ejpmr, 2018,5(3), 104-107

SJIF Impact Factor 4.897

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

ADENOCARCINOMA OF APPENDIX MIMICKING ACUTE APPENDICITIS: A CASE REPORT AND LITERATURE REVIEW

[1]*Dr. Abdalla Saad Abdalla AL-Zawi, [1]Dr. Anita Lazarevska, [1]Dr. Mohammed Omer, [2]Dr. Malgorzata Lange-Ratajczak, [3]Dr. Emraga Abohamod and [1]Dr. Elizabeth Tan

MBBCh, SD, PhD, FRCS*[1], MBBCh, SD[1], MBBCh[1], MbChb, BA[1], MBBCh, PhD[2] and Associate Professor[3]

[1]Basildon University Hospital, Basildon, Essex, United Kingdom.

[2]Department of General, Endocrine & Gastroenterology Oncological Surgery, University of Medical Sciences, Poznan, Poland.

[3]Department of Anatomy, Sebha University Medical School, Libya.

***Corresponding Author: Dr Abdalla Saad Abdalla AL-Zawi**

MBBCh, SD, PhD, FRCS Basildon University Hospital, Basildon, Essex, United Kingdom.

Article Received on 06/01/2018 **Article Revised on 27/01/2018** **Article Accepted on 16/02/2018**

ABSTRACT

Adenocarcinoma of the appendix is a rare neoplasm, often diagnosed in patients of the older age group, who present with a clinical picture of acute appendicitis. This paper presents a case of adenocarcinoma of the appendix in a case initially treated as acute appendicitis.

KEYWORDS: Adenocarcinoma of appendix, Acute appendicitis, Appendicectomy, Right hemicolectomy, Krukenberg tumour.

INTRODUCTION

Primary cancer of the appendix is diagnosed in 0.9 – 1.4% of appendicectomy specimens.[1] The majority of primary malignant appendiceal tumours are carcinoid (up to 90%), followed by mucinous adenocarcinoma.[2,3] In general, primary appendiceal cancer accounts for only 0.4% of all gastrointestinal cancers.[4,5] The most common clinical picture involves features of acute appendicitis, however an abdominal mass or anaemia may also feature. Appendiceal cancer is histologically classified into carcinoids, goblet cell adenocarcinoma (GCA), colonic and mucinous adenocarcinoma.[4,6,7] Right hemicolectomy may be necessary to ensure proper staging and management. This paper presents a case of adenocarcinoma of the appendix in a female patient aged 83 years.

CASE REPORT

An 83-year old female patient presented to the Accident & Emergency Department with a 2-day history of right iliac fossa pain and reduced appetite. She was known to have congestive cardiac failure, atrial fibrillation (on warfarin) and was classed as ASA 3. Blood results showed white cell count $8x10^9$/L and C-Reactive Protein 36mg/L. CT abdomen revealed a fluid-filled dilated (11mm) appendix, surrounded by inflammatory changes [Fig.1&2].

The patient underwent laparoscopic appendicectomy. Intraoperatively, the appendix was found to be inflamed. However, approximately 1 inch distal to the base, there was a tense, pale, grey nodule, possibly cystic in nature. After resection, the lump was measured to be 26mm distal to the base and 16 mm in length [Fig.3]. Histology was consistent with a mucinous, moderately-differentiated adenocarcinoma. The tumour invaded the muscularis propria into the subserosa, but not into the serosal surface. There was no evidence of lympho-vascular invasion [Fig.4]. The tip and base of the appendix were free of tumour involvement, however showed severe transmural acute inflammation with associated serositis. Immunostaining for synaptophysin was negative, but chromogranin staining showed a few cells were positive, with Ki67 positivity in more than 90% of cells. The tumour was graded as T3NxMx. The postoperative histology was discussed with the patient, and further treatment with right hemicolectomy and surveillance was offered. The patient expressed a firm desire to have no further surgery or interventions.

Several months later, a follow-up CT scan showed a malignant-looking lesion in the descending colon [Fig.5&6], with signs of peritoneal deposits [Fig.7]. There was also a complex adnexal mass, suggestive of the presence of a Krukenberg tumour or the possibility of a synchronous ovarian malignancy. The patient was referred to the oncological team for palliative care.

