

When Collagen and Clotting Fail: Catastrophic Postpartum Hemorrhage in a Patient with Ehlers–Danlos Syndrome and Functional Platelet Disorder

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BACKGROUND



Ehlers–Danlos Syndrome (EDS) is a group of connective tissue disorders associated with abnormal collagen synthesis, tissue fragility, and, in some cases, bleeding tendencies.



Obstetric complications include uterine rupture, poor wound healing, and postpartum hemorrhage (PPH), which may be worsened by coexisting hematologic disorders.



PPH, defined as $\geq 1,000$ mL blood loss within 24 hours after delivery, remains a leading cause of maternal morbidity and mortality worldwide.



In patients with platelet dysfunction or connective tissue disease, bleeding may be severe and refractory to standard measures. Interventional radiology (IR) techniques such as uterine artery embolization (UAE) can be lifesaving when conservative measures fail.

CASE PRESENTATION

A 36-year-old G2P1001 with hypermobility-type EDS, functional platelet disorder, chronic hypertension, and prior hemorrhage presented for scheduled cesarean with bilateral tubal sterilization under general anesthesia. Her first delivery was complicated by **uterine inversion** and **hemorrhage** requiring laparotomy, balloon tamponade, and transfusion of seven units PRBCs.

- Given her history, hematology recommended tranexamic acid (TXA) and **avoidance of intramuscular uterotonics**.
- Cesarean delivery was initially uncomplicated until **brisk placental bed bleeding** and intermittent **atony** occurred.
- Despite Bakri balloon placement, rectal misoprostol, TXA, and transfusion, quantified blood loss **exceeded 6 liters**.
- She received 8 units PRBCs, 5 units FFP, 3 units platelets, and 2 units cryoprecipitate.
- Persistent bleeding prompted **transfer to IR**, where bilateral UAE achieved hemostasis.
- Post-procedure, she stabilized with hemoglobin ~11 g/dL. She was discharged on postop day 4 on oral TXA with **close hematology follow-up**.

