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Post-partum aortic dissection in a patient with previously undiagnosed Marfan syndrome.

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Introduction

- Aortic Dissection is a rare event in the general population (3-4/100,000 per year), even more so in women under 40 years old. [1]
- Half of this group are associated with pregnancy, which; together with having Marfan syndrome (MFS), are the two biggest risk factors in this population.[2] Aortic complications arise in 4% of pregnant women with MFS.[3]
- We present the case of a 38-year-old patient who presented with an acute aortic dissection five days postpartum.

Case Report

38-year-old woman with no significant medical history.

- Single episode of reduced fetal movements earlier in pregnancy.
- Delivery via a caesarean section (urgency category three) following induction at term + six days.
- Uneventful post-operative course and discharged the next day.

Five days post-partum

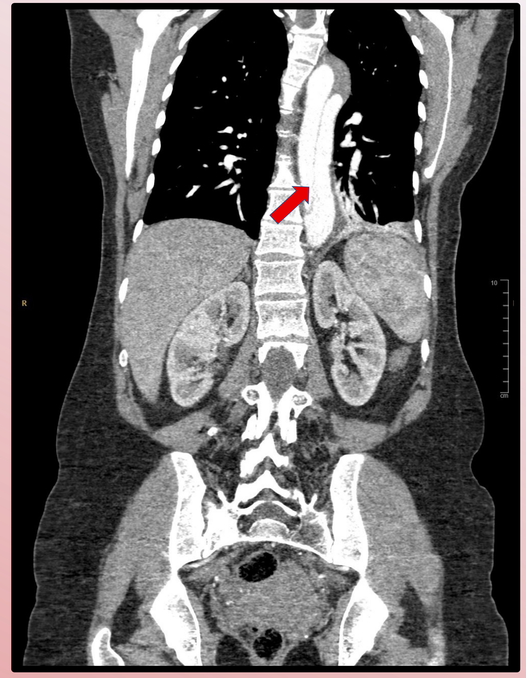
- Presented to A&E with back pain, nausea and pre-syncope.
- CT aorta demonstrated a large type-B aortic dissection, from left subclavian artery into both common iliac arteries.
- All major branches from true lumen.
- Admission to Intensive Care.
- Strict blood pressure management using IV Labetalol.
- Echocardiogram demonstrated no aortic root dilatation.

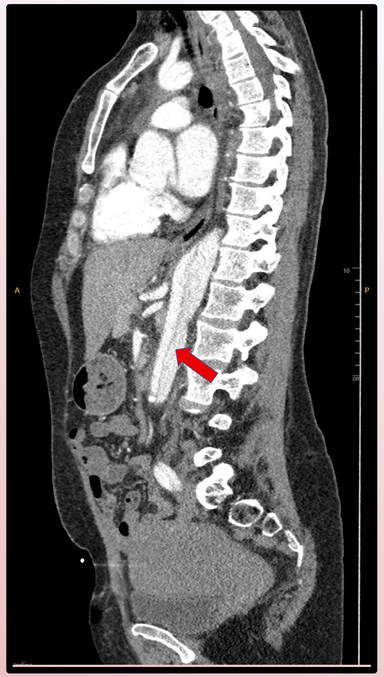
Discharge

- Oral nifedipine and doxazosin initially used to maintain blood pressure.
- Doxazosin stopped when able, to facilitate breastfeeding.
- Plan to introduce atenolol if needed.
- Discharged home five days after admission on oral medication having made a full recovery.

Ongoing management

- Clinical genetic medicine diagnosed patient with MFS.
- Specific mutation (FBN1) identified.
- Familial testing offered.
- Repeat CT aorta planned for three months time.
- Referred to specialist cardiothoracic services.
- No requirement for surgical intervention at present.

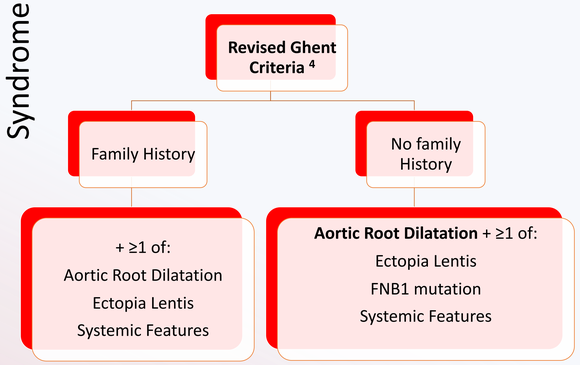




Aortic Dissection

- Acute aortic dissection is a life-threatening event involving a tear in the aortic intima allowing blood to create a false lumen between the intima and media layers.
- This can be divided into type A (involving the ascending aorta) and type B (limited to the descending aorta - distal to the left subclavian artery.)
- Patients can present with severe ‘tearing’ chest/back pain, hypovolaemia, limb blood pressure discrepancy, aortic regurgitation murmur, symptoms of end organ ischaemia including stroke, renal injury, limb ischaemia.
- Type A and some type B dissections (those with compromised blood supply to organs/limbs) are treated surgically as an emergency. Whereas uncomplicated type B dissections can be managed with strict blood pressure control using beta blockers initially, analgesia and close observation.

Diagnosing Marfan Syndrome



Discussion

- How would this patient’s care have been different if she was known to have MFS?
  - Antenatal planning involving the multidisciplinary team including obstetricians, cardiologists, anaesthetists, neonatologists. This would be tailored to the individual including obstetric history and degree of aortic dilatation.
  - Consideration of antenatal beta blockers and strict blood pressure management
  - Planning for delivery, aiming to minimise haemodynamic stress. This may include an early epidural with avoidance of prolonged pushing or an elective caesarian section.
    - N.B. patients with MFS have a high prevalence of dural ectasia, which should be excluded antenatally if considering an epidural.
  - Postnatal observation (unlikely to have been for 5 days).
- Given this, should echocardiogram and/or genetic investigation be considered for pregnant women who are of a ‘marfanoid’ appearance in order to increase antenatal diagnosis and management?

References

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