 DOI: 10.69729/aogp.v19i2a08

Incidental appendicular mucocele in a caesarean section – a case report

Mucocelo apendicular incidental em cesariana – relato de um caso

Joana Torres Silva[1], Mariana Leal[1], Filomena Barreto[2], Carla Duarte[1], Ana Maçães[1]
Department of Obstetrics and Gynaecology, Gaia/Espinho Local Health Unit, Portugal.
Department of Pathological Anatomy, Gaia/Espinho Local Health Unit, Portugal.

Abstract

We report a clinical case of a ruptured appendicular mucocele detected in a caesarean section due to suspicion of cephalopelvic disproportion (CPD). Histopathological examination of the specimen revealed a low-grade appendiceal mucinous neoplasm (LAMN) with peritoneal involvement. Abdominal and pelvic viscera examination during a caesarean section should be a crucial step if abnormal findings are seen during the procedure to identify and diagnose tumours, such as LAMN.

Keywords: Abnormal labour progression; Appendicular mucocele; Caesarean section; Cephalopelvic disproportion; Low-grade appendiceal mucinous neoplasm.

Resumo

Apresentamos um caso clínico de um mucocelo apendicular roto detetado numa cesariana devido a incompatibilidade feto-pélvica. O exame anatomopatológico da peça cirúrgica revelou uma neoplasia mucinosa do apêndice de baixo grau com envolvimento peritoneal. A revisão da cavidade abdominal e pélvica durante a cesariana deve ser um passo fundamental, se forem identificados achados suspeitos, na identificação de tumores, como a neoplasia mucinosa do apêndice de baixo grau.

Palavras-chave: Trabalho de parto estacionário; Mucocelo apendicular; Cesariana; Incompatibilidade feto-pélvica; Neoplasia mucinosa do apêndice de baixo grau.

INTRODUCTION

Appendicular mucocele is a rare condition referring to a mucus filled, distended appendix, encountered in 0.2 to 0.3 percent of appendectomies. Pathogenesis is dependent on its etiology, that can range from benign to malignant findings.

We report a clinical case of a pregnant woman who underwent a caesarean section due to suspicion of cephalopelvic disproportion (CPD), one of the causes of abnormal labour progression. During surgery, it was detected diffuse intra-abdominal mucin originating from a ruptured appendicular mucocele. Appendectomy was performed and histopathological examination revealed a low-grade appendiceal mucinous neoplasm (LAMN) with peritoneal involvement.

1. Department of Obstetrics and Gynaecology, Gaia/Espinho Local Health Unit, Portugal.
2. Department of Pathological Anatomy, Gaia/Espinho Local Health Unit, Portugal.

CASE REPORT

A 28-year-old healthy woman, gravida 2, para 0 (2G 0P), with an uneventful pregnancy, was admitted for labour induction at 41 weeks' gestation. Misoprostol was used for cervical ripening, and six hours after, oxytocin was initiated at 15 millilitres per hour. Amniotomy performed four hours after oxytocin initiation revealed clear amniotic fluid. Labour progression was normal until nine centimetres of dilatation, at which point an occiput posterior fetal position was detected. After four hours without any further progression despite corrective measures, a caesarean section was performed due to suspected CPD.

We performed a Pfannenstiel incision and opened the abdominal wall in layers. Copious amounts of yellow-tinged mucoid material were found in the peritoneal cavity. A male neonate (2755 grams) was delivered via Kiwi® vacuum-assisted extraction device due to significant insinuation on the maternal pelvis, with Apgar scores of 9, 10, and 10 at 1, 5, and 10 minutes, respectively. A meticulous pelvic inspection was performed, revealing a vesicular and friable pattern of uterine, annexal and small intestinal serosa and a ruptured tumoral mass at the appendix apex, suggesting a ruptured mucocele of the appendix. Appendectomy with invagination of colon wall was performed in collaboration with general surgeons. The specimen and mucin was sent to histopathological examination. Umbilical cord blood gas analysis was performed and was normal.

