E-ISSN: 2616-3470
P-ISSN: 2616-3462

www.surgeryscience.com
2022; 6(4): 74-76
Received: 08-06-2022
Accepted: 13-07-2022

Dr. Manaswi Ganvir
Junior Resident, Department of General Surgery, Dr. D.Y. Patil Medical College and Hospital, Navi Mumbai, Maharashtra, India

Dr. Shshank Jain
Assistant Professor, Department of General Surgery, Dr. D.Y. Patil Medical College and Hospital, Navi Mumbai, Maharashtra, India

Dr. Kalpesh Chaudhari
Assistant Professor, Department of General Surgery, Dr. D.Y. Patil Medical College and Hospital, Navi Mumbai, Maharashtra, India

Corresponding Author:
Dr. Manaswi Ganvir
Junior Resident, Department of General Surgery, Dr. D.Y. Patil Medical College and Hospital, Navi Mumbai, Maharashtra, India

Diagnosis and management of retroperitoneal mucinous adenocarcinoma possibly originating from previously operated appendix: A rare case

Dr. Manaswi Ganvir, Dr. Shshank Jain and Dr. Kalpesh Chaudhari

DOI: https://doi.org/10.33545/surgery.2022.v6.i4b.952

Abstract
Retroperitoneal mucinous adenocarcinoma [RPMA] is a rare condition usually diagnosed incidentally, after surgery. The study reports a case of incidentally diagnosed retroperitoneal mucinous adenocarcinoma in a 78-year-old male. The retroperitoneal cystic mass was resected by careful dissection from the surrounding structures and samples from the cyst and cystic fluid were sent for histopathological evaluation. The histopathological examination suggested a mucinous adenocarcinoma possibly arising from the previously operated appendix. Following the surgery, the patient was stable, however, he suffered a fatal cardiac arrest on day 9 post-operatively. The patient could not be revived despite cardiopulmonary resuscitation.

Keywords: Retroperitoneal mass, mucinous adenocarcinoma, case report, cystic mass

Introduction
RPMA is a rare condition, as evidenced by the few examples published since Roth initially defined it in 1977 [1]. This tumour's histogenesis is yet unknown. RPMA, like other retroperitoneal masses, may produce clinical symptoms or is noticed by patients only when it grows significantly larger in size. Preoperative diagnosis of RPMA is challenging since tumour markers such as CA-125, CEA, and CA19-9 may not increase and may lack specificity, making it difficult to distinguish the lesion from other tumours such as ovarian cyst, cystic mesothelioma, cystic lymphangioma, non-pancreatic pseudocyst, and renal cyst [2]. Ultrasonography, computed tomography, and magnetic resonance imaging are frequently utilised to localise the tumour. However, these modalities are incapable of distinguishing between benign and malignant neoplasms [3]. Surgeons should consider RPMA in the preoperative diagnosis of a retroperitoneal cystic lesion that displaces the colon, kidney, or ureter medially, according to Yang *et al*. [4].
Surgical resection is the conventional treatment for RPMA, as chemotherapy has not been proven to be an effective treatment option for this tumour [5].

Case Report
History
This is the case of a 78-year-old male who attended the surgical out patient department (OPD) with an abdominal lump that was gradually increasing in size for 2 years accompanied by constipation and reduced appetite. He had a history of diabetes and hypertension. However, he was not on any medications for the same. Patient was vitally stable and routine investigations were normal on admission. The patient had an operative history of appendicectomy 6 years ago.

Examination
On examination, the lump was found to be 20 cm × 20 cm, soft and nontender. It was present in the right lumbar region extending to the right iliac region. The lump was globular, firm, non-tender, nonmobile, had dilated veins on its surface, well defined, non-ballotable and parietal.

Investigations (Biochemical and radiological)
Patient was further evaluated by performing computed tomography of abdomen and pelvis which was suggestive of large right retroperitoneal cystic lesion more likely representing retroperitoneal paraganglioma.

