Pseudomyxoma extraperitonei: a rare presentation of a rare tumour

Carmina Diaz-Zorrilla,¹ Antonio Ramos-De la Medina,¹ Peter Grube-Pagola,² Alfredo Ramirez-Gutierrez de Velasco¹

¹Department of Surgery, Hospital Español de Veracruz, Veracruz, Mexico
²Department of Pathology, Hospital Español de Veracruz, Veracruz, Mexico

Correspondence to
 Dr Antonio Ramos-De la Medina, ramos.antonio@inbox.com

SUMMARY

Pseudomyxoma extraperitonei is rare lesion resulting from the rupture of an appendiceal mucocele into the extraperitoneal tissues. We report a case of an 80-year-old woman with a medical history for a left hemicolectomy and a laparoscopic cholecystectomy 11 and 6 years, respectively, referred to our hospital for abdominal pain of increasing severity localised to the right hemiabdomen. The abdominal examination revealed a mobile mass a multidetector CT was performed; the patient was taken to surgery which was performed with no complications. Histopathological analysis of the tumour reported a pseudomyxoma associated to a moderately differentiated adenocarcinoma. The patient remains asymptomatic at a 1-year follow-up.

BACKGROUND

A rare presentation of a rare tumour.

CASE PRESENTATION

An 80-year-old female patient was referred to our hospital with a 3-year history of abdominal pain of increasing severity localised to the right hemiabdomen. Her medical history was relevant for a left hemicolectomy for colonic poliposis and a laparoscopic cholecystectomy for gallstone disease 11 and 6 years, respectively, prior to her admission.

INVESTIGATIONS

On physical examination, she did not appear to be in acute distress. The abdominal examination revealed a mobile mass located to the right of the midline that extended from just below the costal border to the right iliac crest that was painful to palpation. There were no cervical, axillary or inguinal lymphadenopathies and there was not ascites. The rest of her exam was irrelevant. A multidetector CT (MDCT) was performed that showed a well-circumscribed mass anterior the right rectus muscle that measured 251×92 mm. The appendix and ovaries were not visible (figures 1 and 2).

TREATMENT

The patient was taken to operation where an extra-peritoneal encapsulated mass was found without any involvement of neighbouring structures or intra-abdominal organs. It was completely resected without breaking the capsule or entering the abdominal cavity. Histopathological analysis reported a pseudomyxoma associated to a moderately differentiated adenocarcinoma. Immunohistochemistry was positive for CK20 and CDX2 and negative for CK7 and calretinin (intestinal origin) (figure 3).

OUTCOME AND FOLLOW-UP

The patient was discharged 2 days after the surgery without any complications and continues to do well after 1-year follow-up.

Figure 1 A multidetector CT showing an extraperitoneal mass.

To cite: Diaz-Zorrilla C, Ramos-De la Medina A, Grube-Pagola P, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-007702

Figure 2 A multidetector CT showing an extraperitoneal mass.

DISCUSSION

Pseudomyxoma peritonei was first described by Werth¹ in 1884 as a rare condition consisting of mucinous ascites. In 1948, Bonann² reported a pseudomyxoma involving only the retroperitoneum and it was first described by Coppil³ in 1950. Twenty years later, Early⁴ described a retroperitoneal mucocoele of the appendix which contained 10 litres of mucus that had not ruptured and a complete curative excision was possible; this was termed by Moran⁵ as *pseudomyxoma extraperitonei* (PE) in 1988. Shelton *et al*⁶ later named it pseudomyxoma retroperitonei. Pseudomyxoma extraperitonei is a rare condition resulting from

the rupture of an appendiceal mucocoele into the extraperitoneal tissues. It can also arise from implantation of mucosal cells of the primary site accompanied of modified glandular cells with the ability to behave in an invasive tumour-like manner. Tumours of the appendix are very rare, with mucinous adenocarcinomas constituting 8% of all malignant neoplasms of the appendix with an estimated incidence of 0.2/100 000 population/year.⁷ Mucinous implants in the peritoneal cavity are almost always of appendiceal or ovarian origin.⁴ In a recent report, van Ruth *et al*⁸ suggest that the appendix is the primary source of most pseudomyxoma peritonei, which may then spread to sites like the ovaries. Unusual

Figure 3 (A) Neoplastic epithelium with complex architecture, also appreciate abundant mucin (H&E, $\times 5$). (B) Note cytological atypia and loss in nuclear polarity (haematoxylin and eosin, $\times 40$). (C) CK20 and (D) CDX2 positive in neoplastic epithelium.