Histopathological examination of the specimen revealed an irregular, mucus-filled appendix (Figure 1). It had a serrated muco-secretory epithelium without atypia and a mucosa with multifocal ulceration and inflammatory reaction (Figure 2). Margins of surgical top of the specimen were clear. The examination of mucoid material revealed scattered muco-secretory epi-thelium cells (Figure 3). These histopathological findings suggested a LAMN, pT4a Nx M1b R0 (AJCC 8th Edition/UICC 2017)[6], with peritoneal involvement, due to perforation and extravasation of mucinous content.

The patient and her baby had an unremarkable postoperative course and were discharged home on the fourth post-caesarean day.

TABLE I. LITERATURE REVIEW OF LAMN DIAGNOSES DURING CAESAREAN SECTION.

Author	Maternal and gestational age	Clinical findings	Caesarean indication	Surgical and pathological findings	Staging	Postpartum treatment
Inubashiri *et al.* (2019)[1]	24 yrs 38 wks	None	Cephalopelvic disproportion	LAMN	CT scan 1 mo after caesarean	None
Yohannes *et al.* (2019)[2]	31 yrs 38 wks	None	Abnormal 2nd stage labour progression	LAMN with mucinous epithelium involving the serosa + appendiceal	Lost to follow-up	Lost to follow-up
Baron *et al.* (2020)[3]	31 yrs 40 wks	None	Abnormal 2nd stage labour progression	LAMN with PMP	CT scan 3 wks after caesarean	CRS/HIPEC
Ribeiro *et al.* (2024)[4]	36 yrs 41 wks + 1 d	None	Two previous caesarean	LAMN	–	–
Bowles *et al.* (2022)[11]	38 yrs 40 wks + 6 d	Cystic structure in RIF incidentally found	Fetal bradycardia	LAMN with extracellular mucin with inflammatory cells	CT scan + Colonoscopy	Cecectomy at the same operative time + Right hemicolectomy latter

CRS/HIPEC: cytoreductive surgery/heated intraperitoneal chemotherapy; CT: computed tomography; d: days; LAMN: low-grade appendiceal mucinous neoplasms; PMP: pseudomyxoma peritonei; RIF: right iliac fossa; Wks: weeks; Yrs: years.

