

Giant Unruptured Mucocoele of the Appendix Diagnosed in Pregnancy

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BACKGROUND

Diagnosis and management of adnexal masses in pregnancy is an ongoing challenge faced by obstetricians and radiologists alike. Pregnancy contributes several complicating factors when diagnosing such pathologies, including anatomical changes secondary to the gravid uterus, as well as limiting the imaging modalities available. Extensive collaboration between obstetricians, radiologists, neonatologists, anaesthetists and general surgeons, to ensure the best possible outcome for both mother and baby, is required.

An appendiceal mucocoele is a distended, mucus-filled appendix, often diagnosed incidentally during investigation of an unrelated complaint (1). A low-grade appendiceal mucinous neoplasm (LAMN) is a rare gastrointestinal malignancy, found in less than 0.3% of all appendectomy specimens (2). Symptoms vary depending on clinical manifestations, but most present with symptoms suggestive of acute appendicitis. The treatment course of LAMN is not well defined within the literature; however, an appendectomy-only approach in non-metastatic disease or neoplasms with uncertain malignant potential is recommended as a first line approach (2). If it ruptures, either spontaneously or during surgical resection, the release of mucin and epithelial cells into the peritoneal cavity may give rise to Pseudomyxoma Peritonei (PMP), which has an extremely high morbidity and mortality rate (3). PMP is the diffuse collection of gelatinous material in the abdomen and pelvis, usually associated with perforation of epithelial tumours of the appendix (3).

CASE

The patient was a female in her late 30's, gravida 4 para 2, with an incidental finding of a right sided solid-cystic mass on a first trimester ultrasound at 12 weeks' gestation, which was not seen in previous pregnancies. Serial ultrasounds throughout the first and second trimesters of pregnancy demonstrated progressive migration of the mass (which did not increase in size), with a scan at approximately 27 weeks' gestation confirming the mass to be separate from the patient's right ovary and fallopian tube. Sonographically, the mass was thought to be highly suggestive of an appendiceal mucocoele (with typical "gut signature"), with a small concern for malignancy and/or rupture. As a result, an abdominal and pelvic MRI was performed at 28 weeks' gestation, with findings in keeping with the diagnosis of an appendiceal mucocoele. At the time of the MRI, there was no evidence of rupture or malignancy. An urgent referral was made to the colorectal surgeons. It was recommended by the general surgical team that a joint caesarean section and appendectomy be performed between 37- and 38-week's gestation, to minimize the risk of rupture of the appendiceal mucocoele in the setting of vaginal delivery.

The patient remained asymptomatic throughout her pregnancy, despite the 6cm abdominal mass. An elective lower uterine incision caesarean section, bilateral salpingectomy and appendectomy were performed at 37+4 week's gestation.

Histopathology confirmed low grade appendiceal mucinous neoplasm (LAMN), with surgical margins uninvolved. On sectioning of the specimen, the appendix lumen was dilated up to 25mm, filled with gelatinous purulent material. As the tumour was resected completely, the colorectal surgeons did not consider intensive follow-up necessary. However, it was recommended the patient complete a colonoscopy for confirmation of resection and exclusion of co-existing lesions. This was subsequently performed, with a tubular adenoma with low-grade dysplasia confirmed on histopathology removed. A repeat colonoscopy in 5 years' time was recommended.

Key MRI features of an appendiceal mucocoele (4):

- Tubular right lower quadrant T1-hypointense and T2-hyperintense structure attached to the caecum (*image 1*)
- Mucin or hemorrhage can increase the signal intensity on non contrast T1-weighted images

CONCLUSION

The incidence of newly diagnosed adnexal masses in pregnancy continues to increase, as the use of ultrasound in pregnancy becomes more accessible (5). Although many of these masses found during pregnancy are functional or benign, accurate diagnosis and appropriate follow-up to exclude malignancy is strongly recommended. An article published in the *Journal of Gynaecological Surgery* in March 2021 emphasizes the importance of diagnosis and management of such masses for both maternal and neonatal outcomes (5). The importance of a multidisciplinary approach to these patients cannot be underestimated.

