

# Management of Desmoid Tumor in a Patient with Familial Adenomatous Polyposis

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## INTRODUCTION

- Desmoid tumor can occur sporadically or secondarily as a complication of Familial Adenomatous Polyposis (FAP)
- This locally invasive tumor rarely metastasizes, but can cause significant adverse effects due to rapid growth and compromise of surrounding organ architecture
- Due to its variable presentation, location and disease course, optimal management of desmoid tumor is controversial

## BACKGROUND

- The patient is a 50 year old female with a prior medical history significant for Familial Adenomatous Polyposis (FAP), Gardner Syndrome, and Human Immunodeficiency Virus (HIV)
- Importantly, at the time of her presentation, the patient was two years status-post open total proctocolectomy with an ileo-anal pouch and diverting ileostomy, due to a prior diagnosis of FAP. The patient reported that since her colectomy, she also suffered from loose non-bloody stools

## CLINICAL COURSE

### Chief Complaint

- Sharp left sided abdominal pain without radiation
- Increased abdominal girth
- Early satiety, worsening post-prandial pain and vomiting

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### Preliminary Imaging

- CT of the abdomen and pelvis showed a hypodense mass centered within the mesentery, extending from the pelvis to upper abdomen
- CT guided-needle biopsy of the mass showed spindle cell proliferation

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### Continued Course

- Medical management was sufficient for five years until the patient returned presenting with recurrent episodes of abdominal pain and fever
- Multiple trials of image-guided drainage and J-tube insertion were required for infected desmoid abscess

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### Surgery

- Goals were for adhesiolysis, abdominal wall fistula and desmoid tumor excision
- Several bowel loops were found within the desmoid tumor
- Many attempts were made for dissection while avoiding enterotomy
- After significant dissection, it was determined that further resection would risk short gut and malnutrition; surgery was discontinued

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### Clinical Exam

- Tender abdomen with marked distension
- Large, firm mid-abdominal mass identified
- Trace pedal edema

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### Disease Course

- Because the tumor was initially deemed inoperable due to size, medical management was initiated (sulindac + tamoxifen), to which initial response was positive
- HIV treatment also continued

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### Change in Management

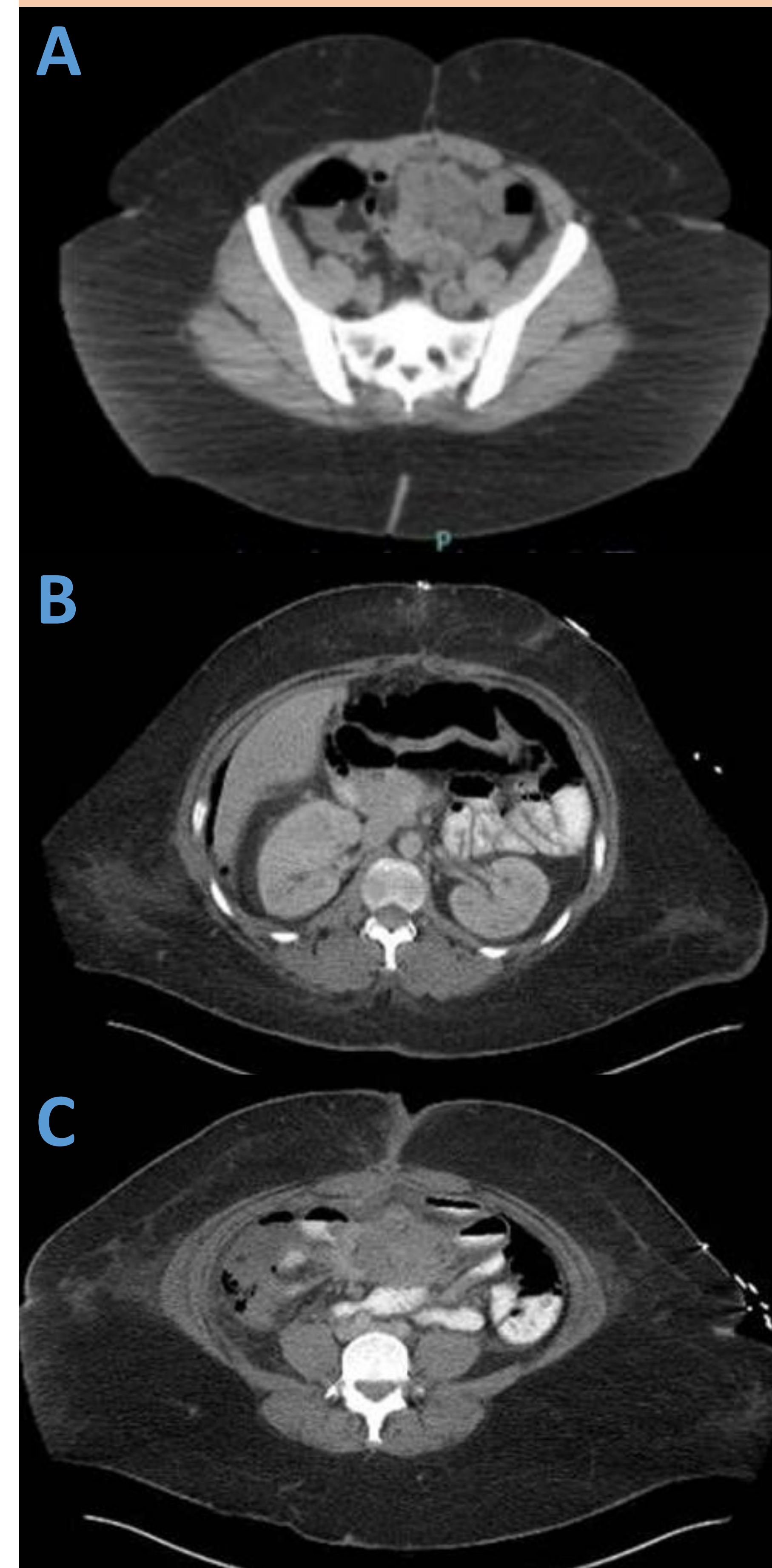
- Recurrent infection raised suspicion for a desmoid abscess (figure 1)
- Surgical evaluation was deemed appropriate due to decreased tumor size secondary to medical management and a 30 day RCRI class II risk of 6%
- Exploratory laparotomy was performed