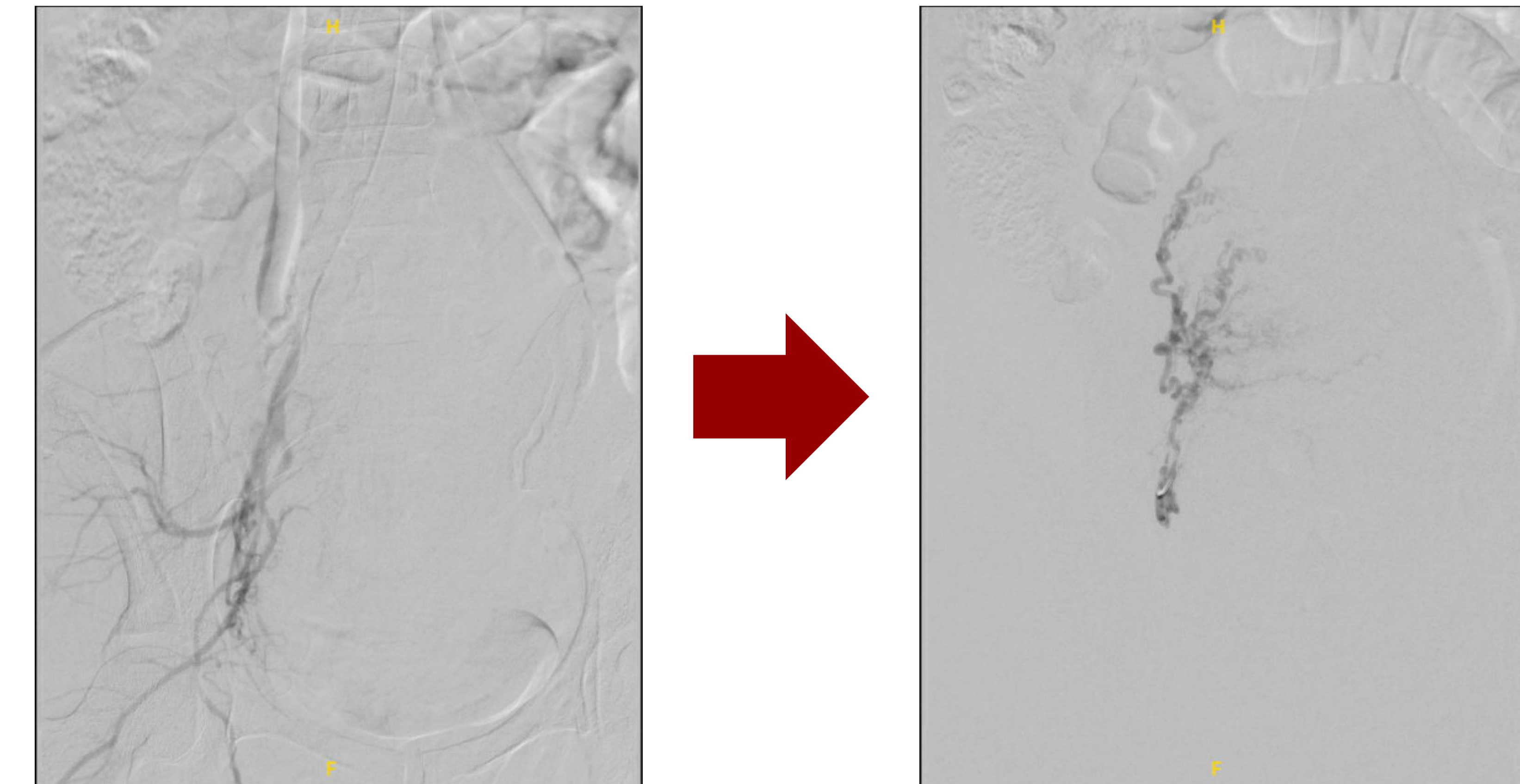


Image 1: DSI of right uterine artery before and after embolization

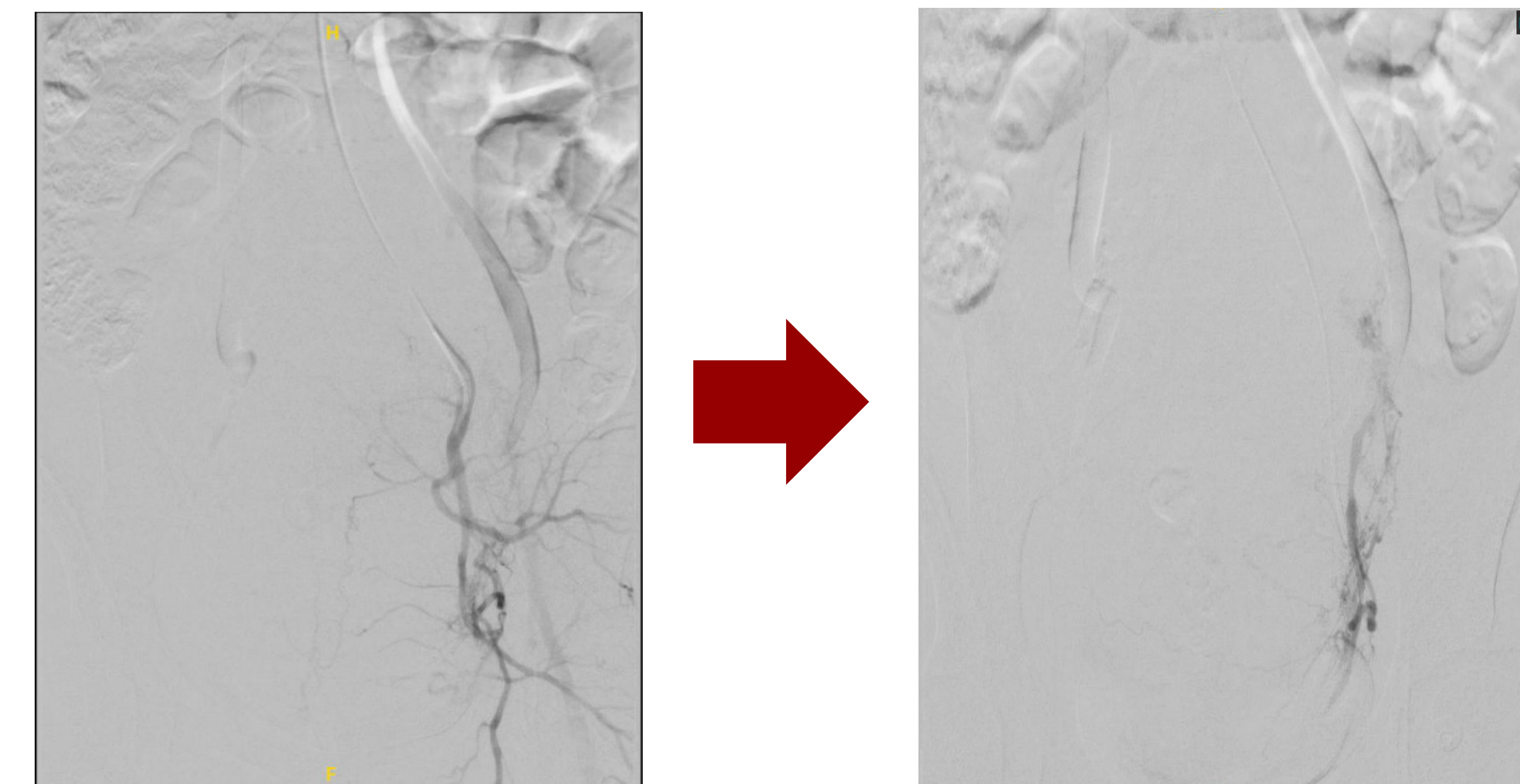


Image 2: DSI of left uterine artery before and after embolization



TEACHING POINTS

This case illustrates the synergistic hemorrhagic risks of EDS and platelet dysfunction, underscoring the importance of individualized hemorrhage management protocols in medically complex obstetric patients.