Table 1 Pseudomyxoma extraperitonei reported cases

Age	Sex	Presentation	Primary tumour	Treatment	Outcome	Reference
74	M	Abdominal pain and mass	Appendiceal mucinous adenocarcinoma	Debulking right hemicolectomy chemotherapy	Recurrence 6 months, fistula year	Ioannidis ¹⁴
68	F	Lumbar pain and mass	Appendiceal mucinous adenocarcinoma	Debulking right hemicolectomy chemotherapy	Disease free at 3 years.	Ioannidis ¹⁴
48	F	Abdominal pain and mass	Ovary	Debulking right oophorectomy and chemotherapy	Recurrence at 4 years	Chamisa ¹⁵
51	F	Right flank purulent and mucinous discharge, palpable mass.	Appendiceal mucinous adenocarcinoma	En block resection with a portion of iliac bone appendectomy, systemic chemotherapy and radiotherapy	NA	Cakmak ¹⁶
80	F	Lower abdominal mass	Appendiceal mucinous adenocarcinoma	Resection of mucous and ileum, cecum both ovaries and uterus	NA	Niwa ¹⁷
57	M	Lower abdominal pain and mass	NA	Excision of cyst radiotherapy chemotherapy	Alive and disease free at years	Solkai ¹⁸
55	F	Right lower abdominal pain and mass, retroperitoneal abscess	Ascending solon cancer	Right hemicolectomy oophorectomy, mucous removal. Systematic chemotherapy	Alive, no recurrence 5 months	Hirokawa ¹⁹
78	F	Right abdominal tumour and pain	Appendiceal cystadenocarcinoma	Right hemicolectomy	NA	Kojima ²⁰
NA	M	NA	Mucinous paraenteric cyst	NA	NA	Angelescu ²¹
68	F	NA	Appendiceal mucinous adenocarcinoma	Appendectomy debulking of mucous, intraoperative chemotherapy, systemic chemotherapy	Recurrence at 5 month, disease free at 3 and a half years	Liu ²²
58	F	Abdominal pain, retroperitoneal mass	Mucinous cystadenoma (primary or secondary)	NA	NA	Matsuoka ²³
46	M	Skin fistula, abdominal pain and mass	Appendiceal mucinous adenocarcinoma	Right hemicolectomy	Recurrence at 3 months	Koizumi ²⁴
53	F	Retroperitoneal abscess	Appendiceal mucinous adenocarcinoma	Right hemicolectomy and debulking systemic chemotherapy	Alive at 1 year	Edrees ²⁵
NA	NA	NA	NA	NA	NA	Fujimura ²⁶
69	M	Abdominal mass and pain, weight loss	Appendiceal mucinous adenoma	Debulking	NA	Tsai ²⁷
NA	NA	NA	NA	NA	NA	Hemet ²⁸
NA	NA	NA	NA	NA	NA	Oyama ²⁹
56	M	Appendiceal abscess	Appendiceal mucinous adenocarcinoma	Drainage, radiotherapy	Recurrence, Death 39 months	Stevens ³⁰
65	M	Abdominal mass	Appendiceal cystadenocarcinoma	Debulking and appendectomy	NA	Mor ³¹
39	NA	Right back pain and right lower abdominal mass	Appendiceal mucinous adenocarcinoma	Right hemicolectomy, resection of right iliopsoas muscle, partial peritonectomy	NA	Tamai ³²
NA	NA	NA	NA	NA	NA	Nobusawa ³³
NA	NA	Polyuria and back pain	Appendiceal mucinous cystadenoma	NA	NA	Baba ³⁴
81	M	Abdominal mass	Appendiceal mucinous cystadenoma	Debulking and appendectomy	Persistent discharge	Shelton ⁶
47	M	Retroperitoneal mass with anterior thigh extension	NA	Excision	Disease free at 3 years	Fann <i>et al</i> ³⁵
NA	NA	NA	NA	NA	NA	Koyama ³⁶
NA	NA	NA	Appendix	NA	NA	Vladimirtseva ³⁷
58	M	Right loin pain and abdominal mass	NA	Drainage, appendectomy and repeat drainage. Chemotherapy, radiotherapy mucolytic agents, radiofrequency hyperthermia	Recurrence at 3 weeks, recurrence at 5 years with sinus formation. Alive at 10 years	Moran ⁵
67	M	Flank pain and mass	Appendix	Surgical debulking retroperitoneal chemotherapy	Recurrence at 2 years. Recurrence at 5 years	Brady ³⁸
NA	NA	NA	NA	NA	NA	Shimabukuro ³⁹
NA	NA	NA	NA	NA	NA	Torgunakov ⁴⁰
NA	NA	NA	NA	NA	NA	Kayser ⁴¹
NA	NA	NA	Appendix	NA	NA	Chetcheva ⁴²
NA	NA	NA	Appendiceal cyst	NA	NA	Chekharina ⁴³
NA	NA	NA	Probable appendicular	NA	NA	Coppini ³
37	M	Tender abdominal mass	Mucocele of appendix	Evacuation of cyst in 2 stage procedure	NA	Bonnan ²
57	M	Abdominal mass	Mucocele of appendix	Debulking of tumour	Alive at 2 years	Early ⁴
50	F	Total uterine prolapse	Appendix	Excision, omentectomy, total hysterectomy and oophorectomy	No evidence of disease 2 years	Snyder and Vandivort. ⁴⁴

