

Vascular Ehlers-Danlos Cesarian Section

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Background

- Vascular Type Ehlers-Danlos Syndrome (vEDS) is a result of a mutation in COL3A1 gene encoding type III collagen
- Carries elevated risk of vascular, uterine, and bowel complications, especially in pregnancy
- Pregnancy is contraindicated due to increased risk of mortality (fatal in 1 in 20 women)¹

Case

- **38-year-old G1P0 at 32 weeks** with history of asthma and prior spontaneous **R renal artery dissection**
- Presented with spontaneous **left leg numbness/pain**
- Found to have:
 - **2 L common iliac artery dissections extending to common femoral artery (images 1 and 2)**
 - **1.7 cm Splenic artery aneurysm (image 3) and MCA aneurysm**
- Started on **anticoagulation and beta-blockade**
- Genetic counseling positive for **Vascular Ehlers-Danlos**
- Planned **cesarean section in main OR at 37 weeks** with **vascular surgery on standby for catastrophic vascular injury**
- Initially declined, then consented to **general anesthesia** after risk discussion
- **Arterial line** placed; induction and intubation uneventful
- **Infant delivered** via C-section; **APGARs: 1, 6, 9**
- **Both discharged POD 4** in stable condition

Decision: General vs Neuraxial Anesthesia

General Anesthesia (GA)

Pro

- Controlled general anesthetic for possible vascular injury and emergent repair
- Avoids emergent airway if needed to convert to general anesthesia
- Avoids sympathetic block that could potentiate hypovolemia caused by possible hemorrhage

Con

- Potential hemodynamic swings with induction and intubation
- Possible unstable C-spine in vEDS²
- Increased chance of aspiration during intubation in parturient

Neuraxial with Combined Spinal Epidural (CSE)

Pro

- Patient's original preference
- Allows awareness for mother during delivery
- Greater postoperative pain control abilities potentially improving hemodynamic control

Con

- EDS patients have been shown to have reduced sensitivity to local anesthetics³
- Increased epidural tissue laxity in vEDS makes dosing unpredictable and increases chance of dural rupture and PDPH^{3, 4}
- vEDS has increased risk of bleeding and epidural hematoma with neuraxial procedures⁴

Management

- Planned operation was performed in main operative room area as opposed to OB floor
- Vascular surgery originally planned to perform post-delivery vascular repair, but opted to remain on standby in case of catastrophic vascular complication
 - Hemodynamic goals provided by vascular surgery included SBP <130 mmHg and HR <80
- Multidisciplinary conversation regarding best anesthetic method; patient originally preferred neuraxial but ultimately consented for general anesthesia
- Preoperative arterial line placed after risk/ benefit discussion of vascular injury
- Miscommunication among teams led to induction of general anesthesia before OB team was ready and NICU team arrival
- Uneventful rapid sequence induction utilizing fentanyl and esmolol for hemodynamic control
- Dexmedetomidine infusion and esmolol boluses to maintain hemodynamic goals intraoperatively
- Qualitative blood loss of 791 mL
- Oxytocin infusion used after delivery to aid in increasing uterine tone

Discussion

Decision- Making

- The case highlighted the need for multidisciplinary decision-making to manage the patient and prepare for possible catastrophic vascular injury.
- General anesthesia was chosen as the most conservative approach after a thorough risk-benefit discussion, emphasizing preparation for the worst-case scenario.
- GA allowed the team to control as many variables as possible and limit potential complications caused by neuraxial anesthesia

Miscommunication

- Coordinating multiple teams in an unfamiliar setting (performing a C-section in the main OR instead of the OB OR) led to unique challenges.
- A miscommunication occurred, leading to the induction of anesthesia before the surgical team was fully ready.
- Despite this, quick action led to good outcomes.
- A postoperative debrief was held, and a Quality Improvement (QI) project was initiated to improve communication for cesarean sections

