Mucinous cystadenoma of the appendix presenting with an elevated carcinoembryonic antigen (CEA): Report of two cases and review of the literature

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ABSTRACT

INTRODUCTION: Despite the fact the mucocoele of the appendix is a rare entity it has been the subject of much interest in the literature. The term mucocoele refers to cystic dilatation of the appendix filled with mucin resulting from proximal obstruction of the appendix lumen.

PRESENTATION OF CASE: We report two patients presenting with mucinous cystadenoma of the appendix with elevation of serum carcinoembryonic antigen (CEA), which has rarely been reported. Both patients had mild symptoms and a normal colonoscopy preoperatively. The diagnosis was not suspected in one patient prior to surgery. The elevated CEA prompted additional diagnostic radiologic investigations including ultrasonography, and computed tomography scans. The patients had uneventful appendicectomies with subsequent normalization of their CEA levels.

DISCUSSION: Mucinous cystadenoma of the appendix is a rare pathological entity characterized by a dilated mucous filled appendix. The presence of an elevated CEA associated with the presence of this entity is a rare presentation.

CONCLUSION: Because the diagnosis is rarely suspected prior to surgery patients with an enlarged appendix with associated elevated CEA levels should have careful investigations to exclude malignancy.© 2013 The Authors. Published by Elsevier Ltd on behalf of Surgical Associates Ltd. All rights reserved.

1. Introduction

Appendiceal mucocoele (AM) is a rare pathological entity characterized by cystic dilatation of the lumen of the appendix, which contains mucous. It may present with a variety of clinical syndromes or it can occur as an incidental finding. The prevalence of AM is 0.2–0.4% of appendectomies. This pathological entity of the appendix was first described by Rokitansky and divided into benign and malignant types. The term mucocoele is non-specific and refers to a cystic mass containing mucin. These masses have been more clearly defined based on histopathological changes to the underlying epithelium of the appendix as mucosal hyperplasia (25%), mucinous cystadenoma (63%), mucinous cystadenocarcinoma (12%) and retention cysts. Synchronous colon cancers have been recognized to occur with AM and this association is between 19.5% and 21.4%. Different treatment modalities based on the results of detailed investigations will be required to differentiate between the pathological variants.

The surgical management of this entity remains quite controversial with a significant difference in the fundamental approach, whether open or laparoscopic. Complete excision of the mucocoele without rupture is of paramount importance since although a benign disease, complications may manifest from rupture resulting in pseudomembrana peritonei and the risk of malignancy. Careful adherence to oncological principles is therefore important. The application of laparoscopy to appendiceal disorders other than appendicitis has seldom been reported. Justification for the use of this approach in the management of cases other than confirmed appendicitis requires careful judgement. Reported cases of patients with mucocoele of the appendix presenting with elevated carcinoembryonic antigen (CEA) has rarely been reported. Herein we report two cases, which were treated with both a laparoscopic and open approach respectively with good results.

2. Case report

A 72-year-old female who underwent a routine yearly medical examination by her private physician in Jamaica was found to have an elevated CEA level of 26 ng/ml. She was asymptomatic and in good health. Significantly in her past medical history she had been screened for both colon and breast cancer. A physical examination was normal. Colonoscopy done for bright red blood per rectum 6 months prior to presentation was normal apart from internal haemorrhoids.

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MDCT scans confirmed the presence of a cystic pelvic mass. Blood investigations were normal apart from an elevated CEA of 26 ng/ml (N 0.5 ng/ml). A standard 3-port laparoscopic appendicectomy was performed and a large unruptured cystic swelling of the appendix with some adhesions around the terminal ileum was found. The postoperative course was uneventful and she was discharged the following day. The histology confirmed a veriform appendix with part of the mucosa replaced by a multilocular, mucus-filled cyst lined by tall columnar mucinous epithelium. There were areas of lymphoid aggregates and areas of dystrophic calcification. The features were consistent with a mucinous cystadenoma of the appendix. The patient remained well following discharge with a return to normal in her CEA level and CEA repeated at 6 months was normal.

3. Case report 2

A 54-year-old man was seen with a 1-year history of recurrent right iliac fossa pain of moderate severity. There were no constitutional symptoms. Initial investigations were normal apart from an elevated CEA level of 128 ng/ml.