Continued

Table 1 Continued

Age	Sex	Presentation	Primary tumour	Treatment	Outcome	Reference
63	M	Retroperitoneal mass	Appendix	Excision	No evidence of disease	Early <i>et al</i> ⁴
41	M	Pleural mass	Appendix	Chemotherapy	Death of disease 2.5 years	Radosavljevic <i>et al</i> ⁴⁵
65	M	Inguino labial hernia	Appendix	Excision	Alive with disease 9 years	Ben-Hur <i>et al</i> ⁴⁶
33	M	Scrotal mass	Rectum	Debulking, omentectomy	No evidence of disease at 9 months	Baker <i>et al</i> ⁴⁷
38	M	Pleural mass	Appendix	Excision, chemotherapy	Alive with disease at 2.5 years	Peek and Beets ⁴⁸
53	M	Retroperitoneal mass with skin fistula	Appendix	NA	NA	Koizumi and Noiguchi ⁴⁹
75	M	Scrotal mass	Appendix	Chemotherapy	Dead of disease	Al-Bozom ⁹

sources of mucinous adenocarcinoma causing pseudomyxoma peritonei have included colon, uterus, common bile duct, pancreas and the stomach. There are over 44 cases of PE or retroperitonei reported in the literature, most commonly presented as an extra-peritoneal abdominal mass (table 1).

The pathogenesis for extraperitoneal pseudomyxoma peritonei in most cases is explained by one of the following reasons: (1) a communication through hernial sac; (2) increased intra-abdominal pressure (cases of uterovaginal prolapse) or (3) a leak through the peritoneum (retroperitoneal presentation associated with intraperitoneal pseudomyxoma). In the absence of peritoneal pseudomyxoma, a variant of the anatomy of the appendix (retroperitoneal location) may be the explanation; however, it is still speculative.⁹ In the present case, we could not demonstrate a communication with the abdominal cavity. The port sites used for the laparoscopic cholecystectomy did not appear to be the source of access to the preperitoneum. Preoperative diagnosis is very rare; symptoms like fatigue, decreased appetite and slowly progressing pain are common. Ultrasound and MDCT may contribute to the diagnosis, typical findings of pseudomyxoma peritonei are ascites, localised fluid collection throughout the abdomen and pelvis, omental thickening and deformity of the liver surface may be absent in PE.^{10 11} Several treatments have been used. Sugarbaker *et al*¹² advocate radical surgery together with preoperative intraperitoneal and systemic chemotherapy. The reported 5-year survival of patients with Pseudomyxoma peritonei varies from 11% to 75% (mean 50%).¹³ The role of adjuvant treatment in Pseudomyxoma extraperitonei and survival rates are not well known owing to the scarcity of reported cases. However, the disease is benign and the prognosis is likely to be better than Pseudomyxoma peritonei as the vital abdominal structures are not involved.⁵