References

1. Wright GL, Wen T, Engel DJ, Guglielminotti J, Andrikopoulou M, Booker WA, D'Alton ME, Friedman AM. Delivery Outcomes and Postpartum Readmissions Associated with Ehlers-Danlos Syndrome. *Am J Perinatol*. 2024 May;41(S 01):e3045-e3051. doi: 10.1055/a-2185-4149. Epub 2023 Oct 4. PMID: 37793432.
2. Campbell N, Rosaeg OP. Anesthetic management of a parturient with Ehlers Danlos syndrome type IV. *Can J Anaesth*. 2002 May;49(5):493-6. doi: 10.1007/BF03017928. PMID: 11983666.
3. Arendt-Nielsen, L., Kaalund, S., Bjerring, P. and Høgsaa, B. (1990), Insufficient effect of local analgesics in Ehlers Danlos type III patients (connective tissue disorder). *Acta Anaesthesiologica Scandinavica*, 34: 358-361. <https://doi.org/10.1111/j.1399-6576.1990.tb03103.x>
4. Wiesmann, T., Castori, M., Malfait, F. *et al*. Recommendations for anesthesia and perioperative management in patients with Ehlers-Danlos syndrome(s). *Orphanet J Rare Dis* **9**, 109 (2014). <https://doi.org/10.1186/s13023-014-0109-5>