Fluorodeoxyglucose positron emission tomography (FDG PET/CT) scan of the whole body reported a multilobulated cystic lesion in the right lumbar region extending into the pelvis on the right side with a low-grade metabolic activity in the periphery.

Management

Resection of the cystic mass was carried out by careful dissection around the cyst wall, adhesions between bowel and cyst were broken with fine dissection and cyst was excised. Intraoperative findings suggested a retroperitoneal mass was attached laterally to the lateral peritoneal wall, medially attached to ileo-caecal junction, caecum, ascending colon and posteriorly the mass was extending till the psoas muscle. Samples from the cyst (Figures 1 and 2) and cystic fluid were sent for histopathological evaluation. The cyst was approached retro-caecally and through the right paracolic gutter during which the cyst ruptured with the spillage of its contents in the paracolic gutter. The histopathology reports suggested a mucinous adenocarcinoma possibly arising from the previously operated appendix.

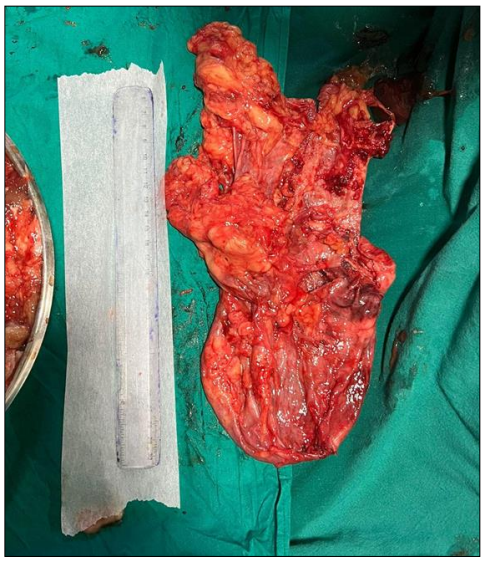

Fig 1: Image depicting size of the mass

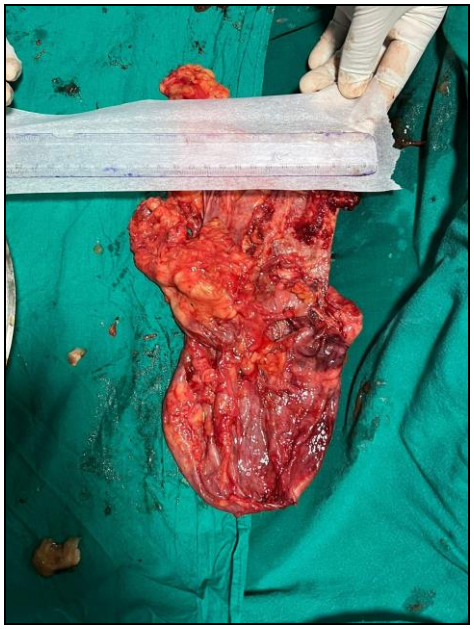

Fig 2: Image depicting size of the mass

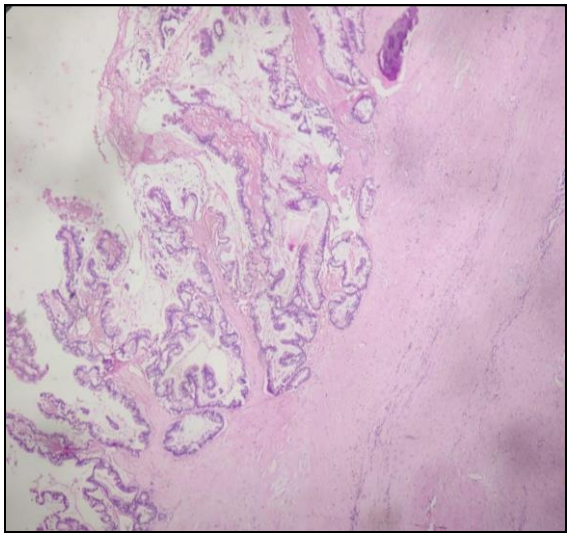

Fig 3: Image showing histopathological findings

Outcome

On postoperative day 2, the right sided subhepatic drain showed bilious with feculent matter hence the patient was taken for emergency exploratory laparotomy. Intraoperative findings suggested a spontaneous ileal perforation with gangrenous caecum. During the resection of the cyst, the large intestine along with its mesentery was mobilised to the left, which might have led to the occlusion of the mesenteric vascular pedicle causing gangrenous changes of the caecum. Resection of the caecum and terminal ileum followed by ascending colon-ileal anastomosis with loop ileostomy was done. The sample was sent for histopathological evaluation. The patient was stable during the immediate postoperative period. Postoperative leucocyte counts were increasing steadily. So, the patient was shifted from intravenous administration of meropenem to tigecycline. Unexpectedly, the patient suffered a fatal cardiac arrest on day 9 post-operatively. The patient could not be revived despite cardio-pulmonary resuscitation.

Histopathology

Histopathological examination of the resected specimens of the cyst wall, cyst fluid and of caecum with ileum was done. Histopathology report showed (Figure 3) large multilobulated cyst with focal papillary area and fibro-collagenous and fibromuscular wall lined largely by foamy macrophages. Very focally, mucin-containing columnar cell lining with goblet cells (intestinal type epithelium) displaying features of dysplasia and superficial invasion into the cyst wall were seen. Diagnosis in favour of mucinous adenocarcinoma possibly arising from appendix/caecum was reported, considering the mass was closely adherent to the caecum and appendix was not visualised intraoperatively. Section studied from a single segment of the intestine had features which were consistent with multiple perforations (2 ileal and 1 caecal) with submucosal lipoma. Immunohistochemistry with CDX2 shows positivity in the lining epithelium.