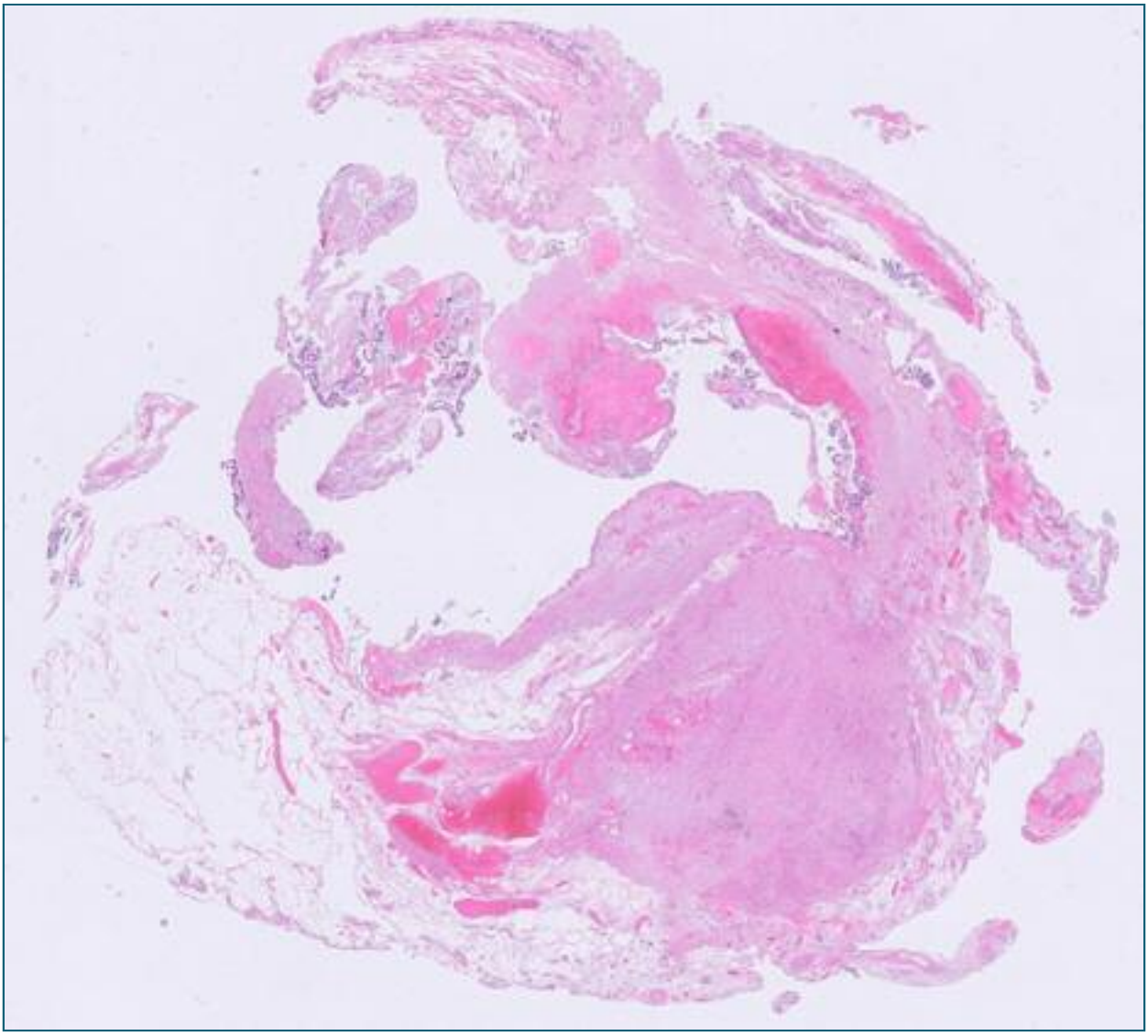

FIGURE 1. Ruptured appendix (hematoxylin and eosin stain, 20x).

The woman was referenced for a General Surgery consultation at our hospital. A postoperative thoracoabdominopelvic computed tomography (CT) scan revealed no locoregional or distant disease. She was subsequently transferred to the Peritoneal Disease Group of the Portuguese National Oncology Center and was considered to cytoreductive surgery (CRS) and heated intraperitoneal chemotherapy (HIPEC). The patient declined fertility preservation, stating no desire for future pregnancies.

At the time of this report, after six months of follow-up, the woman is asymptomatic.

DISCUSSION

Appendicular mucocele is a mucus-filled, distended appendix, encountered in approximately 0.2 to 0.3 percent of appendectomies, with a higher prevalence among females in their 50s and 60s. This condition can be either benign, including simple mucoceles or retention cysts, or malignant, encompassing serrated polyps, mucinous appendiceal neoplasms, and mucinous adenocarcinomas of the appendix.

Appendiceal mucinous neoplasms are characterized by low-grade, non-invasive epithelial proliferation,