Abdominal examination was normal except for mild tenderness in the right iliac fossa and the hypogastrium. Computed tomography scans were performed and revealed a 15 cm × 8 cm cystic mass in the pelvis adjacent to the rectum. Colonoscopy performed and was normal. A provisional diagnosis of rectal duplication cyst was made.

At open laparotomy performed via a lower midline incision and a 15 cm × 8 cm cystic appendiceal mass found in the pelvis adjacent to the rectum (Fig. 1). Appendicectomy was performed without incident and he made an uneventful recovery. The histology confirmed mucinous cystadenoma of the appendix. CEA levels repeated after surgery fell significantly and returned to normal after 3 months. These were repeated 6 months after surgery and the levels remained normal.

4. Discussion

The cases presented here are unique since their presentation was characterized by elevated serum CEA levels, a phenomenon that has rarely been reported. This finding has been documented previously in cases of cystadenocarcinoma, which is a rare malignant variant of cystadenoma. There have been at least two other reports in the literature of elevated CEA levels with cystadenoma. This apparent rarity may be explained by the fact that routine CEA levels are not usually requested in patients with cystadenoma, but can be explained by the fact that this antigen is often produced by neoplasms of the colon.

CEA levels can be elevated in a number of medical conditions and an abnormal level may be associated with pancreatic, breast, thyroid, gastric colon and lung disease. It is also associated with benign conditions such as inflammatory bowel disease and cirrhosis and it may also be elevated in 3% of healthy individuals. Review of the literature on mucocoeles, reveals that 50% of patients are asymptomatic and in 50% it is an incidental finding at the time of surgery.

Preoperative diagnosis is difficult because of the paucity of symptoms and the non-specific presentation of these patients. Typically non-invasive modalities are employed namely ultrasound and CT scans. Computed tomography scans have been used in the identification of mucocoeles and characteristically show a well encapsulated, round, thin walled cystic mass with areas of calcification in 55% of cases. The appearance of enhancing nodules in the mucocoele wall suggests a diagnosis of cystadenocarcinoma. Mucocoeles less than 2 cm are rarely malignant. Larger mucocoeles 6 cm or more in size may be associated with cystadenoma or cystadenocarcinoma with a higher perforation rate of 20%. Other CT findings that may suggest malignancy include visual scalloping which results from mucin producing cells adherent to sites of relative stasis dislodged by peristaltic movement. Ultrasound may be helpful and may show a cystic, encapsulated lesion, firmly attached to the caecum and sometimes an “onion skin” like appearance. Colonoscopy is occasionally useful and may reveal a yellowish submucosal mass over the caecum. In rare circumstances, the appearance of the appendiceal orifice in the centre of the mound has been described as the volcano sign. Despite the use of several diagnostic modalities, the correct diagnosis is realized in only 30% of patients. Patients who are diagnosed with a mucocoele of the appendix require full evaluation for the presence of other tumours.

The extent of the surgical treatment is based on the suspected pathology. Appendicectomy is recommended for simple mucocoele or for cystadenoma. Frozen section of the base of the appendix has been recommended to rule out the presence of a cystadenocarcinoma. Simple appendectomy and resection of the mesentery is also thought to be adequate even in patients with cystadenocarcinoma without mesenteric lymph node or adjacent organ involvement. Whenever there is doubt, caecal resection or right hemicolectomy is advised. All patients should have complete exploration of the abdomen regardless of the approach used. Surgical resection for benign mucocoele of the appendix, mucosal hyperplasia and mucinous cystadenoma provides a uniformly good outcome with 5-year survival rates of 90–100% reported. Recurrent disease caused by pseudomyxoma peritonei or the presence of metachronous colonic carcinoma has rarely been reported. Cystadenocarcinoma, which does not involve the base of the appendix or adjacent organs, is also associated with an excellent outcome. Five-year survival rates fall to 25% in cases with peritoneal involvement or pseudomyxoma peritonei.