Discussion

Because of its rarity, RPMA poses a diagnostic challenge to surgeons. There are four primary hypotheses that have been offered to explain the tumour's histogenic origin, so far. According to one theory, the tumour develops from a teratoma with significant mucinous epithelium [6], whereas other authors postulate that it is caused by intestinal duplication, also known

as enterogenous genesis [7]. In our case, the intestinal-like epithelioma encircling the cystic tumours may provide support to this theory. The third theory proposes that the tumour develops from heterotopic ovarian tissue [5]. Previously, a fourth hypothesis stating that tumours start from peritoneal epithelial invagination and undergo metaplasia during foetal growth was also widely accepted [8].

Laparotomy is required to allow for proper decision-making and treatment. The investigators have reached an agreement on the total excision of the lesion. However, the extent of the procedure is still debatable. In addition to oophorectomy, total hysterectomy was advised by Lee *et al*. on the basis that RPMA is thought to occur in heterotopic ovarian tissue. However, if the uterus and ovaries are macroscopically normal, hysterectomy and salpingo-oophorectomy are not appropriate for the treatment of RPMA. Laparoscopic removal of the tumour has been recommended to preserve these women's fertility. RPMA affects predominantly women, with only a few male cases reported in the literature [5], making this case-report admissible.

Chemotherapy for RPMA is not well established because the benefits of adjuvant chemotherapy have not been proven. Lee *et al*. described 5 patients who received adjuvant chemotherapy after resection. One of these patients suffered para-ovarian recurrence after receiving cyclophosphamide treatment for 21 months, and two died from extensive metastases 4 months and 18 months following surgery, respectively [9]. Certain authors have claimed that because RPMA's histogenesis is similar to that of ovarian mucinous tumour, chemotherapy should be administered, as it is in the case of this later tumour. Tenti *et al*. reported two cases of RPMA with cystic rupture; the patient who had adjuvant chemotherapy was tumour-free for 33 months after surgery [10]. Chemotherapy is advocated in cases where the tumour had ruptured during surgery or had infiltrated neighbouring structures [5]. Paclitaxel in combination with cisplatin or carboplatin treatment may potentially be successful.

