



Mixed Connective Tissue Disorder Presenting As Eclampsia In A Primigravida: A Case Report

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ABSTRACT

A 23 year old primigravida was diagnosed as a case of Mixed Connective Tissue Disorder (MCTD) after she presented with petechie, synovitis and raynauds phenomenon in the first trimester of pregnancy and was treated with steroids. In the third trimester she presented with Eclampsia and was treated with antihypertensives and magnesium sulphate. She delivered vaginally a live preterm child and was discharged healthy along with her baby.

Keyword: *Mixed Connective Tissue Disorder (MCTD), ribonucleoprotein (RNP), ANA , Eclampsia*

INTRODUCTION

Connective tissue diseases are a group of disorders of unknown etiology. Their classification depends upon identifying clusters of clinical and laboratory features. There are problems to classify individual patient into one of the defined autoimmune rheumatic disease because of the overlapping nature of most of the clinical and laboratory features and the signs and symptoms occur sequentially over time. However, identification of specific disease is valuable for treatment, prognosis and research ^[1]. Mixed Connective Tissue Disorder (MCTD) was first described by Sharp et al (1971-72) as an overlap syndrome with features of SLE, Systemic

sclerosis, polymyositis and rheumatoid arthritis in patients who had antibodies to extractable nuclear antigen and Ribonucleoprotein (RNP) specificity ^[2]. It is predominantly a disease of females and many times diagnosed for the first time during pregnancy. The incidence of MCTD is 1 in 22,000 pregnancies ^[3]. Conflicting reports describe the effects of pregnancy on the course of MCTD and the effects of MCTD on the foetus. Therefore we are presenting this rare case report.

CASE REPORT

A 23 year old primigravida developed bleeding gums and petechiae at 8 weeks of gestational age

and was found to have thrombocytopenia. Few weeks later she had synovitis with effusion in bilateral knee joints and raynaud's phenomenon with puffy fingers. ANA screening showed her to be ANA positive with coarse speckled pattern. There was no family history of connective tissue disorders. She was strongly positive for U1RNP and Ro-52. Hence she was diagnosed as a case of Mixed Connective Tissue Disorder (MCTD) and was on regular steroids since then. At 30 weeks of gestational age patient got admitted with Eclampsia. At the time of admission she was drowsy with a Glasgow Coma Score of 8. She had pallor along with generalised oedema and was hypertensive (168/106 mm of Hg). Further examination revealed ecchymotic patches over left arm and left leg and purpuric spots all over body. Her investigations showed anaemia with thrombocytopenia, elevated ESR and proteinurea. Total count, Differential count, Liver function test, thyroid function test, renal function test, ECG and ECHO cardiography were normal. USG showed a single live intrauterine foetus of wt. 1.5kg. She was transfused packed red blood cells and platelets. After giving injection Magnesium Sulphate and antihypertensives she was induced with prostaglandins. She delivered vaginally a preterm live female child of weight 1.5kg.

After delivery her blood pressure remained under control on antihypertensives and she was discharged on fifth post natal day along with her child.

DISCUSSION

MCTD is predominantly a disease of females, with female to male ratio of 16:1. A study from Japan showed Incidence of 2.7 per 1, 00,000[4]. It is an autoimmune disease with autoantibodies affecting multi organs like skin, joints, kidneys, lungs, heart, liver and nervous system. There is both T-cell & B cell response with less immune complex formation. The characteristic lesions in the involved organs are intensive obliterative, proliferative vascular lesions in large, medium and small vessels with less inflammatory infiltrates [1]. The exact cause of mixed connective tissue disease is unknown. In

some cases it may be genetic as it sometimes occurs in patients with a family history of connective tissue disorders. . Some environmental agents like Vinyl chloride and silica have been associated with MCTD. Drug induced MCTD may occur in TNF-alpha therapy [5]. The "classic" symptoms of MCTD are: Raynaud phenomenon, swollen "sausage-like" fingers, myositis, synovitis, serositis and increase in proteinurea. The most common skin change is the Raynaud phenomenon, which usually presents early in the course of the disease as in our case [6]. The diagnosis is based on complaints, symptoms and organ involvement and on the presence of anti-U1RNP antibodies in high titre. It is the only connective tissue disease for which one specific type of antibody is necessary to make a diagnosis. The present case also showed strong positivity to anti-U1RNP. Nonspecific hematologic and laboratory abnormalities are common in MCTD: approximately 75 percent of patients have a low-grade anaemia. Pregnancy affects maternal MCTD onset and activity. In ten patients reported in a study, five had their disease onset during pregnancy and three patients had disease flare during pregnancy [7]. During pregnancy it may progress to abortion, preeclampsia, HELLP syndrome, eclampsia, premature delivery, anaemia, thrombocytopenia, intrauterine growth retardation (IUGR), and DVT [8],[9]. Maternal mortality is also increased due to renal failure, hypertension and cardiopulmonary complication. The mechanism for pregnancy complications is probably an autoimmune reaction against placental tissues, as immunostaining studies show deposits of fibrinogen, IgG, IgM, IgA, and complement 3 (C3) localized to the trophoblast basement membrane [7]. There are cases reported of babies with chondrodysplasia punctata born to MCTD mothers with high titre anti-RNP [10]. The fetal wastage is increased in MCTD. Just like neonatal lupus syndrome due to crossing of maternal antibodies the foetus can have foetal hemolytic anemia, thrombocytopenia, leukopenia and congenital heart block [7].

The limited data that is available suggests a small risk of disease flare during pregnancy and many authors conclude that the complications are not more than the expected rate. None of the reports in literature account for patient or physician preference in mode of delivery. The case emphasises that with early intervention and adequate treatment with steroids the female with MCTD can go through her pregnancy with minimal complications and deliver a healthy child.

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