Learning points

- Preoperative diagnosis is very rare.
- The disease is benign and the prognosis is likely to be better than Pseudomyxoma peritonei as the vital abdominal structures are not involved.
- The rarity of pseudomyxoma extraperitonei makes outcomes difficult to predict.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Werth R. Pseudomyxoma peritonei. *Arch Gynaecol* 1884;24:100–18.
- 2 Bonnan LJ, Davis JG. Retroperitoneal mucocoele of the appendix. A case report with characteristic roentgen features. *Radiology* 1948;51:375–82.
- 3 Coppini B. Su di un rarissimo caso di pseudomixoma retroperitoneale di probabile origine appendicolare. *G Clin Med* 1950;31:601–12.
- 4 Early KS, Stephenson DV, Davis WC. Giant retroperitoneal mucocoele simulating pseudomyxoma peritonei and mucinous adenocarcinoma. *Am J Surg* 1968;116:439–43.
- 5 Moran CJ, Morgan RH. Pseudomyxoma extraperitonei. *J Roy Soc Med* 1988;81:668–9.
- 6 Shelton MW, Morian JP, Radford DM. Pseudomyxoma retroperitonei associated with appendiceal cystadenoma. *Am Surg* 1994;60:958–60.
- 7 Deans GT, Spence RAJ. Neoplastic lesions of the appendix. *Br J Surg* 1995;82:299–306.
- 8 Van Ruth S, Acherman YIZ, van de Vijwer MJ, *et al*. Pseudomyxoma peritonei: a review of 62 cases. *Eur J Surg Oncol* 2003;29:682–8.
- 9 Issam A, Al-Bozom MD. Extraperitoneal presentation of pseudomyxoma peritonei as a scrotal mass: case report and review of the literature. *Ann Saudi Med* 2000;20:3–4.
- 10 Yeh H, Shafir M K, Slater G, *et al*. Ultrasonography and computed tomography in pseudomyxoma peritonei. *Radiology* 1984;153:507–10.
- 11 Seshul MB, Coulam CM. Pseudomyxoma peritonei: computed tomography and sonography. *Am J Roentgenol* 1981;136:803–6.
- 12 Sugarbaker PH, Kern K, Lack E. Malignant pseudomyxoma peritonei of colonic origin: natural history and presentation of a curative approach to treatment. *Dis Colon Rectum* 1987;30:772–9.
- 13 Sherer D, Abulafia O, Eliakim R. Pseudomyxoma peritonei: a review of current literature. *Gynecol Obstet Invest* 2001;51:73–80.
- 14 Ioannidis O, Cheva A, Paraskevas G, *et al*. Pseudomyxoma retroperitonei: report of 2 cases and review of the literature. *Rev Esp Enferm Dig* 2012;104:268–75.
- 15 Chamisa I. Pseudomyxoma extraperitonei of ovarian origin mimicking a psoas abscess. *Gynecol Surg* 2011;8:373–5.
- 16 Cakmak A, Karakayali F, Bayar S, *et al*. Pseudomyxoma retroperitonei presenting with a skin fistula. *Turk J Gastroenterol* 2009;20:79–80.
- 17 Niwa H, Hiramatsu T, Ishihara Y. Clinical challenges and images in GI. Pseudomyxoma retroperitonei. *Gastroenterology* 2007;133:14, 372.
- 18 Solkar MH, Akhtar NM, Khan Z, *et al*. Pseudomyxoma extraperitonei occurring 35 years after appendectomy: a case report and review of literature. *World J Surg Oncol* 2004;2:19.
- 19 Hirokawa F, Ono K, Hayashido M, *et al*. A case of retroperitoneal pseudomyxoma originated from mucinous adenocarcinoma of the ascending colon (in Japanese). *J W akayama Med Soc* 2004;55:154–8.
- 20 Kojima M, Takahashi N, Yazawa K, *et al*. A case of pseudomyxoma retroperitonei (in Japanese). *Surgery (in Japanese)* 2001;63:742–5.
- 21 Angelescu N, Bordea A, Popa E, *et al*. Pseudomixomul peritoneal (peritonita gelatinoasa). *Chirurgia (Bucur)*. 2001;96:443–51.
- 22 Liu ZL, Jiang YY. Pseudomyxoma retroperitonei due to the primary appendiceal mucinous adenocarcinoma (in Chinese). *World J Med Today* 2001;4:292–3.
- 23 Matsuoka Y, Masumoto T, Suzuki K, *et al*. Pseudomyxoma retroperitonei. *Eur Radiol* 1999;9:457–9.
- 24 Koizumi J, Noguchi H. Pseudomyxoma retroperitonei with spontaneous skin fistula. *Abdom Imaging* 1999;24:193–5.
- 25 Edrees WK, Hannon RJ. Pseudomyxoma extraperitonei: a rare presentation mimicking an iliacus abscess. *Ulster Med J* 1999;68:38–9.
- 26 Fujimura T, Yonemura Y, Fujita H, *et al*. Chemohyperthermic peritoneal perfusion for peritoneal dissemination in various intra-abdominal malignancies. *Int Surg* 1999;84:60–6.

