



Case Report

Rare presentation of low-grade appendiceal mucinous neoplasms (LAMN) as an appendicular lump: A case report

Kinza Fatima^a, Ghayasuddin^a, Arsala Mushtaq^b, Mohammad Hasan^{c,*}

^a KVSS Hospital, Karachi, Pakistan

^b Agha Khan Hospital, Karachi, Pakistan

^c Jinnah Postgraduate Medical Center (JPMC), Karachi, Pakistan



ARTICLE INFO

Keywords:

Appendicular lump
Low-grade appendiceal mucinous neoplasms
Mucocele
Incidental diagnosis

ABSTRACT

Introduction: The prevalence of appendiceal mucinous neoplasms (AMN) is about 0.2%–0.3% in the specimens of the appendix. The LAMN may appear unremarkable or can present as mucin-filled, crystalline dilated tissues. The diagnosis of early-stage AMN is mostly incidental. It is of vital importance to know the features of LAMN for a timely diagnosis

Case presentation: A 46-year-old male came with the complaint of right iliac fossa swelling along with severe intensity pain and a single episode of vomiting. A 4 × 4cm tender, soft, cystic, mobile swelling was found upon the examination. Contrast-enhanced Computerized tomography (CT) scan of the abdomen revealed the appendix diameter of 10mm well-circumscribed cystic measuring 2.1 × 2 cm. Therefore, an open resection surgery was performed. The histopathological report revealed an intraepithelial lesion composed of flat mucinous epithelial cells having eosinophilic cytoplasm and low-grade nuclear atypia.

Case discussion: The AMNs are very rare neoplasms. The imaging modalities that can be diagnostic methods are abdominal and transvaginal ultrasonography (US), and abdominal computed tomography (CT). The low-grade features of the appendiceal mucinous neoplasms have the possibility of recurrence. The CT-scan findings of appendiceal lump > 1.3cm along with cystic dilation and the calcification of the wall are the suggestive features of AMN. There is a high chance of dissemination and port site seeding in case of pneumoperitoneum.

Conclusion: This rare case suggests the significance of adding AMNs as a differential diagnosis in patients with abdominal pain and choosing the right approach to treat such patients to avoid complications.

1. Introduction

The malignant appendicular tumors include neuroendocrine tumors (typical carcinoid), mucinous epithelial neoplasms, lymphomas, adenocarcinomas, goblet/ex-goblet cell or composite carcinoid, and lymphoid or mesenchymal sarcomas. Among these, 65% of the tumors histologically are of neuroendocrine origin. The prevalence of appendiceal mucinous neoplasms (AMN) is about 0.2%–0.3% in the specimens of the appendix [1]. Among all the tumors, they are one of the rarest tumors reported to be about 1% of all the tumors related to the gastrointestinal system [2]. Their treatment is quite controversial regarding the extent of surgery and chemotherapy which includes hyperthermic intraperitoneal chemotherapy (HIPEC) and early postoperative intraperitoneal chemotherapy (EPIC).

On examination, the low-grade AMN (LAMP) may appear unremarkable or can present as mucin-filled, crystalline dilated tissues. The wall of the appendix can be seen as thin, hyalinized, fibrotic, or calcified with a granular or smooth appearance. Low-grade AMN can be further categorized into flat or villous with atrophied lymphoid tissues [3]. According to the early literature, AMN was considered a benign disease with multiple terminologies such as cystadenoma, cystadenocarcinoma, and appendiceal mucocele. However, Carr et al. described it as a neoplasm having uncertain malignant potential with features of low-grade neoplasms with acellular mucin within or beyond the appendiceal wall [4].

The clinical course of LAMN is mainly determined by the stage at which it has been diagnosed as well as the histopathological features reflecting their cellular differentiation. For instance, the advanced stage

* Corresponding author. Postgraduate Medical Center (JPMC), Abdullah Haroon Rd, Artillery Maidan, Karachi, Karachi City, Sindh, Pakistan.