5. Conclusion

Cystadenoma of the appendix remains a diagnosis that is rarely considered prior to elective surgery. If the diagnosis is made postoperatively careful abdominal imaging is useful for excluding the presence of peritoneal disease or other concomitant malignancies. The presence of an elevated CEA requires careful investigation to exclude malignancy and requires detailed pathological examination of the resected specimen to identify atypical pathological features.
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References
1. Yakan S, Caliskan C, Uguz A, Korkut M, Coker A. A retrospective study on mucocoele of the appendix presenting with acute abdomen or acute appendicitis. Hong Kong Journal of Emergency Medicine 2011;18(3):144–9.
2. Minini F, Petrella M, Morgante A, Santini D. Giant mucocoele of the appendix. Diseases of the Colon & Rectum 2001;44:1034–6.
3. Fujitani T, Hizuta A, Iwashita I, Masuno T, Hamada M, Tanaka N. Appendiceal mucocoele with concomitant colonic cancer. Diseases of the Colon & Rectum 1996;39:232–6.
4. Woodruff R, McDonald JR. Benign and malignant cystic tumors of the appendix. Surgery, Gynecology and Obstetrics 1940;71:750.
5. Higa E, Rosai J, Pizzimbono CA, Wise L. Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix: a re-evaluation of appendiceal mucocoele. Cancer 1973;32:1525–41.
6. Zullo A, Botto G, Pasturmero M. A case of appendiceal mucocoele associated with colon cancer. Minerva Chirurgica 1995;50(12):1095–8.
7. Versaci A, Rossitto M, Giuffre M, Leonello G, Pagano G, Terranova M. Mucocoele of the appendix with concomitant colonic cancer: case report. Giornale di Chirurgia 1999;20:397–401.
8. Noritake N, Ito Y, Yamakita N, Asuma S, Shimokawa K, Miura K. A case of primary mucinous cystadenocarcinoma of the appendix with elevated serum carcinoembryonic antigen (CEA). Japanese Journal of Medicine 1992;29:642–6.
9. Shimizu T, Shimizu M, Kawaguchi K, Yomura W, Ihara Y, Matsumoto T. Mucinous cystadenoma of the appendix with raised serum carcinoembryonic antigen concentration: clinical and pathological features. Journal of Clinical Pathology 1997;50:613–4.
10. Oshimo Y, Otsu N, Ota M, Inamura M, Kim S, Masanga T, Hirano N, Nakashima K. A case of appendiceal mucocoele with elevated CEA level in the cystic fluid detected by colonoscopic examination. Gastroenterological Endoscopy 2000;42(1):41–5.
11. Takahashi S, Furukawa T, Ueda J. Case report: mucocoele of the tip of the appendix. Clinical Radiology 1998;53:149–50.
12. Pai RK, Longacre TA. Appendiceal mucinous tumors and pseudomyxoma peritonei: histologic features, diagnostic problems, and proposed classification. Advances in Anatomic Pathology 2005;12:291–311.
13. Aho AJ, Heinonen R, Lauren P. Benign and malignant mucocoele of the appendix. Histological types and prognosis. Acta Chirurgica Scandinavica 1973;139:392.
14. Pickhardt PJ, Levy AD, Rohrmann CA, Kende AI. Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. Radiographics 2003;23:645–62.
15. Isacs KL, Warshawer DM. Mucocoele of the appendix: computer tomographic, endoscopic, and pathologic correlation. American Journal of Gastroenterology 1992;87:787–9.
16. Chion SS, Pitman MB, Hahn PF, et al. Rare benign and malignant appendiceal lesions: spectrum of computed tomography findings with pathologic correlation. Journal of Computer Assisted Tomography 2003;27:622–5.
17. Haritopoulos KN, Brown DC, Lewis P, Mansour F, Eltayar AR, Labruzzo C, Hakim NS. Appendiceal mucocoele: a case report and review of the literature. International Surgery 2001;86(October–December (4)):259–62.
18. Ponsky JL. An endoscopic view of mucocoele of the appendix. Gastrointestinal Endoscopy 1976;23:42–3.
19. Hamilton DL, Stormont JM. The volcano sign of appendiceal mucocoele. Gastrointestinal Endoscopy 1989;35:453–6.
20. Ruiz-Tovar J, Teruel DG, Castiñeiras VM, Dehesa AS, Quindós PL, Molina EM. Mucocoele of the appendix. World Journal of Surgery 2007;31(March (3)):542–8.

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