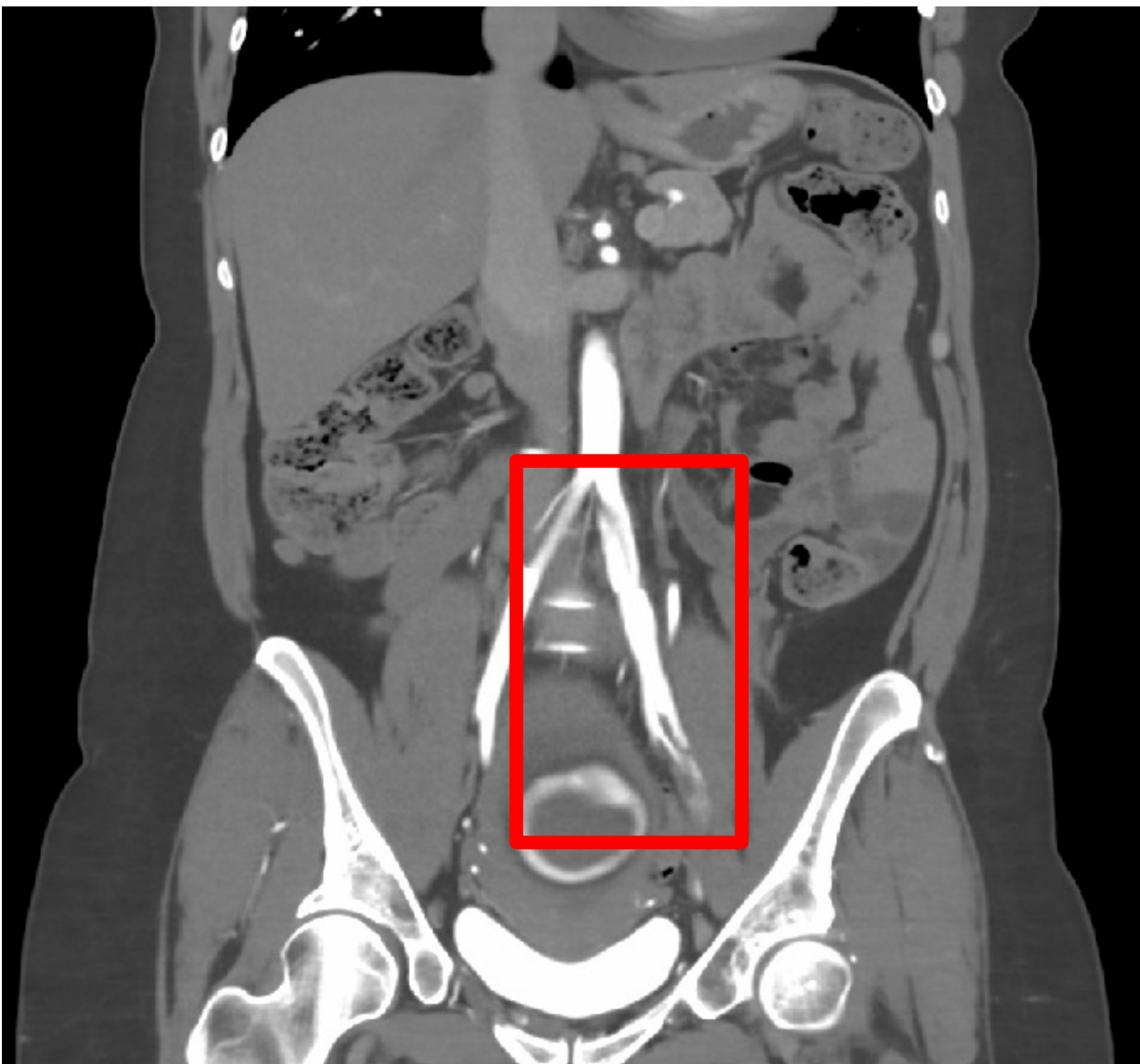


Image 1

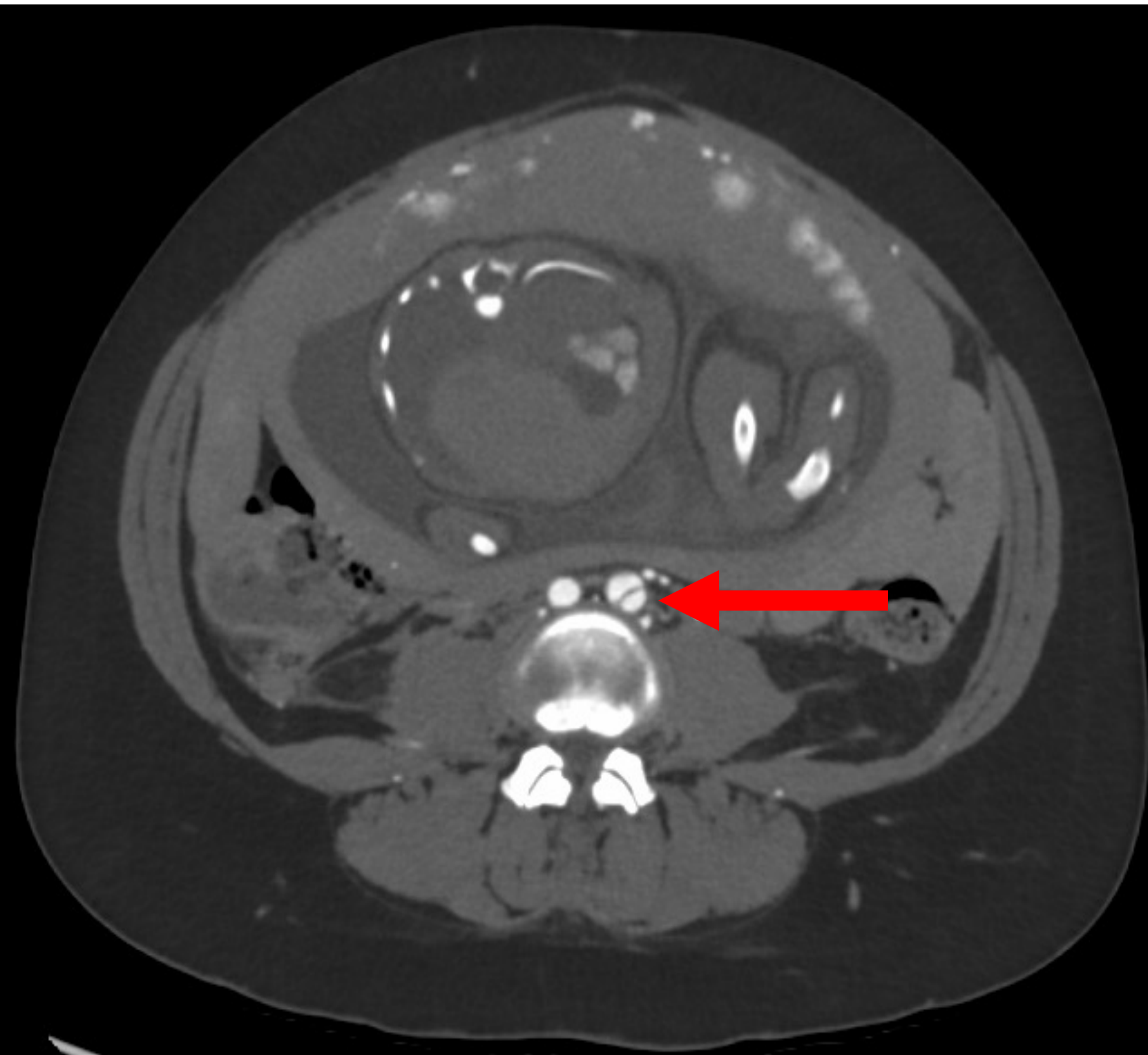