E-mail addresses: kinza_zaidi110@hotmail.com (K. Fatima), mghayas1@yahoo.com (Ghayasuddin), arsala.mushtaq@aku.edu (A. Mushtaq), M_Hasan_96@yahoo.com (M. Hasan).

<https://doi.org/10.1016/j.amsu.2022.104848>

Received 10 August 2022; Received in revised form 21 September 2022; Accepted 30 October 2022

Available online 7 November 2022

2049-0801/© 2022 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

is characterized by the presence of mucinous ascites that lead to pseudomyxoma peritonei (PMP) and has the worst prognosis [1,2]. The common associations of AMN include herniation, dissections, diverticula, and rupture [3]. The diagnosis of early-stage AMN is mostly incidental therefore the histologic differentiation is of vital significance as the well-differentiated mucin-producing appendiceal tumors have lower mortality rates and better prognosis as compared to the poorly differentiated neoplasms. The treatment depends on the histology and the staging [4].

We present this unique case where the appendicular lump changes into a mucocele and presents with findings of residual inflammation in the right iliac fossa with mucocele formation and surrounding peri-appendiceal fluid.

Our Case Report is compliant with SCARE 2020 Guidelines [5]. A complete SCARE 2020 checklist has been provided as a supplementary file.

2. Case presentation

The 46-year-old male with no known comorbid presented in the emergency department, KulsumBai Valika Social Security SITE (KVSS) Hospital with a complaint of right iliac fossa swelling & severe intensity pain in the same region along with a single episode of vomiting. He reported no family history of cancer and no personal history of drug use. Laboratory baseline results which include CBC, UCE, LFT, and PT/INR were unremarkable, and the patient denied any history of hypertension, diabetes, tuberculosis, or malignancy. On Examination, 4 × 4cm tender, soft, cystic, mobile swelling in the right iliac fossa was found.

Ultrasound (US) whole abdomen (WAB) was performed which confirmed the presence of a complex cystic area with minimal peripheral vascularity in the right iliac fossa due to an appendicular lump. The appendix is not separately visualized from the cystic area. Furthermore, Contrast-enhanced Computerized tomography (CT) scan of the abdomen revealed the appendix diameter of 10mm well-circumscribed cystic measuring 2.1 × 2 cm from the tip of the appendix suggestive of residual inflammation in the right iliac fossa with mucocele formation as shown in the Fig. 1. Hence, he was referred to our department for surgical intervention.

On enquiring, the patient gave a similar history of moderate-intensity pain in the right iliac fossa almost 01 years ago, but no swelling was felt by the patient. Previous Ct-scan revealed an appendicular lump as seen in Fig. 2.

Open resection surgery was performed that revealed the appendix of 4 × 1.3 cm and the cystic tissue of 5 × 2.7 × 2.3 cm as seen in Figs. 3 and 4. The cyst was filled with gelatinous material. The average wall thickness of the cyst was 0.1 cm. The lumen of the appendix is partly filled with gelatinous material. The histopathological report revealed intraepithelial lesions composed of flat mucinous epithelial cells having eosinophilic cytoplasm and low-grade nuclear atypia. The lesion invaginates mesoappendix forming cysts filled with mucinous pools consistent with low-grade appendicular mucinous neoplasm (LAMN).

Postoperatively, the patient was stable with no surgical complications. There were no signs or symptoms of recurrence within the follow-up period of 6 months. He was advised to follow up for at least 5 years.

3. Discussion

The AMNs are often confused with acute appendicitis, adnexal mass, or a retroperitoneal tumor when they are discovered during endoscopy, radiology, or in the operation theatre [6]. These misdiagnoses are further caused due to variations in the diagnostic imaging modalities [7]. The physicians suggest using ultrasound (US) as a diagnostic modality to distinguish the AMNs from the more commonly presented disease acute appendicitis defined as an appendiceal outer diameter of equal to or more than 15mm and the visualization of mucinous effusion as defined by the US criteria. In cases of preoperative diagnosis,



Fig. 1. CT-scan of the patient showing mucocele.

contrast-enhanced CT is a better radiological tool. The findings of appendiceal lump >1.3cm along with cystic dilation and the calcification of the wall on the CT scan are suggestive of AMN [8,9].

