

A Case of Marfan Syndrome with Ruptured Ectopic Pregnancy – Case Report.

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Abstract:

- Marfan syndrome is an inherited autosomal dominant connective tissue disorder due to multiple mutations of fibrillin-1 gene present on long arm of chromosome 15 leading to defective fibrillin protein causing generalized tissue weakness . It may range from mild subclinical cases to severe disorder resulting in scoliosis, joint laxity, cardiovascular complications like mitral valve prolapse, aortic aneurysm or aortic dissection(which appears more commonly during pregnancy).
 - Incidence of marfan syndrome is 2 to 3 per 10,000 individuals and is with out racial or ethnic predilection. It affects both sexes equally.
 - Thus the syndrome can make a pregnancy fall in high risk pregnancy, that too with ruptured ectopic pregnancy.
 - We report a rare case of 26 year oldprimi, a known case of marfan syndrome who is on treatment for aortic dissection with ruptured ectopic and was surgically managed.
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I. Case Report

- A 26 year old primi presented in emergency department with complaints of acute lower abdominal pain with bleeding p/v preceded by 2 months of amenorrhea. On history she was a known case of marfan syndrome with aortic dissection and on regular medications for it. No other significant history.
- On examination she was conscious, looking pale, afebrile, with fair hydration, with pulse rate 104 bpm and blood pressure 100/60mmhg and saturation of 99% with room air. On abdominal examination – soft, tenderness present in right iliac fossa, no distention. On speculum examination, cervix and vagina healthy, no discharge. On vaginal examination, cervix downwards, uterus anteverted, normal size, right forniceal tenderness present, other fornices free, cervical motion tenderness present.
- All routine investigations were sent with TC-9500cells/cc , Hb-8.8gm% , other parameters normal. On ultrasound, a gestational sac in right fallopian tube with free fluid in pouch of douglas identified.
- The patient was shifted immediately to operation theatre and emergency laparotomy was proceeded with right partial salphingectomyunder spinal anaesthesia. She improved well. She was adviced to continue cardiac medications with regular follow up.

II. Discussion

- Pregnancy in patients with Marfan syndrome remains a controversial subject. The 2010 thoracic aortic disease guidelines advocate avoidance of pregnancy if the aortic route diameter exceeds 40mm and recommend prophylactic aortic route replacement in those who desire pregnancy.
- Pregnancy causes a slight increase in aortic route diameter. In long run, women with enlarged aortic route diameters at pregnancy, show a slightly accelerated aortic route growth and therefore will have elective aortic route surgery at a younger age. Prophylactic beta blocker therapy has become the standard medical approach for pregnant women with Marfan syndrome because it reduces haemodynamic stress on the ascending aorta and slows the rate of dilation. Vaginal delivery with regional analgesia and assisted second stage seem safe for women with aortic route diameter < 4cm. When the aortic route measures 4-5cm or greater elective cesarean delivery is recommended with consideration of post partum replacement of the proximal aorta with a prosthetic graft.
- Donnelly et al. did a prospective study with 98 women with an aim to assess the impact of pregnancy and the rate of aortic growth as well on short and long term clinical outcomes. Of the 199 pregnancies there were 170 live births, 26 spontaneous abortions and 2 ectopic pregnancies. In conclusion , women with

marfan syndrome, without previous cardiac complications seem to tolerate pregnancy well, upto an aortic route diameter of 45mm, with good clinical care before, during and after pregnancy. Pregnancy should be discouraged in women with previous aortic dissection because of high risk of aortic complications.

- In conclusion, all women especially with enlarged aortic route diameters, the pros and cons of pregnancy should be fully discussed as well as the alternatives (childlessness, adoption, surrogacy).

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