Abstract:

Idiopathic myelofibrosis is a rare myeloproliferative disorder characterized by marrow fibrosis, extramedullary hematopoesis and splenomegaly. The cause of this disease is not known and no specific therapy exists. Among all myeloproliferative disorders the prevalence of Idiopathic myelofibrosis in women of child bearing age group is very less and the prognosis is variable but generally poor. So far only eight pregnancies in four patients with Idiopathic myelofibrosis have been reported in the literature. We report such a rare case of Idiopathic myelofibrosis in pregnancy who had a favorable outcome with close antepartum surveillance inspite of increased perinatal and maternal risks.

Keyword: Idiopathic myelofibrosis, women, pregnancy, child bearing age group

CASE REPORT:

A 22 yr old third gravida with H/o 3 months amenorrhoea presented to our OPD with severe anaemia. She was admitted and evaluated. She had H/o 2 abortions at 5 months amenorrhoea before this pregnancy. Clinically she had massive splenomegaly without ascites with mild hepatomegaly without generalized lymphadenopathy. Her reports showed Hb-3.7g%, WBC-2000 cells/cumm, platelets-30,000/cumm. LFT, RFT and LDH were normal. ANA & Coomb’s test were negative. Portal vein doppler was normal. Peripheral smear showed normochromic normocytic RBC’s with few macrocytes and target, tear drop cells. Bone marrow biopsy revealed megakaryocytosis with preserved erythroid and granulocytic progenitors with patchy fibrosis. She was periodically admitted and treated supportively with repeated packed cell and platelet transfusion. Her hematological parameters improved with supportive therapy alone and pregnancy continued till term. She delivered a healthy boy baby of 2.75 kg by LSCS done for fetal distress. After post partum period patient
was referred to Hematology department for further management.

**DISCUSSION:**

Idiopathic myelofibrosis is a chronic myeloproliferative disorder characterized by splenomegaly, immature granulocytes and erythroblastosis in the blood, distorted tear drop shaped red cells and bone marrow fibrosis. Idiopathic myelofibrosis is the least prevalent of all the myeloproliferative diseases in women of child bearing age group (0.02-0.06/1,00,000) [1]. The prognosis is variable but generally poorer than other myeloproliferative disorders. Even though pancytopenia & leukemic transformation is common in primary myelofibrosis, thrombosis is also a dominant clinical complication [7]. Most of the patients are asymptomatic at presentation and the disease is usually detected by the presence of splenic enlargement and abnormal blood count. In Idiopathic myelofibrosis the blood smear shows the characteristic feature of extramedullary hematopoiesis i.e tear drop cells, nucleated red cells, myelocytes, promyelocytes. Myeloblasts may also be present. WBCs and platelets may be increased/decreased/normal. Marrow is usually in aspirable due to fibrosis. Biopsy will reveal a hypercellular marrow with trilineage hyperplasia and in particular increased number of megakaryocytes in clusters and with large dysplastic nucleus. Autoimmune abnormalities such as immune complexes, Antinuclear antibodies, Rheumatoid factor or a positive Coomb’s test can also be observed in Idiopathic myelofibrosis. Cytogenetic analysis of blood is useful both to exclude chronic myeloid leukemia and for prognostic purpose in Idiopathic myelofibrosis[6]. Presence of complex karyotypic abnormalities in Idiopathic myelofibrosis has poor prognosis. Idiopathic myelofibrosis has to be differentiated from Polycythemia vera and Chronic Myeloid Leukemia because similar clinical picture can be observed in all three conditions. In this case the presence of anaemia, leucopenia, thrombocytopenia, splenomegaly, presence of tear drop cells, target cells, myelocytes in peripheral smear, hypercellular marrow with megakaryocytosis and patchy fibrosis favours the diagnosis of Idiopathic myelofibrosis and excludes Polycythemia vera and Chronic Myeloid Leukemia. As thrombosis is an important complication of Idiopathic myelofibrosis (Cervantes et al, 2006)[2] the H/o two abortions in this patient may be due to placental infarction which might have caused due to thrombosis. Similar h/o of abortions was also noted in pregnant women with Idiopathic myelofibrosis in a study by Gotic et al (2001)[3] and Sameer tulpule et al (2008)[5]. In his study, Sameer tulpule successfully managed these patients with aspirin and low molecular weight heparin who presented with thrombocytosis to avoid recurrent pregnancy loss. Even though our patient had h/o recurrent pregnancy loss, aspirin and LMWH was not given as she presented with thrombocytopenia instead of thrombocytosis. As far as treatment is concerned no specific treatment strategy exist for Idiopathic myelofibrosis. Anaemia can be corrected with recombinant erythropoietin and packed cell transfusion. Erythropoietin may worsen splenomegaly and will be ineffective if the serum erythropoietin level is >125mU/L[6]. Erythropoietin was not given to this patient as it may worsen splenomegaly. Splenectomy can be done only in urgent indications or in selective cases like symptomatic splenomegaly, mechanical
discomfort, refractory thrombocytopenia, hypercatabolic symptoms and portal hypertension (Lana macuknovic-golubovic et al,2004)[7]. Splenectomy was not done in our patient as it was not indicated. The role of alpha-interferon in Idiopathic myelofibrosis is still undefined. But successful use of alpha interferon in pregnant women with primary myelofibrosis was documented by Sameer tulpule et al[5] and Gotic M et al[3] in their study. In this case hematological parameters were corrected with supportive treatment like packed cell transfusion and platelet transfusion. Patient general condition improved and pregnancy was continued till term with close antepartum surveillance and a normal healthy male baby was delivered by LSCS done for fetal distress.

Conclusion:
Inspite of increased perinatal and maternal risk, favourable outcome is possible with close antepartum surveillance in pregnant women with Idiopathic myelofibrosis.

REFERENCES:


6 Harrison’s textbook of Internal Medicine 17th edition (p674 – p675)