Mucinous Cystadenocarcinoma Of The Appendix: A Management Dilemma

A Bhat, H Shinoy, P Bhat

Citation

A Bhat, H Shinoy, P Bhat. *Mucinous Cystadenocarcinoma Of The Appendix: A Management Dilemma*. The Internet Journal of Surgery. 2004 Volume 6 Number 1.

Abstract

Appendiceal neoplasms are a rare entity and account for less than 0.5% of all intestinal tumours. In most cases, a pre-operative or intra-operative diagnosis is rarely achieved and patients frequently require a second operation for definitive therapy. Once diagnosed, the traditional treatment for a mucinous cystadenocarcinoma of the appendix has been surgical in the form of a right hemicolectomy and aggressive debulking added if found associated with pseudomyxoma peritoneii (intra-abdominal metastasis) which is seen in upto 50% of patients. We report a case of a mucinous cystadenocarcinoma of the appendix which presented as a mass in the right iliac fossa and the problems encountered during surgery.

CASE REPORT

A 42 year old male patient attended the surgical outpatient with pain in the right iliac fossa, fever and vomiting of 10 days duration. He had consumed varying antibiotics for what was suspected to be acute appendicitis. On examination, he was febrile (102o F) had a pulse rate of about 100 beats/minute and was normotensive. A firm, tender mass of about 8 x 10 cms was found in the right iliac fossa. Remainder of the abdomen was soft and bowel sounds were absent. Clinically, a diagnosis of the lesion being an appendicular abscess was considered. Laboratory investigations revealed leucocytosis (total w.b.c. count of 14100) with 85% neutrophils. An ultrasound examination of the abdomen showed a dilated, non-compressible, aperistaltic tubular lesion in the right iliac fossa with an adjacent isoechoic lesion with echogenic foci. The possibility of the lesion being an inflamed appendix with mucocoele was suggested.

In view of the clinical suspicion of an appendicular abscess, an emergency laparotomy was performed via a right lower paramedian incision. A 6 x 5 cm mass was found about 2 cms distal to the base of the appendix, consisting of a dilated, inflamed appendix and adjacent loops of matted small bowel. The Caecum and the base of the appendix were found to be separate from the mass. On attempting to separate the bowel loops from the mass, 15 – 20 ml of yellow, non foul-smelling, thick fluid was found. Appendicectomy was performed, stump invaginated, a drain was placed in the right iliac fossa and the abdomen was closed. On the second post-operative day patient developed pain and swelling of the left leg. Deep vein thrombosis was suspected and a Duplex scan confirmed thrombosis of the left popliteal vein for which the patient was started on heparin infusion. The histopathological report of the specimen was available on the fifth post-operative day and had found the lesion to be a mucinous cystadenocarcinoma of the appendix. The diagnosis was discussed with the patient and definitive surgery in the form of a right hemicolectomy was proposed, but deferred in view of his recent DVT.

Two weeks after his first operation, and after bowel preparation and a negative fibreoptic colonoscopic examination, a laparotomy was performed. Dense adhesions were found between the bowel loops and the anterior abdominal wall, and loops of bowel were also found to be plastered to each other in the right iliac fossa. On attempting separation of the bowel loops, serosal tears were caused in a couple of areas. Serosal tears were sutured and the procedure had to be abandoned because of the technical difficulty. Patient was explained about the difficulties encountered and the need for another operation later on for definitive therapy to which he expressed his unwillingness. Considering the possibility of local dissemination of the disease following spillage, patient was offered chemotherapy and has completed six cycles of single agent 5-fluorouracil given for 5 days in a month. Patient has been doing well and visiting

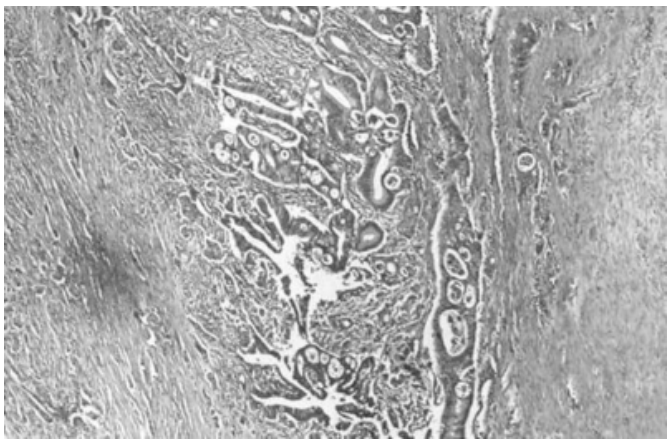
the hospital for follow-up regularly after his surgery and at the end of 2 years has no clinical or sonological evidence of residual or recurrent disease.