Image 2

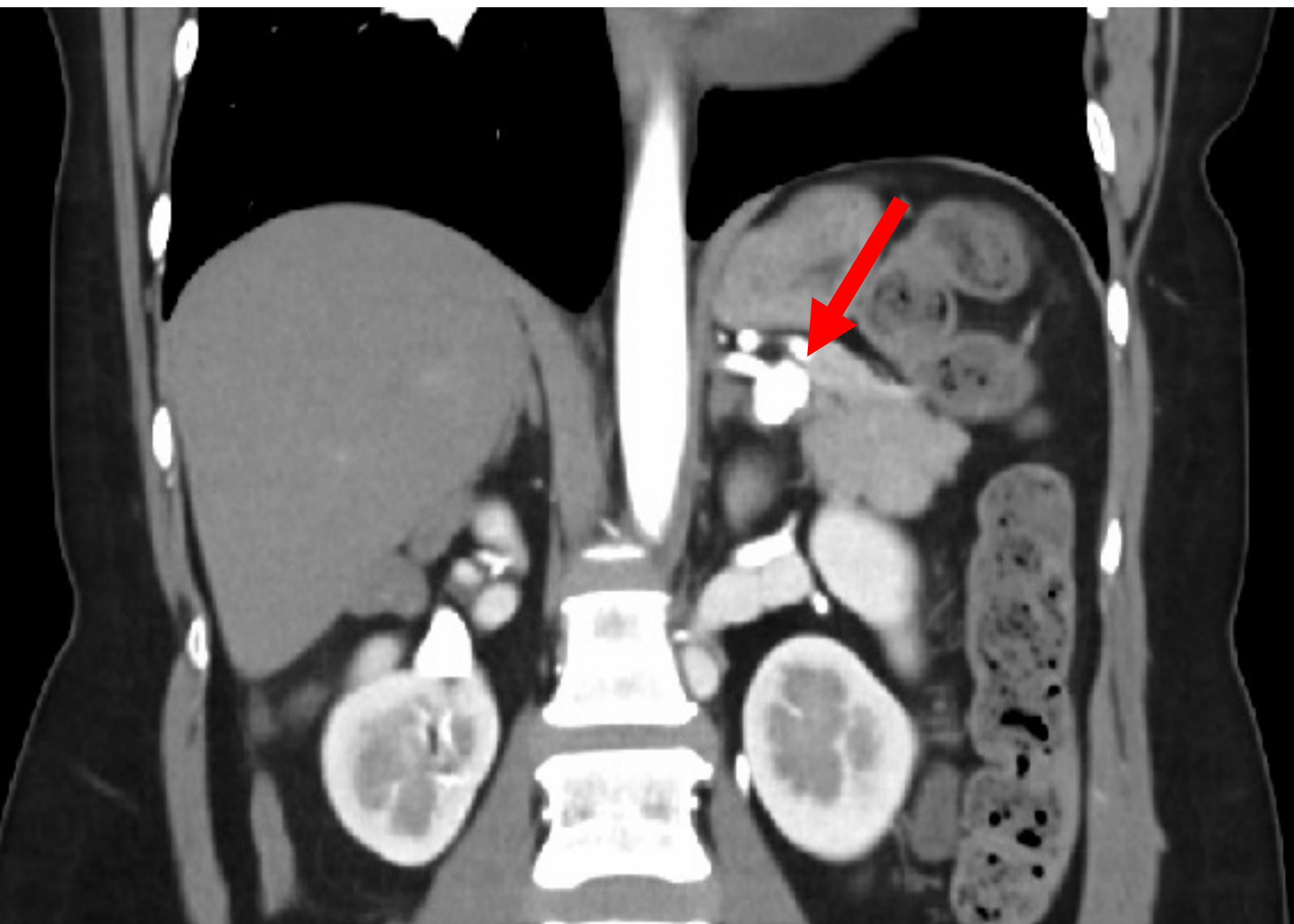


Image 3