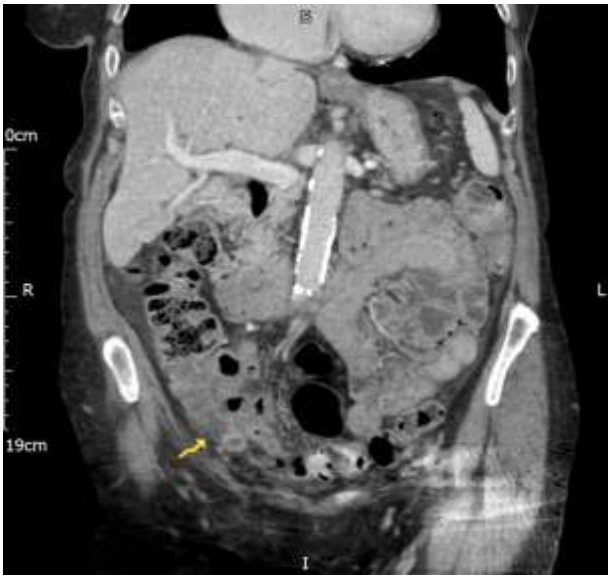

Fig 1: CT abdomen - coronal view: The appendix is dilated to 11 mm, fluid-filled, and surrounded by minor inflammatory changes.

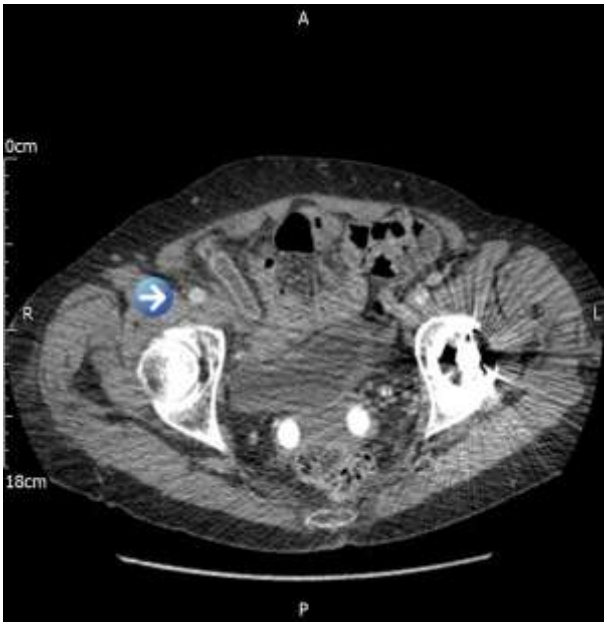

Fig 2. CT abdomen - axial view: The appendix is dilated to 11 mm, fluid-filled, and surrounded by minor inflammatory changes.

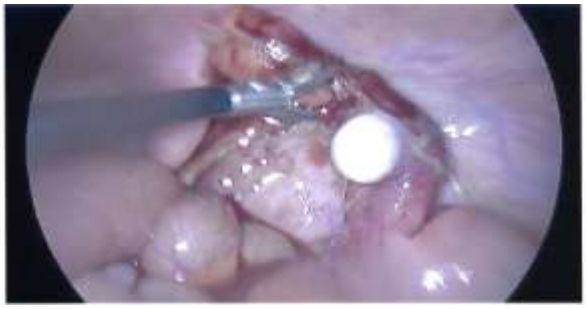

Fig. 3: A cystic mass seen distal to the base of the appendix during laparoscopic appendicectomy, initially diagnosed as a mucocele.

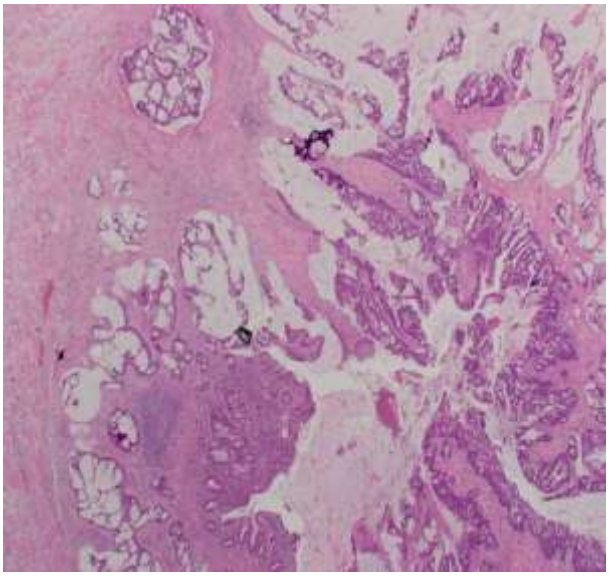

Fig.4: Appendix showing extensive infiltration of the wall by a mucinous adenocarcinoma.