For anesthesiologists, perioperative care required anticipating massive transfusion, tailoring uterotonic therapy, and coordinating with hematology and IR.

Connective tissue fragility may impair vascular integrity and uterine tone, while platelet abnormalities magnify bleeding.

PPH guidelines emphasize early escalation, multidisciplinary involvement, and rapid deployment of surgical or IR interventions.

UAE avoided hysterectomy and preserved uterine integrity, particularly important in EDS due to wound healing concerns.

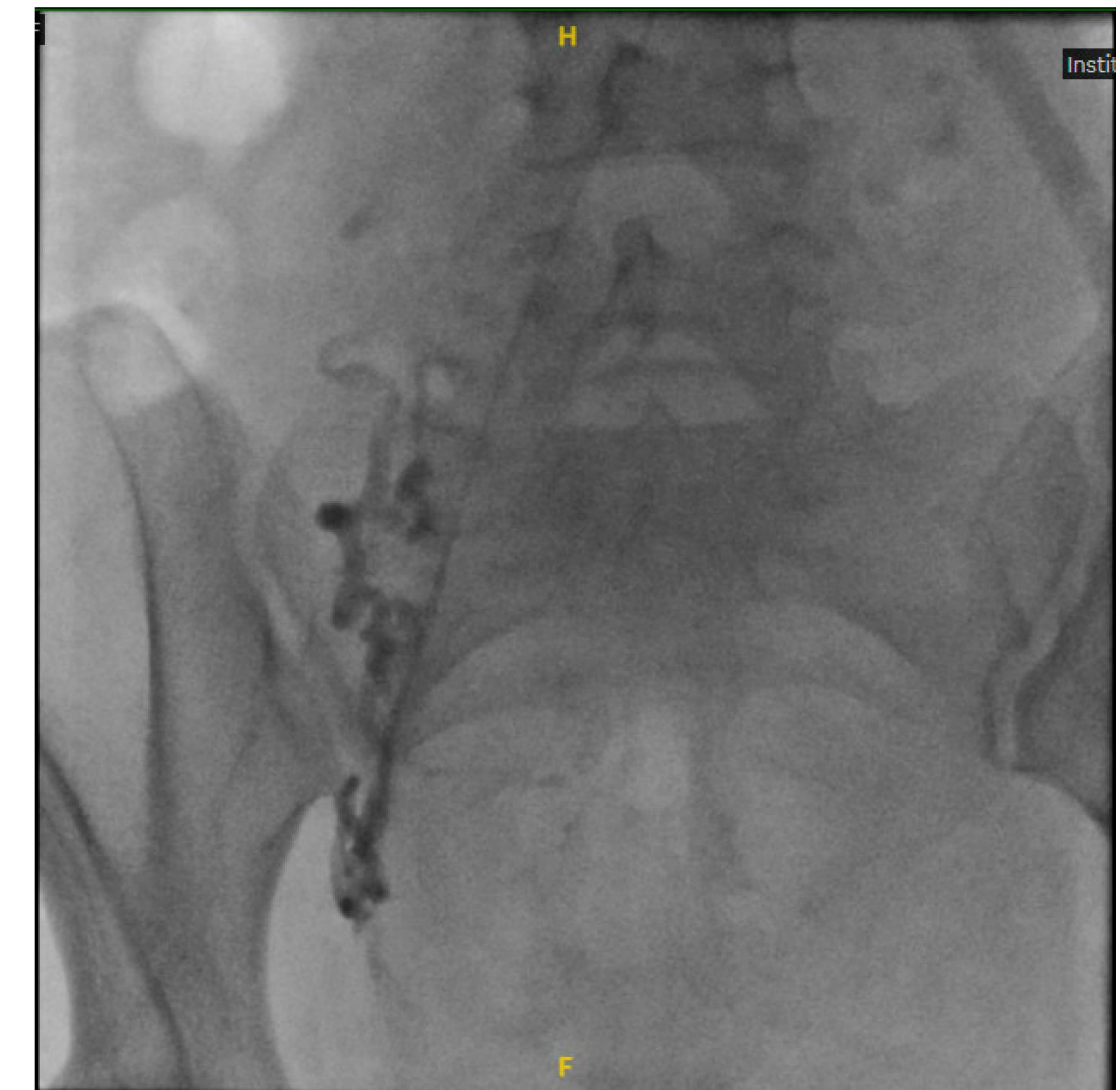


Image 3: Right uterine artery post-embolization

