

A Gravid Marfan Syndrome Patient Presents with Aortic Dissection Undergoing Cesarean Section

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INTRO

- Marfan syndrome is an autosomal dominant genetic connective tissue disorder
- Marfan syndrome affects the FBN1 gene on chromosome 15 and fibrillin-1 production
- Many different organ systems can be affected including cardiovascular, musculoskeletal, and ocular issues
- Marfan syndrome may be associated with local anesthetic resistance

METHODS

- This case report is devoid of patient identifiable information and exempt from IRB review requirements per Medical College of Wisconsin policy
- Patient also gave consent to use this case as a case report

REFERENCES

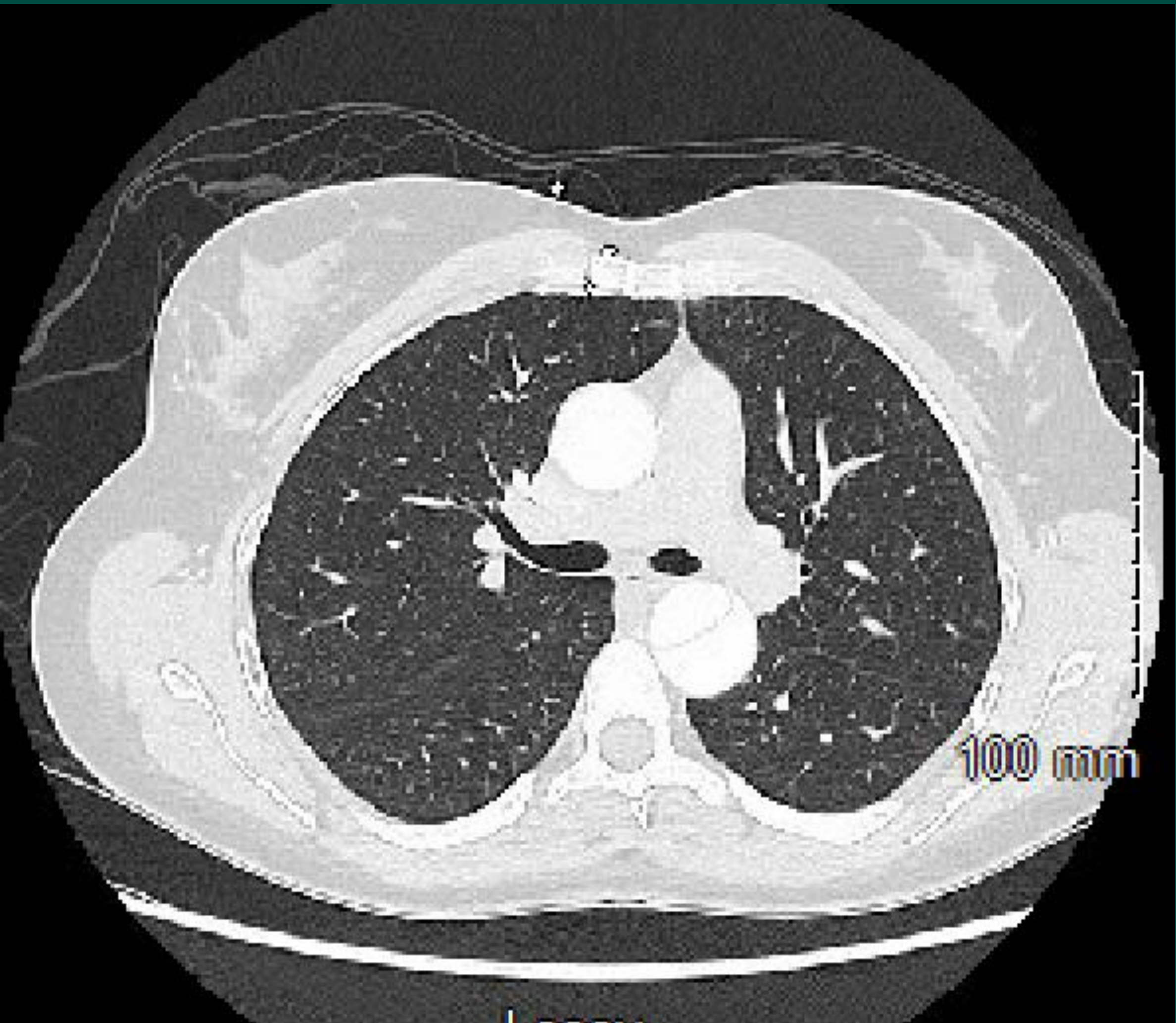
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Systemic and neuraxial local anesthetic resistance can occur in Marfan syndrome patients. The etiology and incidence of resistance remains unclear. One potential hypothesis for local anesthetic resistance is dural ectasia.



CASE REPORT

- 38-year-old gravid female 5’10” tall weighing 85.5 kg and BMI of 27 with history of Marfan syndrome, atrial septal defect status post repair, and aortic root aneurysm status post valve sparing aortic root replacement
- Patient presented with type B aortic dissection
- Decision was made to medically manage patient in ICU while fetus developed with plan for scheduled cesarean section at 34 weeks gestation
- At 32 weeks patient developed preeclampsia and decision was made to pursue cesarean section early
- Given the complexity of the situation and preference to avoid general anesthesia a combined spinal epidural was performed
- The first attempt at a combined spinal epidural was performed at L3-4 level. Loss of resistance was achieved with saline using a 17-gauge Tuohy needle. A spinal was attempted using a 25-gauge pencil tip spinal needle; however, were unable to get return of cerebrospinal and therefore no intrathecal medications administered
- Given this, the decision was made to thread the epidural catheter and the epidural was titrated slowly with 2% lidocaine with epinephrine.
- A total of 20cc of 2% lidocaine with epinephrine was administered. This resulted in some isolated low lumbar coverage but not enough for surgical coverage. The decision was then made to remove the epidural catheter and attempt combined spinal epidural again. This time the combined spinal epidural was placed at the L2-3 level. Loss of resistance was achieved with saline and a spinal was performed using a 25-gauge pencil-point spinal needle.
- Cerebrospinal fluid was obtained before, during, and after intrathecal administration. A total of 1.6 ml of 0.75% hyperbaric bupivacaine, 15 mcg of fentanyl, and 150 mcg of durapmorph were administered intrathecally.
- After administration of intrathecal medications the epidural was not able to be successfully threaded. Despite presumed intrathecal administration no change in level was achieved. Both attempts at combined spinal epidural were performed by an attending anesthesiologist.
- Given this culmination of events there was concern for local anesthetic resistance in the setting of known Marfan syndrome and decision was made to convert to general endotracheal anesthesia. The cesarean section was performed without complication and five days later the patient underwent thoracic endovascular aortic repair without complication.