Conclusion

RPMA poses both diagnostic and therapeutic challenges to the surgeons. It warrants accurate preoperative imaging to localise the tumour and vigilant surgical technique to remove the tumour in toto. Adjuvant chemotherapy may be useful in complex cases. By virtue of their rarity, each case of RPMA should be reported to add evidence in the literature and aid future-generation surgeons to manage this rare entity accordingly.

Conflict of Interest

Not available

Financial Support

Not available

References

1. Roth LM, Ehrlich CE. Mucinous cystadenocarcinoma of the retroperitoneum. Obstet Gynecol [Internet]. [Cited 2022 Oct 12], 1977 Apr, 49(4). Available from: https://pubmed.ncbi.nlm.nih.gov/854250/
2. Tangjitgamol S, Manusirivithaya S, Sheanakul C, Leelahakorn S, Thawaramara T, Kaewpila N. Retroperitoneal mucinous cystadenocarcinoma: a case report and review of literature. Int J Gynecol Cancer [Internet]. [cited 2022 Oct 12], 2002 Jul, 12(4). Available from: https://pubmed.ncbi.nlm.nih.gov/12144691
3. Matsubara M, Shiozawa T, Tachibana R, Hondo T, Osasda K, Kawaguchi K, *et al*. Primary retroperitoneal mucinous cystadenoma of borderline malignancy: a case report and review of the literature. Int. J Gynecol Pathol [Internet]. [cited 2022 Oct 12], 2005 Jul, 24(3). Available from: https://pubmed.ncbi.nlm.nih.gov/15968195/
4. Yang DM, Jung DH, Kim H, Kang JH, Kim SH, Kim JH, *et al*. Retroperitoneal cystic masses: CT, clinical, and pathologic findings and literature review. Radiographics [Internet]. [cited 2022 Oct 12], 2004 Sep, 24(5). Available from: https://pubmed.ncbi.nlm.nih.gov/15371613/
5. Jiang H, Jin K, You Q, Fang W, Xu N. Retroperitoneal primary mucinous adenocarcinoma: A case report. Oncol Lett. 2011 Jul;2(4):633.
6. Papadogiannakis N, Gad A, Ehliar B. Primary retroperitoneal mucinous tumor of low malignant potential: histogenetic aspects and review of the literature. APMIS. 1997 Jun;105(6):483-6.
7. Vara TC, Gustein D, Salvi M, Plata J. [Retroperitoneal enteroid cystadenocarcinoma (possible intestinal origin)]. Rev Esp Enferm Apar Dig [Internet]. [cited 2022 Oct 12], 1984 Oct, 66(4). Available from: https://pubmed.ncbi.nlm.nih.gov/6515089/
8. Subramony C, Habibpour S, Hashimoto LA. Retroperitoneal mucinous cystadenoma. Arch Pathol Lab Med [Internet]. [cited 2022 Oct 12], 2001 May, 125(5). Available from: https://pubmed.ncbi.nlm.nih.gov/11300948/
9. Lee SA, Bae SH, Ryoo HM, Jung HY, Jang SB, Kum YS. Primary Retroperitoneal Mucinous Cystadenocarcinoma: A Case Report and Review of the Literature. Korean J Intern Med. 2007 Dec;22(4):287.
10. Tenti P, Carnevali L, Tateo S, Durola R. Primary mucinous cystoadenocarcinoma of the retroperitoneum: two cases. Gynecol Oncol [Internet]. [cited 2022 Oct 12], 1994 Nov, 55(2). Available from: https://pubmed.ncbi.nlm.nih.gov/7959300/

How to Cite This Article

Ganvir M, Jain S, Chaudhari K. Diagnosis and management of retroperitoneal mucinous adenocarcinoma possibly originating from previously operated appendix: A rare case. National Journal of Clinical Orthopaedics. 2022;6(4):74-76.