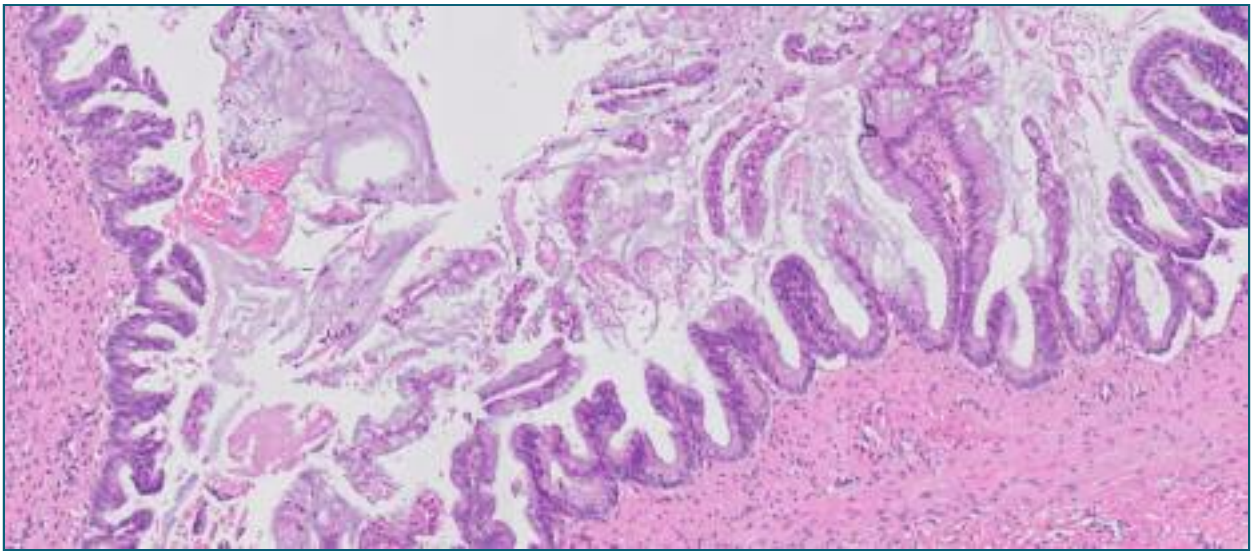

FIGURE 2. Low grade epithelial features in the absence of infiltrative growth (hematoxylin and eosin stain, 20x).

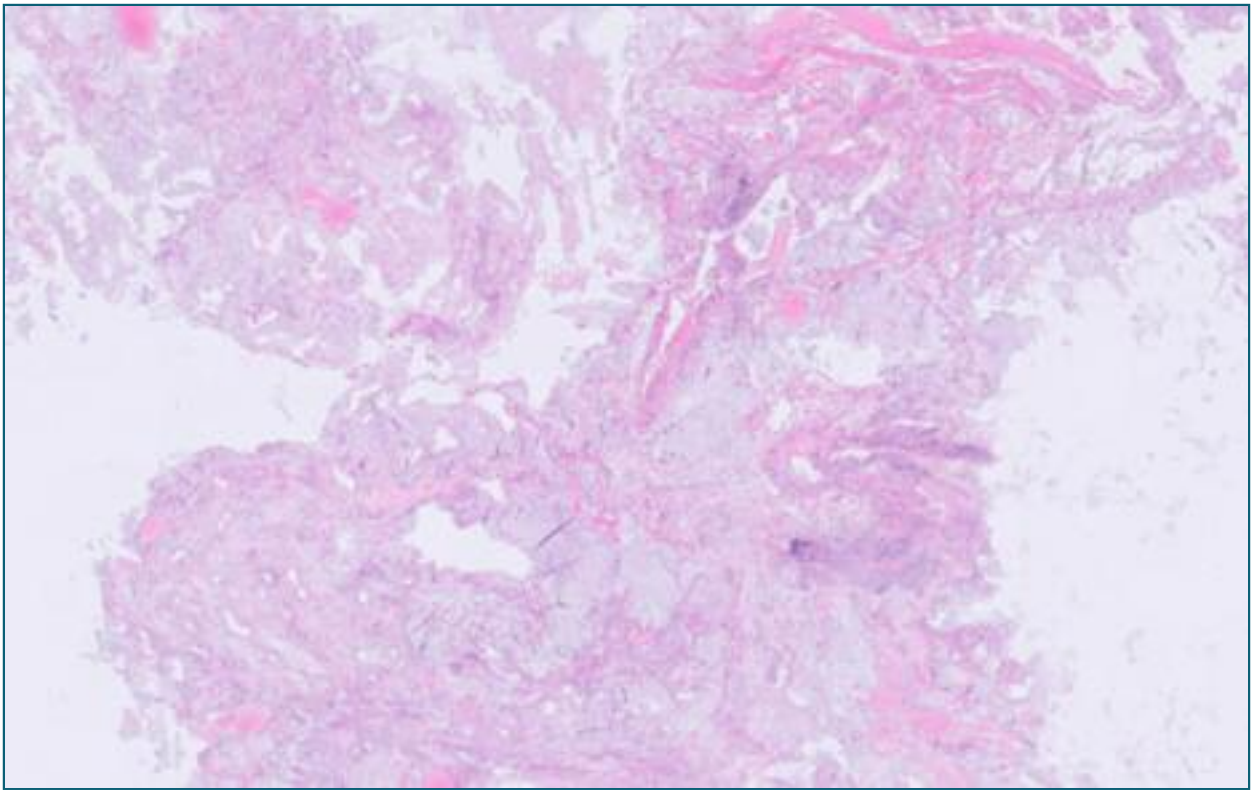

FIGURE 3. Mucous material (hematoxylin and eosin stain, 4x).

which, if the appendix ruptures, can lead to pseudomyxoma peritonei (PMP). A rarer and more aggressive variant, high-grade appendiceal mucinous neoplasm (HAMN), demonstrates high-grade dysplasia.

