerular sclerotic lesions that closely resemble those of primary focal-segmental glomerulosclerosis [6, 7].

The etiology and pathogenesis of focal and segmental glomerulosclerosis (FSGS) in patients with toxemia during pregnancy remain controversial [6]. Results indicate that FSGS may not only be induced by preeclampsia but also be one of the representative glomerular changes in preeclamptic patients with nephrotic syndrome. FSGS-like lesions can occur during pregnancy in "pure" preeclamptic patients but the lesions may not be progressive. The renal lesions appear fully reversible and the disease has no remote cardiorenal effects on patients [5-7].

This report points out the poor fetal prognosis associated with early onset nephrotic syndrome. Since intact renal function is necessary for the physiologic adjustments to pregnancy, such as vasodilatation, lower blood pressure, increased plasma volume and increased cardiac output, pregnant patients with early onset nephrotic syndrome should be carefully evaluated for the presence of chronic renal disease.

In our case, there was apparent preeclampsia and nephrotic range proteinuria at 17 weeks of gestation. Early onset nephrotic syndrome with a histology of focal segmental glomerulosclerosis has been a diagnostic dilemma [4]. This report highlights the need for increased awareness of this rare presentation of nephrotic syndrome. We conclude that primary renal pathology should be included in the differential diagnosis of massive proteinuria in early pregnancy.

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