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- Overall surgery resulted in removal of the abdominal wall fistula and adhesiolysis; partial desmoid tumor remained
- Post-operative treatment with IV antibiotics and patient controlled analgesia pump. Home bicitgravir was also restarted for HIV infection

## RADIOLOGY

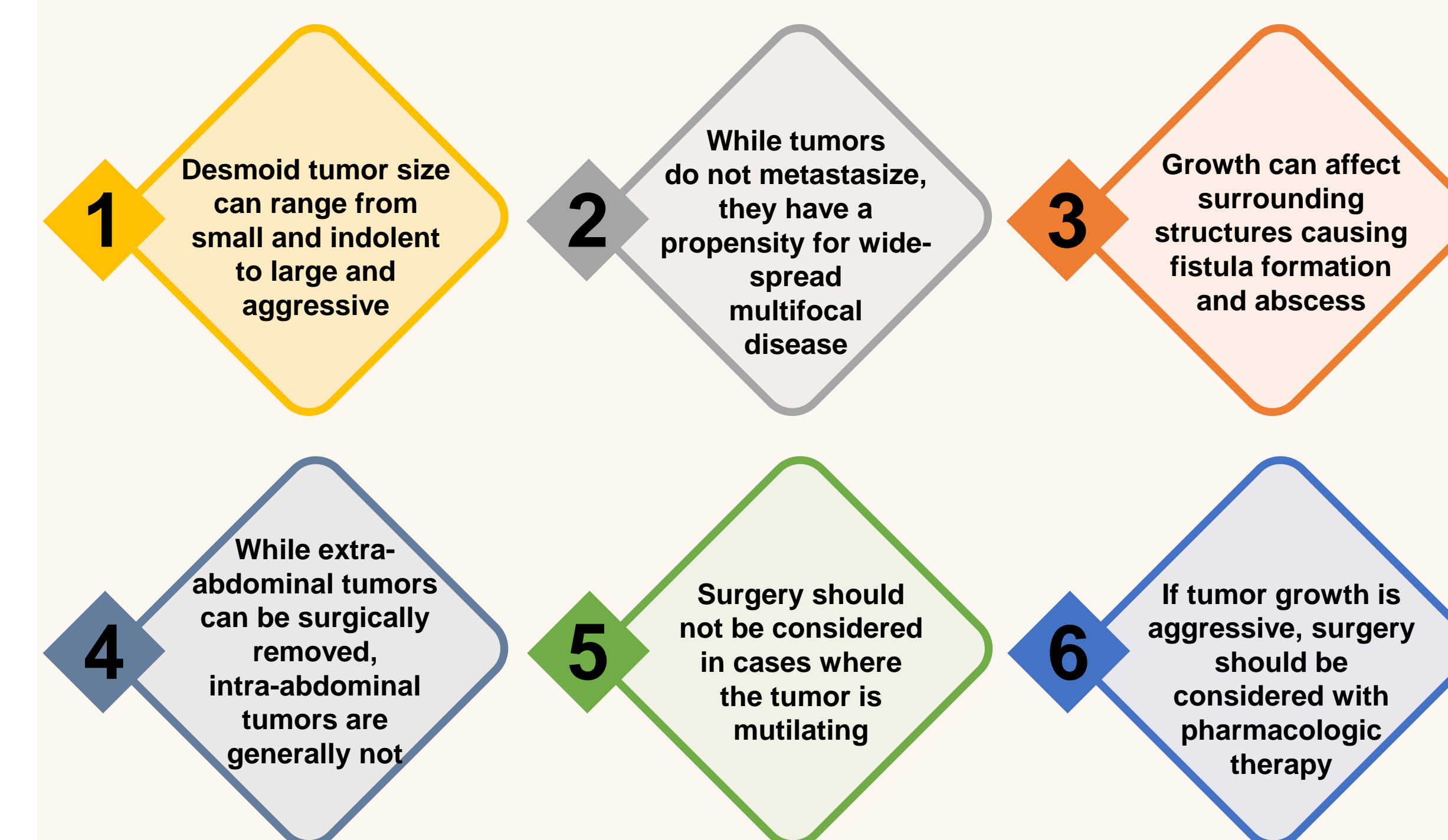


**Panel A:** CT showing an abscess within an unresectable desmoid tumor almost 5 years after initial diagnosis of tumor  
**Panel B & C:** CT with contrast 3 days following adhesiolysis and removal

## CASE DISCUSSION

- Post-operative course was uneventful until day three, when the patient developed a fever to 102.7° and abdominal pain, despite a clean and dry surgical wound
- CT showed pneumoperitoneum (figure 2), suspicious for intestinal perforation
- On day four, another exploratory laparotomy was done and no perforation, infection or other acute intra-abdominal pathology were identified
- Following procedure, no acute events developed post-operatively and the patient recovered
- Antibiotics and anti-retroviral therapy were resumed

## CONCLUSIONS



## REFERENCES

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