The clinical manifestations of LAMN are typically nonspecific and may include abdominal pain,

particularly in the right lower quadrant, a palpable mass, gastrointestinal symptoms such as nausea, vomiting, and bleeding, as well as signs of intestinal intussusception[5]. However, most cases are discovered incidentally during diagnostic or therapeutic procedures. In pregnant women, uterine contractions can obscure clinical symptoms, further complicating diagnosis.

To date, only five cases of LAMN detected during cesarean section have been reported in the literature[1-4,11]. Bowles *et al.* documented a case in which a 10.2 × 3.6 × 5.2 centimetres cystic structure in the right iliac fossa was incidentally diagnosed via first-trimester ultrasound[11]. Other reported cases involved asymptomatic women diagnosed intraoperatively[1-4]. Additionally, Yohannes *et al.* and Baron *et al.* described two cases in which LAMN was identified during cesarean section due to abnormal labor progression[2-3].

LAMN is characterized by the production of abundant mucin and an expansile growth confined to the *muscularis propria*. Appendectomy is typically sufficient when the lesion is localized. Both Inubashiri *et al.* and Ribeiro *et al.* reported cases of LAMN confined to the muscularis propria without mucosal fluid leakage, successfully treated with appendectomy, followed by five years of surveillance using CT, ultrasound, and tumor marker assessments[1,4].

However, LAMN growth can thin the appendiceal wall, increasing the risk of rupture and subsequent mucin spillage, leading to PMP. The mucin can induce fibrosis and obstruction, resulting in complications such as intussusception, volvulus, small bowel obstruction, and ureteral obstruction[2,5]. PMP is associated with a high recurrence rate (33 to 78 percent) and significant morbidity[7-10]. Despite previous concerns, right hemicolectomy does not appear to confer additional benefit over appendectomy alone[7]. Patients with PMP should be referred to specialized centers for peritoneal surface malignancy management, where CRS and HIPEC may be considered. Fertility preservation should also be discussed in affected individuals.

Histopathological examination in our case revealed LAMN with pT4a Nx M1b R0 staging, indicating cellular mucin involvement of the appendiceal serosa (pT4a), mucin deposits confined to the peritoneum (M1b), and clear surgical margins (R0)[6]. Postoperative imaging detected no locoregional or distant disease, and CRS/HIPEC was proposed due to peritoneal involvement.

Only one case of LAMN with PMP has been reported in the literature. Baron *et al.* documented a case where abundant mucin was observed adherent to pelvic organs during cesarean section, originating from the appendix tip. This patient underwent oocyte retrieval for cryopreservation before CRS/HIPEC and successfully conceived a second child using a frozen embryo three years post-treatment[3]. Conversely, Bowles *et al.* reported a case in which histopathological examination revealed extracellular mucin with inflammatory cells but no evidence of malignancy, leading to a decision for right hemicolectomy, which subsequently confirmed the absence of malignant or free mucin[11].

The overall three-year and five-year survival rates for LAMN with extra-appendiceal disease range from 91 to 100 percent and 79 to 86 percent, respectively. Patients require long-term follow-up, including routine imaging (abdominopelvic CT, magnetic resonance imaging, and/or ultrasound) and tumor marker evaluations (carcinoembryonic antigen [CEA] and carbohydrate antigens CA 125 and CA 19.9).

In conclusion, LAMN is a rare clinical entity, often detected incidentally. Even when associated with symptoms or identified as a cystic structure on imaging, suspicion remains low due to the higher prevalence of appendicitis and adnexal pathology. Early and accurate diagnosis is crucial for appropriate management. Thorough examination of abdominal and pelvic viscera during cesarean section should be a standard practice when abnormal findings are encountered, as timely identification of tumors such as LAMN can significantly impact patient outcomes.

