

PREGNANCY WITH FACTOR XIII DEFICIENCY

(A Case Report)

by

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Factor XIII also known as the fibrin stabilizing factor was first recognised by Duckert in 1960. It acts in the common pathway of coagulation where it forms stabilizing, bonds with fibrin strands. It is an α_2 globulin with a molecular weight of 3,20,000. Only 32 cases have been reported throughout the world literature.

CASE HISTORY:

Mrs. S. F. a primigravida was referred on 29-7-1980 as a case of pregnancy with a bleeding disorder.

Her menarche was at the age of 17. Her first period had been profuse and prolonged and had necessitated hospitalization. Her menstrual cycles were regular and uneventful thereafter. Her last menstrual period was on 20-1-1980 and her expected date of delivery was 27-10-1980. Her past history indicated several bleeding episodes following trauma for which blood transfusions had to be given off and on. In the family history her brother had died due to some bleeding disorder.

General examination of the patient showed that her pulse was 78 beats per minute, regular. Blood pressure 120/80 mm. of mercury. There was no odema of the feet. Respiratory and

cardiovascular systems were normal. On obstetric examination the fundal height was 30 weeks size uterus. Breech presentation, fetal heart sounds were 120 beats per minute, and regular.

On investigation, hemoglobin was 12 gms. per cent, blood V.D.R.L. — negative, blood group — A Rh positive, bleeding and clotting time were normal. Fibrinogen levels were 300 mgm. per cent. Prothrombin time was 11.5 seconds, platelet count was normal. Clot solubility test with 1% monochloroacetic acid was positive and so she was diagnosed as having factor XIII deficiency. In spite of repeated requests patient was unwilling to remain in the hospital and she was discharged on 2-8-1980.

She was readmitted on 5-8-1980 with pain in the abdomen and minimal bleeding per vaginum. On examination, her pulse was 80 beats per minute, blood pressure — 120/80 mm. of mercury. There was no edema of the feet. On obstetric examination, the uterus was contracting every 3 minutes and fundal height was 32 weeks. Breech presentation, left sacro-anterior, fetal heart sounds were regular, 132 beats per minute. On vaginal examination the cervix was fully dilated and effaced, membranes had ruptured and station of breech was at +2. She had an assisted breech delivery. Following delivery, 10 units of pitocin was given prophylactically. She was also given one bottle of plasma. The baby weighed 1.8 kilograms, sex female, apgar score at the end of two minutes was ten. Cord blood examination showed that the blood group was AB Rh positive. All the coagulation factors were within normal limits. The puerperal period was uneventful and she was discharged on the eighth day of the puerperium.

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Further follow up of the patient could not be done however, her husband informed us that on 1-11-1980 she had developed severe headache, for which on hospitalization she was diagnosed as having subarachnoid haemorrhage. She died within 24 hours of hospitalization.

Discussion

Factor XIII deficiency is transmitted as autosomal recessive. Though a rare disorder, its diagnosis is simple with urea or monochloroacetic acid clot solubility test. Bleeding from the umbilical cord is very common. This alone should forewarn an obstetrician to investigate further. Hemorrhage into the central nervous system has

been a serious and lethal complication and is significantly more common in this deficiency as compared to other deficiency disorder.

Conclusion

A case on Factor XIII deficiency with pregnancy has been reported.

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