REFERENCES

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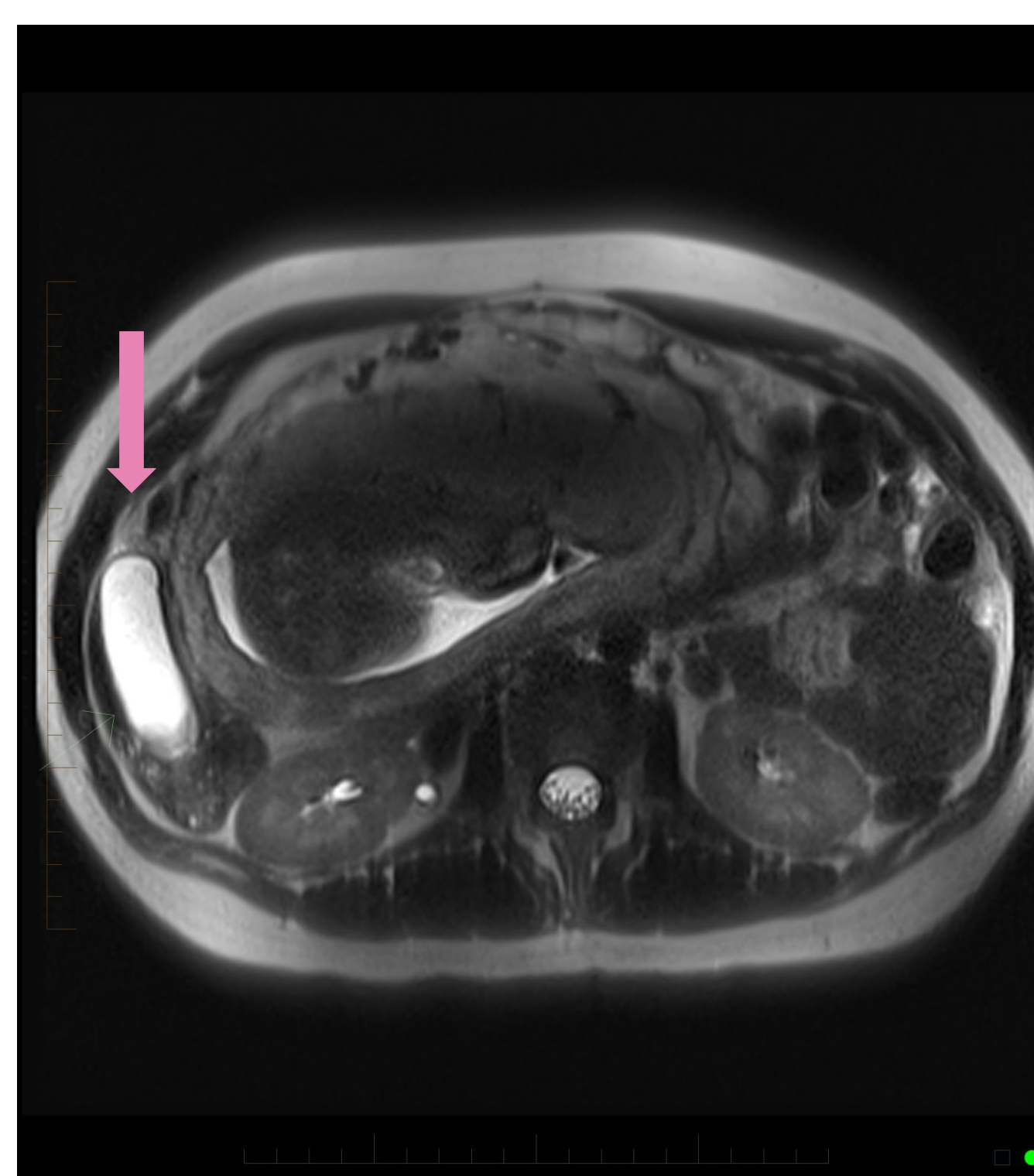


Image 1: Pelvic MRI demonstrating T2-hyperintense structure in right lower quadrant with signal characteristics compatible with mucin



Image 2: 6.28cm x 2.08cm right adnexal mass with a sharply margined border demonstrating the typical 'gut signature' of two echogenic lines with hypoechogenicity between these