REFERENCES

1. Inubashiri E, Watanabe Y, Akutagawa N, Kuroki K, Sugawara M, Deguchi K, et al. An incidental funding of low-grade appendiceal mucinous neoplasm during cesarean section: A case report. JGH Open. 2020;4(2):306-308. doi:10.1002/jgh3.12232.

2. Yohannes N, Watkins JC, Weeks AG, Osmundson SS, Shi C, Kovach AE. Low-grade Appendiceal Mucinous Neoplasm and Endometriosis: Incidental Coincident Pathologies at Cesarean Section. Int J Gynecol Pathol. 2020; 39(5):498-502. doi:10.1097/PGP.0000000000000630.

3. Baron E, Gushchin V, King MC, Nikiforchin A, Sardi A. Management of Low-Grade Appendiceal Mucinous Neoplasm with

Extensive Peritoneal Spread Diagnosed during Pregnancy: Two Case Reports and Literature Review. Case Rep Oncol Med. 2020;2020: 1-9. doi: 10.1155/2020/8853704.

4. Ribeiro NN, Côrtes LS, Araujo FM, Cerutti MLC, Caixeta DV, Lima MSCM, et al. Mucocele: Achado Cirúrgico Raro em Cesárea. Contemporary Journal. 2024; 4(1):2110-2120. doi: 10.56083/RCV4N1-116.

5. Panarelli NC, Yantiss RK. Mucinous neoplasms of the appendix and peritoneum. Arch Pathol Lab Med. 2011;135(10): 1261-1268. doi: 10.5858/arpa.2011-0034-RA.

6. Amin MB, Edge SB, Greene FL, Byrd DR, Brookland RK, Washington MK, et al. American Joint Committee on Cancer (AJCC) Cancer Staging Manual (8th Edition). New York, NY: Springer; 2017. ISBN 978-3-319-40617-6.

7. Yantiss RK, Shia J, Klimstra DS, Hahn HP, Odze RD, Misdraji J. Prognostic significance of localized extra-appendiceal mucin deposition in appendiceal mucinous neoplasms. Am J Surg Pathol. 2009; 33(2):248-255. doi: 10.1097/PAS.0b013e31817ec31e.

8. Pai RK, Beck AH, Norton JA, Longacre TA. Appendiceal mucinous neoplasm: clinicopathological study of 116 cases with analysis of factors predicting recurrence. Am J Surg Pathol. 2009; 33(10):1425-1439. doi: 10.1097/PAS.0b013e3181af6067.

9. Honoré C, Caruso F, Dartigues P, Benhaim L, Chirica M, Goéré D, et al. Strategies for Preventing Pseudomyxoma Peritonei After Resection of a Mucinous Neoplasm of the Appendix. Anticancer Res. 2015; 35(9):4943-4947. PMID: 26254392.

10. Roxburgh CS, Fening YM, Cercek A, Shia J, Rassam RM, Paty PB, et al. Outcomes of Low-Grade Appendiceal Mucinous Neoplasms with Remote Acellular Mucinous Peritoneal Deposits. Ann Surg Oncol. 2019; 26(1):118-124. doi: 10.1245/s10434-018-7003-7.

11. Bowles M, Ng JY, Nabi, H. Delivery of an Incidental Appendiceal Mucinous Neoplasm. Cureus. 2022; 14(6):e26214. doi: 10.7759/cureus.26214.

PATIENT'S INFORMED CONSENT

The patient has given informed consent for publication.

CONFLICTS OF INTEREST

The authors declare to have no conflict of interest.

AUTHORS' CONTRIBUTIONS

Joana Torres Silva: Conception, design, writing, reviewing and final editing of the article. Mariana Leal, Carla Duarte, Ana Maçães: Critical review of article content and final approval of the version to be published. Filomena Barreto: Provision and description of the images from the histopathological study.

CORRESPONDENCE TO:

Joana Torres Silva
E-mail: joanam.torres.silva@gmail.com
https://orcid.org/0009-0001-7053-5233

RECEIVED: 15/10/2024
ACCEPTED: 01/03/2025