The low-grade features of the appendiceal mucinous neoplasms have the possibility of recurrence due to which they have multiple histologic classification schemas so that it could be predicted whether the patient will have the recurrence and will need a follow-up visit or not. Consequently, many researchers have referred to it as “uncertain malignant potential” when the LAMP appears with features that are not clear whether it is malignant or benign [10–13]. The LAMN of <2 cm is rarely malignant and is categorized as a benign retention mucocele. However, the AMNs larger than 6cm have a greater risk of malignancy, greater appendiceal perforation, and more chances of developing the most feared complication of PMP [14]. The AMN manifests histologically with features of atypical glandular cells and epithelial cells with the presence of “pushing invasion” of the malignant cells invading within the appendiceal wall with the possibility of the formation of diverticula [3]. At times, mucinous, goblet, and colonic cells are also frequently found in the AMN [15]. Reports suggest a risk of 35% concurrent GI malignancy in patients suffering from AMN [15].

The decision of a better surgical approach between open resection and laparoscopy is quite controversial. The literature suggests a reduced incidence of malignant mucinous cystadenocarcinoma however while deciding the procedure between open resection and laparoscopy, the surgeon must be careful to minimize the events of rupture and mucinous



Fig. 2. CT-scan of this patient showing the appendiceal lump.

seeding [16]. There is a high chance of dissemination and port site seeding in case of pneumoperitoneum and the removal of specimen via abdominal wall. The study by Fujini et al. suggests the laparoscopic approach to be used considering its benefits of decreased risk of seeding and quicker recovery [17].

Post-appendectomy, the patients are advised to come for a follow-up for at least 5–10 years in which, a thorough physical examination, annual CT-scan imaging, and monitoring of the CA19-9 and CEA (raised levels to suggest recurrence) [18]. There is a need for further studies related to the treatment and prognosis of AMN to propose the definitive treatment of the disease. A close follow-up was recommended for our patient as the 5-year survival rate for localized AMN is 95%.

4. Conclusion

This case report concludes that AMN is one of the rarest diseases and its diagnosis is incidental. As we conclude that there are chances of an appendicular lump leading to a mucocele, there is a need for further studies to diagnose the appendicular lump in its early stages and surgically excise it after 6 weeks to prevent its progression into AMN. Diagnostic modalities vary from radiological imaging to monitoring tumor biomarkers. This case of AMN suggests the importance of considering the possibility of appendiceal neoplasms in patients with abdominal pain and choosing the right approach to treat such patients



Fig. 3. Appendiceal Mucinous neoplasm.