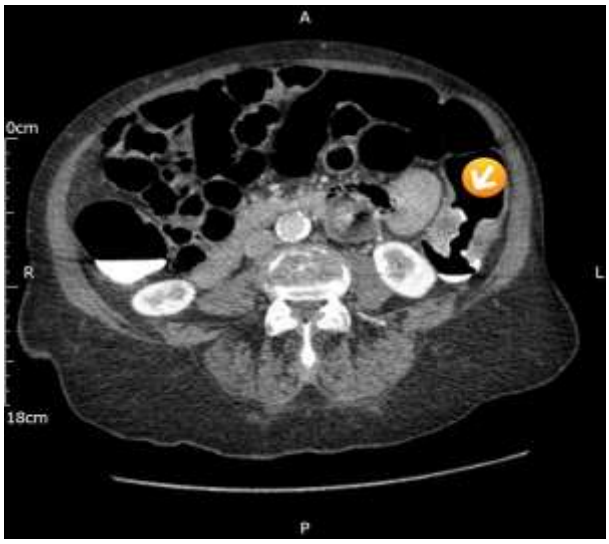

Fig.5: CT Abdomen - axial view: malignant looking stricture within the descending colon.

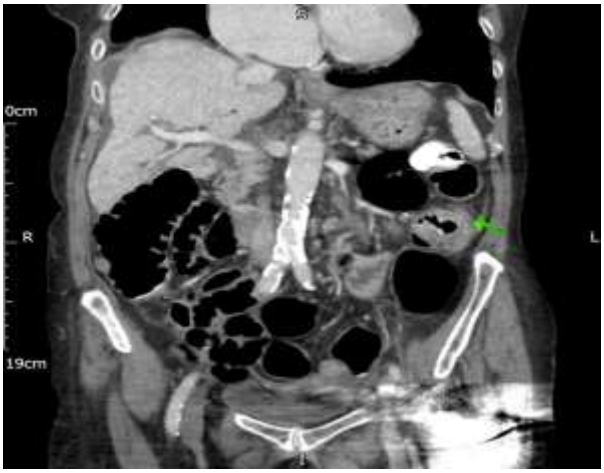

Fig.6: CT Abdomen - Coronal view: Malignant looking stricture within the descending colon.

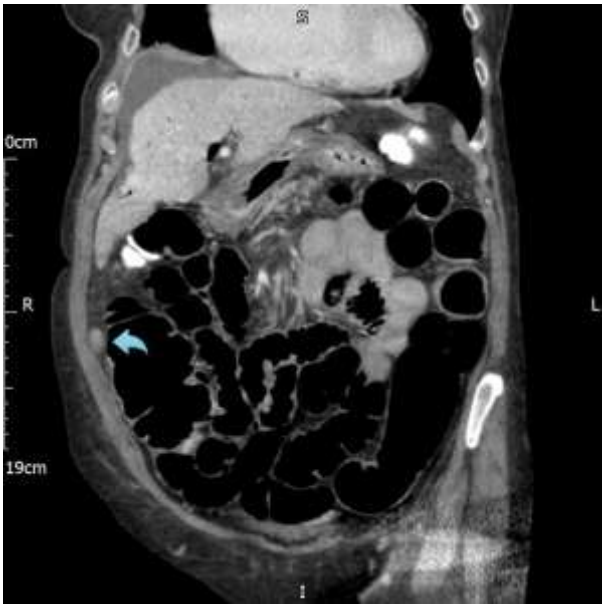

Fig.7: CT abdomen - Coronal view: peritoneal malignant deposit.

DISCUSSION

Adenocarcinoma of the vermiform appendix is a rare neoplasm, usually diagnosed incidentally and most often affecting adults.[8] It was first described by Berger in 1882[4]; the patients often present with the clinical picture of acute appendicitis, particularly with right iliac fossa abdominal pain.[9] Other rare presentations also reported include an abdominal mass[10], right thigh mass[5,11], caecal intussusception[12], peritonitis, anaemia[13], hydronephrosis[14], or Krukenberg tumour of the ovary.[15]

In general, the incidence of appendiceal neoplasm varies from 0.01 to 0.2 per 100,000 persons per year.[7] The majority of primary appendiceal cancers occur in the age group of 55–65 years, with the exception of the malignant carcinoid, which has been reported in patients with a mean age of 38 years.[16] Patients of an older age group and presenting with symptoms of acute appendicitis should be investigated for the possibility of an underlying neoplasm[8]; a CT scan is suggested in a such scenarios. The tumour may be diagnosed either as an isolated diagnosis or associated with another condition, such as ulcerative colitis[17], ovarian malignancy[18] or De Garengeot hernia (femoral hernia containing the appendix).[19] It is also possible for adenocarcinoma of the appendix to arise in a post appendicectomy stump.[20] The reported complications from appendiceal adenocarcinoma include torsion, disease dissemination, hydronephrosis[14], intussusception, and bowel perforation.[5]