DISCUSSION

Appendiceal mucinous cystadenocarcinoma is a rare tumour and accounts for less than 0.5% of all intestinal tumours.[1] The age of affection of a mucinous cystadenocarcinoma is lower than the typical patient with an adenocarcinoma of the appendix.[2] In 15-20% of cases, a second malignancy in the gastrointestinal tract is associated with the disease.[2] Malignancies of the appendix are never suspected pre-operatively and seldom post-operatively, the diagnosis being usually made at histopathology of the specimen.[3,4] In the event of finding an 'atypical' appendix during surgery a prompt frozen section examination might help to obviate the need for a second operation for definitive surgery on the occasion of having discovered a malignancy of the appendix.[1] At histopathology, invasion of the appendiceal wall by atypical glands and identification of epithelial cells in any intraperitoneal mucinous collection are the features that suggest the diagnosis of a malignancy.[1] (Fig. 1)

Figure 1

Figure 1: Epithelium seen undergoing malignant change with formation of glands of varying sizes and shapes lined by columnar cells having hyperchromatic and pleomorphic nuclei, and eosinophelic to vacuolated cytoplasm infiltrating all the layers of the appendix. (H & E x 50)



Once a diagnosis of a mucinous cystadenocarcinoma has been achieved, either by frozen section examination during surgery or at histopathological examination post-operatively, a right hemicolectomy has been the accepted form of definitive therapy.[5] These tumours have a slower growth rate which translates to longer survival than with appendiceal adenocarcinomas.[2] Surgical removal of the tumour when detected early is said to offer a cure. The 10-year survival after right hemicolectomy has been found to be 65% in contrast to 37% in patients subjected to appendicectomy alone.[2] Patients with disseminated disease are benefited by aggressive debulking, omentectomy and oophorectomy and often require repeated operations for intestinal obstruction or fistulation.[2] Radiotherapy and chemotherapy have not been found to be effective in the setting of disseminated disease.[2] The role of adjuvant therapy in the isolated case with the possibility of tumour spillage at surgery, or infiltration of adjacent viscera is a question that might never be answered in view of the rarity of the disease.

CORRESPONDENCE TO

Dr. Akhil Krishnanand Bhat Department of Surgery, Kasturba Medical College & Hospital, MANIPAL – 576104, INDIA. E-mail: akhil.bhat@kmc.manipal.edu Phone: 91-820-2571201 Extn: 22213. Facsimile: 91-820-2570061

References

1. Rutledge RH, Alexander JW, Primary appendiceal malignancies: rare but important. Surgery 1992; 111:244-250.
2. Lee J, Gardiner KR, Wilson BG. Incidental mucinous cystadenocarcinoma of the appendix. Postgrad Med J. 1996; 72(843):55-56.
3. Gilhome RW, Johnston DH, Clark J, Kyle J. Primary adenocarcinoma of the vermiform appendix: report of a series of ten cases, and review of the literature. Br. J. Surg. 1984; 71:553-555.
4. Caponnetto A, Bergantino A. Mucinous cystadenocarcinoma of the appendix. A rare case. Minerva Chir. 1997 Jan-Feb; 52(1-2):103-5.
5. Lo NS, Sarr MG. Mucinous cystadenocarcinoma of the appendix. The controversy persists: a review. Hepatogastroenterology. 2003 Mar-Apr; 50(50):432-7.

Author Information

Akhil Krishnanand Bhat, MBBS, MS
Kasturba Medical College and Hospital

H. Divakar Shinoy, MBBS, MS
Kasturba Medical College and Hospital

Padmanabh R. Bhat, MS, FRCS
Kasturba Medical College and Hospital