Image 3: Tubular mass seen separate to ovary, kidney and liver at 27 weeks' gestation

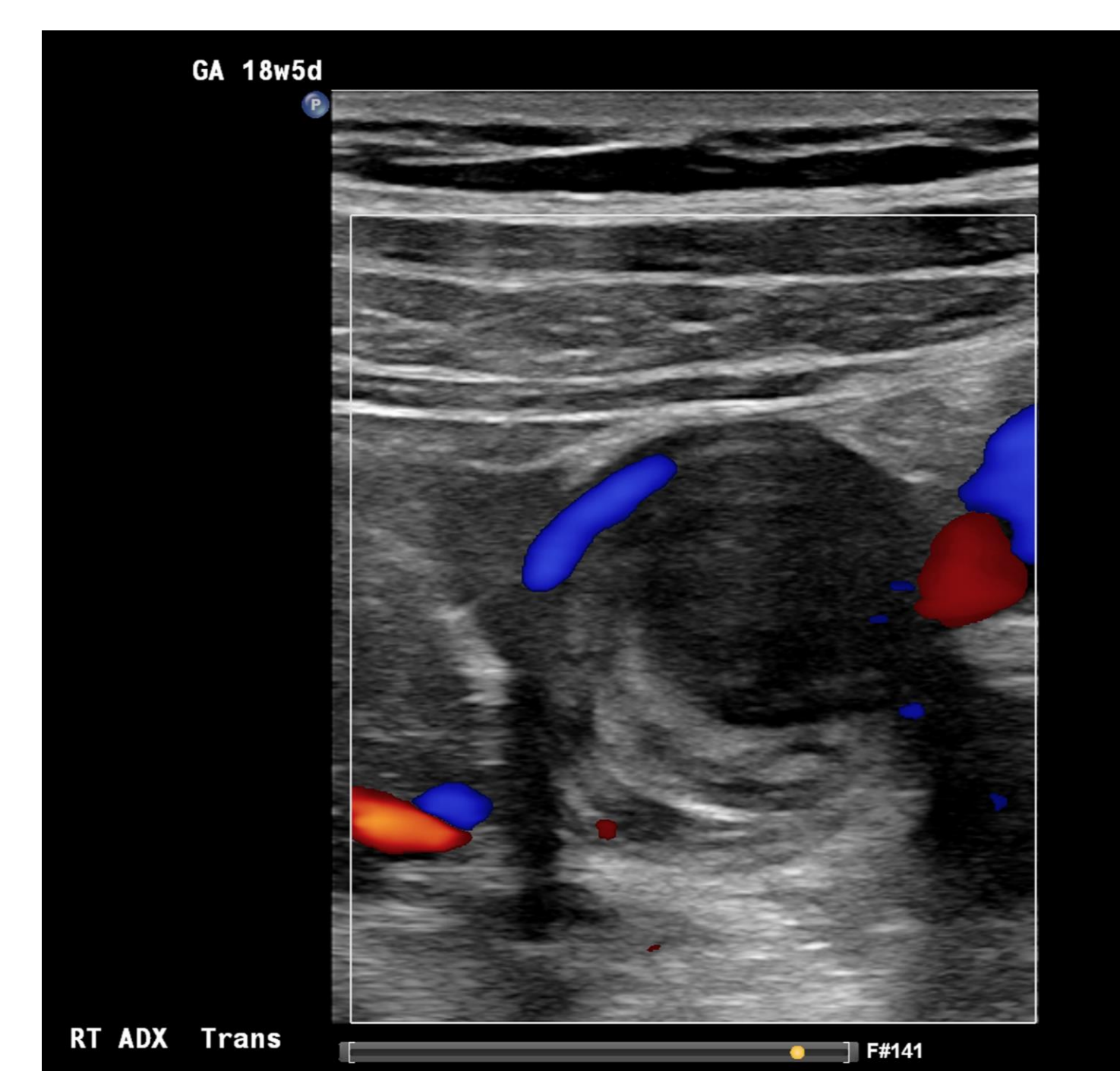


Image 4: Doppler ultrasound demonstrating minimal peripheral vascularity of the right adnexal mass