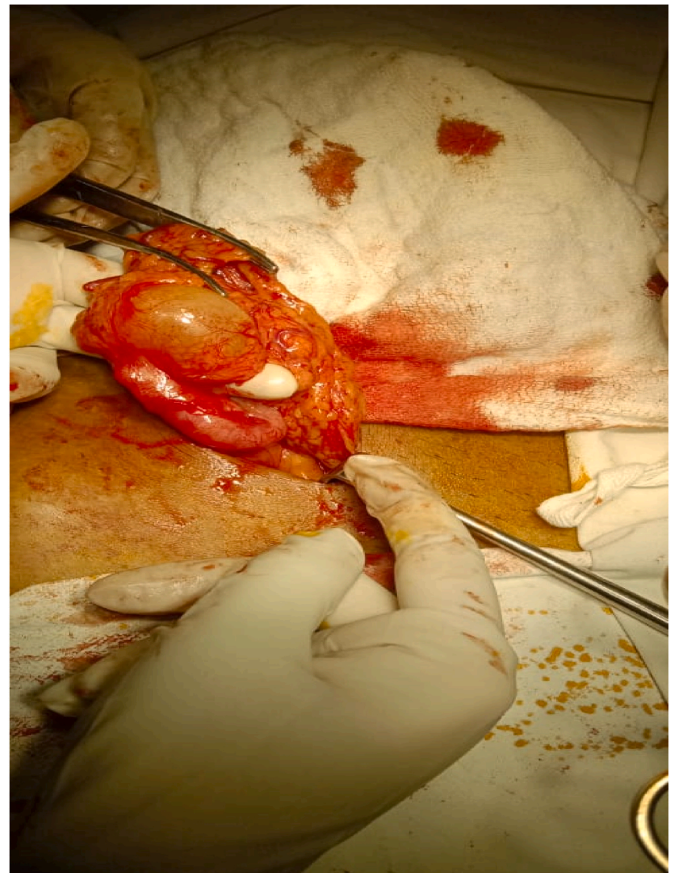


Fig. 4. Resection of LAMN

and prevent the recurrence, seeding, and development of PMP.

Ethical approval

This is a case report and it didn't require ethical approval from the

ethics committee according to our institution's policy.

Sources of funding

This study did not receive any funding from governmental or private organizations.

Author contributions

1. The study concept or design was determined by Kinza Fatima, Ghayasuddin
2. Collection of data and interpretation is done by Arsalan Mushtaq, Kinza Fatima
3. Writing of the manuscript is done by Kinza Fatima, Mohammad Hasan,
4. Manuscript editing is done by Mohammad Hasan, Arsalan Mushtaq

Consent

Written informed consent was taken from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Research registration

NA.

Guarantor

Kinza Fatima.

Declaration of competing interest

The authors report no conflicts of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2022.104848>.