Carcinomas arising in the appendix are classified macroscopically into four forms: ulcerative, vegetative, infiltrative or mixed. They are histologically classified into four histopathological distinct subtypes: carcinoid, goblet cell adenocarcinoma (GCA), colonic and mucinous adenocarcinoma.[4,6,7]

The most common primary appendiceal malignant tumour is carcinoid (up to 90%)[4,5], followed by mucinous adenocarcinoma.[2,3] This mucinous variety most often presents following perforation of the primary tumour, with a spread of mucin and tumour cells throughout the peritoneal cavity. This results in a clinical syndrome of abdominal distention, secondary to mucinous ascites; it is known as *pseudomyxoma peritonei*. The condition is usually diagnosed at operation for appendicitis, peritonitis, possible ovarian malignancy, or on diagnostic imaging.[21] Several other conditions of the appendix must also be differentiated from primary mucinous appendiceal adenocarcinomas, including acute appendicitis, mucocele[22], carcinoid, caecal carcinoma, lymphoma[5] or rarer tumours such as ganglioneuroma.[23] The colonic type is found in less than 0.1% of cases, where the cystic sub-variety is characterised by its tendency for perforation and dissemination.

The surgical management of appendiceal carcinoma is controversial; is appendicectomy enough or should it be extended to right hemeicolectomy? Appendicectomy alone is justified when the tumour is limited to the mucosa and does not extend beyond the muscularis mucosae and has no vascular infiltration. The only exception for this is in goblet cell adenocarcinoma, as 20 - 40% of these cases show regional lymph node metastasis. Therefore, for appropriate management, right hemicolectomy and staging investigations are highly recommended for adequate management regardless of primary tumour size.[21,24] For all cases of goblet cell adenocarcinoma, locally advanced adenocarcinoma or carcinoids, postoperative staging is suggested, followed by right hemicolectomy and systemic chemotherapy as required.[21] Positive results have also been reported by Sugarbaker, following development of a definitive cyto-reductive surgery using hyperthermic intraoperative intraperitoneal chemotherapy.[25]

After surgery and adjuvant treatment, patients should be followed up with clinical assessment and endoscopy imaging in the same way as for colonic cancer.

CONCLUSION

Patients of an older age group presenting with symptoms of acute appendicitis should be investigated for the possibility of an underlying malignancy; a CT scan is suggested in such scenarios. Detection of a vermiform appendiceal adenocarcinoma is essential, as it ensures the appropriate management of a right hemicolectomy instead of appendicectomy alone.

REFERENCES

1. Connor SJ, Connor SJ, Hanna GB, Frizelle FA. Appendiceal tumors: retrospective clinicopathologic analysis of appendiceal tumor from 7970 appendectomies. Dis Colon Rectum, 1998; 41: 75–80.
2. McCusker ME, Cote TR, Clegg LX, Sobin LH. Primary malignant neoplasm of the appendix: a population-based study from the surveillance, epidemiology and end-results program, 1973-1998. Cancer, 2002; 94: 3307-3312.
3. Mcgory M, Maggard M, Kang HE et al. Malignancies of the appendix: Beyond case series reports. Dis Colon rectum, 2005; 48: 2264. https://doi.org/10.1007/s10350-005-0196-4.
4. León Arellano M, González-Domínguez Y, Molina-Ortiz F, et al. Primary adenocarcinoma of the appendix: experience at La Paz University Hospital of Madrid (1967–2014). Int J Surg Open, 2016; 4: 23–6.
5. Handler M, Anand N, Wei L, Snieckus P. Adenocarcinoma of the Appendix Presenting as a Palpable Right Thigh Mass. Journal of Radiology Case Reports, 2017; (11): 4.
6. KO YH, PARK SH, JUNG CK, WON HS, HONG SH, PARK JC, ROH SY, WOO IS, KANG JH, HONG YS, BYUN JH. Clinical characteristics and prognostic factors for primary appendiceal carcinoma. Asia-Pacific Journal of Clinical Oncology, 2010; 6: 19–27. doi: 10.1111/j.1743-7563.2010.01276.x.
7. Komm M, Kronawitter-Fesl M, Kremer M, Lutz L, Holinski-Feder E, Kopp R. Primary Mucinous Adenocarcinoma of the Vermiform Appendix with High Grade Microsatellite Instability. Journal of Cancer, 2011; 2: 302-306.
8. Pickhardt PJ, Levy AD, Rohrmann CA Jr, et al. Primary neoplasms of the appendix manifesting as acute appendicitis: CT findings with pathologic comparison. Radiology, 2002; 224: 775–81.
9. Wang YT, Li YR, Ke TY. Adenocarcinoma Ex Goblet Cell Carcinoid of Appendix: Two Case Reports. Case Reports in Pathology, 2017. Article ID 5930978.https://doi.org/10.1155/2017/5930978.
10. Kalpande S, Pandya J, Sharma T. Adenocarcinoma mimicking appendicular lump: a diagnostic dilemma—a case report. World Journal of Surgical Oncology, 2016; 14: 28. DOI 10.1186/s12957-016-1036-9.
11. Kim MK, Lee HY, Song IS, et al. A case of a giant mucocoele of the appendiceal stump presented with a palpable mass in the right thigh: pre-operative diagnosis based on characteristic multidetector CT findings. The British Journal of Radiology, 2010; 83(994): e220-e223. doi:10.1259/bjr/98679856.
12. Lee CT, Lien WC, Wang HP, Lin BR, Huang PH.Lin JT. Primary appendiceal adenocarcinoma with caeco-colic intussusception. Journal of Gastroenterology and Hepatology, 2006; 21: 1079–1081. doi:10.1111/j.1440-1746.2006.04105.
13. Yamada T, Murao Y, Nakayama T, Tabuse H, Miyamoto S, Imai S, et al. Primary adenocarcinoma of the appendix, colonic type associated with perforating peritonitis in an elderly patient. J Gastroenterol, 1997; 32: 658–62.
14. Lee J, Lim H, Lee S. Ureteral obstruction caused by mucocele of the appendix: MDCT findings. Australasian Radiology, 2007; 51: B59–B61. doi:10.1111/j.1440-1673.2007.01800.x
15. Thomas R, Bumhill D, Worsham F, Hoskins W. Krukenberg tumor of the ovary from un occult appendiceal primary: case report and review of the literature. Obstet Gynecol, 1985; 65: 955–85.
16. Rouff C, Hanna L, Zhi W, Shahzad G,Gotlieb V, Saif MW. Cancers of the Appendix: Review of the Literatures. Oncology, 2011. Article ID 728579, DOI:10.5402/2011/728579.
17. Stahn B, Hartmann C, Heinig, Scheit L. Ulcerative Colitis and Adenocarcinoma of the Appendix: Is there a Causal Link?. Dtsch med Wochenschr, 2017; 142(17): 1300-1303.DOI: 10.1055/s-0043-106879.
18. Kashani M, Levy M. Primary adenocarcinoma of the appendix with bilateral Krukenberg ovarian tumors. J Surg Oncol, 1983; 22(02): 101-105.
19. Ryan JW, O'Riordan I, Gorey T, Geoghegan T. de Garengeot hernia with a mucinous neoplasm of the appendix, two clinical rarities combine to yield a first for the literature. BMJ Case Rep, 2017. doi:10.1136/bcr-2017-220830.
20. Xiong L, Li TG, Zhao H. A Rare Case of Appendiceal Stump.Adenocarcinoma and Review of Literature. Kuwait Medical journal, 2014; 46(1): 57-59.
21. Kelly KJ. Management of appendix cancer. Clin Colon Rectal Surg, 2015; 28: 247–55. doi: 10.1055/s-0035-1564433.
22. Saad Abdullah A. Mucocele of the appendix. Kuwait Medical Journal, 1998; 30(4): 329-330.
23. Lockhart ME, Smith JK, Canon CL, Morgan DE, Heslin MJ. Appendiceal ganglioneuromas and pheochromocytoma in neurofibromatosis type I. American Journal of Roentgenology, 2000; 175(1): 132–134.
24. Pape U F, Perren A, Niederle B. et al. ENETS Consensus Guidelines for the management of patients with neuroendocrine neoplasms from the jejuno-ileum and the appendix including goblet cell carcinomas. Neuroendocrinology, 2012; 95(2): 135–156.
25. Sugarbaker PH. New standard of care for appendiceal epithelial neoplasms and pseudomyxoma peritonei syndrome? Lancet Oncol, 2006; 7: 69–76.