References

- [1] R.M. Smeenk, M.L. Van Velthuysen, V.J. Verwaal, F.A. Zoetmulder, Appendiceal neoplasms and pseudomyxoma peritonei: a population based study, *Eur. J. Surg. Oncol.* 34 (2) (2008 Feb 1) 196–201, <https://doi.org/10.1016/j.ejso.2007.04.002>.
- [2] V. Ramaswamy, Pathology of mucinous appendiceal tumors and pseudomyxoma peritonei, *Indian J. Surg Oncol.* 7 (2) (2016 Jun) 258–267, <https://doi.org/10.1007/s13193-016-0516-2>.
- [3] J. Misdrjaji, R.H. Young, Primary epithelial neoplasms and other epithelial lesions of the appendix (excluding carcinoid tumors), *InSeminars. Diagn. Pathol.* 21 (No. 2) (2004 May 1) 120–133, <https://doi.org/10.1053/j.semmp.2004.11.005>. WB Saunders.
- [4] W.L. Shaib, R. Assi, A. Shamseddine, O.B. Alese, C. Staley III, B. Memis, V. Adsay, T. Bekaii-Saab, B.F. El-Rayes, Appendiceal mucinous neoplasms: diagnosis and management, *Oncol.* 22 (9) (2017 Sep) 1107–1116, <https://doi.org/10.1634/theoncologist.2017-0081>.
- [5] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, A.J. Beamish, A. Noureldin, A. Rao, B. Vasudevan, B. Challacombe, The SCARE 2020 guideline: updating consensus surgical Case Report (SCARE) guidelines, *Int. J. Surg.* 84 (2020 Dec 1) 226–230, <https://doi.org/10.1016/j.ijsu.2020.10.034>.
- [6] K.F. von Rokitsansky, *A Manual of Pathological Anatomy*, Blanchard & Lea, 1855.
- [7] M.E. McFarlane, J.M. Plummer, K. Bonadie, Mucinous cystadenoma of the appendix presenting with an elevated carcinoembryonic antigen (CEA): report of two cases and review of the literature, *Int. J. Surg. Case Rep.* 4 (10) (2013 Jan 1) 886–888, <https://doi.org/10.1016/j.ijscr.2013.07.007>.
- [8] F.U. Malya, M. Hasbahceci, A. Serter, G. Cipe, O. Karatepe, E. Kocakoc, M. Muslumanoglu, Appendiceal mucocele: clinical and imaging features of 14 cases, *Chirurgia (Buchar.)* 109 (6) (2014 Nov 1) 788–793.
- [9] A. Rouchaud, L. Glas, M. Gayet, M.J.D.I.I. Bellin, Appendiceal mucinous cystadenoma 95 (1) (2014) 113–116.
- [10] J.D. Seidman, A.M. Elsayed, L.H. Sobin, F.A. Tavassoli, Association of mucinous tumors of the ovary and appendix. A clinicopathologic study of 25 cases, *Am. J. Surg. Pathol.* 17 (1) (1993 Jan 1) 22–34, <https://doi.org/10.1097/00000478-199301000-00003>.
- [11] N.J. Carr, W.F. McCarthy, L.H. Sobin, Epithelial noncarcinoid tumors and tumor-like lesions of the appendix. A clinicopathologic study of 184 patients with a multivariate analysis of prognostic factors, *Cancer* 75 (3) (1995 Feb 1) 757–768, [https://doi.org/10.1002/1097-0142\(19950201\)75:3<757::AID-CNCR2820750303>3.0.CO;2-F](https://doi.org/10.1002/1097-0142(19950201)75:3<757::AID-CNCR2820750303>3.0.CO;2-F).
- [12] N.J. Carr, L.H. Sobin, Unusual tumors of the appendix and pseudomyxoma peritonei, *InSeminars. Diagn. Pathol.* 13 (4) (1996 Nov 1) 314–325.
- [13] R.K. Pai, T.A. Longacre, Appendiceal mucinous tumors and pseudomyxoma peritonei: histologic features, diagnostic problems, and proposed classification, *Adv. Anat. Pathol.* 12 (6) (2005 Nov 1) 291–311, <https://doi.org/10.1097/01.pap.0000194625.05137.51>.
- [14] V. Padmanaban, W.F. Morano, E. Gleeson, A. Aggarwal, B.L. Mapow, D.E. Stein, W. B. Bowne, Incidentally discovered low-grade appendiceal mucinous neoplasm: a precursor to pseudomyxoma peritonei, *Clin. Case. Rep.* 4 (12) (2016 Dec) 1112, <https://doi.org/10.1002/ccr3.694>.
- [15] L. Narasingamoorthy, P. Rajagopalan, D. Subramaniam, Primary mucinous adenocarcinoma of appendix: a rare case report, *J. Evol. Med. Dent. Sci.* 5 (65) (2016 Aug 15) 4678–4680, <https://doi.org/10.14260/jemds/2016/1066>.
- [16] S. Hassan, A. Dhebri, L. Lin, M. Haque, Appendiceal mucocele: a missed diagnosis, 2013 Jan 11, *Case Rep.* (2013), <https://doi.org/10.1136/bcr-2012-007983>. bcr2012007983.
- [17] S. González Moreno, B.M. Shmookler, P.H. Sugarbaker, Appendiceal mucocele, *Surg. Endosc.* 12 (9) (1998 Sep) 1177–1179, <https://doi.org/10.1007/s004649900811>.
- [18] B. Papaziogas, I. Koutelidakis, P. Tsiaousis, O.C. Goula, S. Lakis, S. Atmatzidis, J. Makris, G. Paraskevas, K. Atmatzidis, Appendiceal mucocele. a retrospective analysis of 19 cases, *J. Gastrointest. Cancer* 38 (2–4) (2007) 141–147, <https://doi.org/10.1007/s12029-008-9030-2.PMID:19089669>.