- 27 Tsai CJ. Ultrasound features of disseminated adenomucinosis (pseudomyxoma). *Br J Radiol* 1998;71:564–6.
- 28 Hemet J, Lemoine F, Irimescu D, *et al.* Le pseudomyxome extraperitoneal. A propos de 3 observations. *Ann Pathol* 1998;18:137–40.
- 29 Oyama T, Kabayashi H, Minoda T, *et al.* Case report. Pseudomyxoma retroperitonei (in Japanese). *Nippon Igaku Hoshasen Gakkai Zasshi* 1997;57:620–1.
- 30 Stevens KJ, Dunn WK, Balfour T. Pseudomyxoma extraperitonei: a lethal complication of mucinous adenocarcinoma of the appendix. *Am J Gastroenterol* 1997;92:1920–2.
- 31 Mor Y, Leibovitch I, Garniek A, *et al.* Case report: pseudomyxoma extraperitonei: an unusual presentation mimicking a huge hydronephrotic kidney. *Clin Radiol* 1996;51:221–2.
- 32 Tamai O, Muto Y, Shiraishi H, *et al.* Mucinous cystadenocarcinoma of vermiform appendix involving retroperitoneum (in Japanese). *J Jpn Soc ColoProctol* 1995;48:496–501.
- 33 Nobusawa H, Hashimoto T, Munechika H, *et al.* CT findings of primary retroperitoneal cystic tumors: with special emphasis on the distinction between benignancy and malignancy (in Japanese). *Nippon Igaku Hoshasen Gakkai Zasshi* 1995;55:861–6.
- 34 Baba Y, Nakajo M, Kajiya Y, *et al.* A case of retroperitoneal pseudomyxoma. *Radiat Med* 1995;13:247–9.
- 35 Fann JJ, Vierra M, Fisher D, *et al.* Pseudomyxoma peritonei. *Surg Gynecol Obstet* 1993;177:441–7.
- 36 Koyama A, Ichikawa T, Honma M, *et al.* A case of retroperitoneal pseudomyxoma (in Japanese). *Jpn J Clin Radiol (Rinsho Hoshasen)* 1992;37:519–21.
- 37 Vladimirtseva AL, Chattalos MG, Poletaev VA. Pseudomiksoma retrotsekal'no raspolozhennogo cherveobraznogo otrostka s prorastaniem v zabriushinnoe prostranstvo i aruzhnyimi svishchami. *Arkh Patol* 1989;51:74–6.
- 38 Brady MB, Ewing RH, Robinson AE, *et al.* Flank mass and pain in a 72-year-old man. *Invest Radiol* 1986;21:419–23.
- 39 Shimabukuro Y, Miyagi T, Irabu T, *et al.* A case of giant pseudomyxoma in the retroperitoneum (in Japanese). *Okinawa Ikaishi* 1984;21:486–7.
- 40 Torgunakov AP, Pavliuk VD. Pseudomiksoma briushnoi polosti i zabriushinnogo prostranstva. *Vestn Khir Im I I Grek* 1975;114:139–40.
- 41 Kayser HW. Pseudomyxoma retroperitoneale. *Geburtshilfe Frauenheilkd* 1960;20:730–3.
- 42 Chetchueva TA. Zabriushinnaia psevdomiksoma cherveobraznogo otrostka. *Arkh Patol* 1957;19:64–6.
- 43 Chekharina EA. Pseudomiksoma zabriushinnogo prostranstva iz kisty cherveobraznogo otrostka. *Vopr Onkol* 1955;1:103–5.
- 44 Snyder TE, Vandivort MR. Mucinous cystadenocarcinoma of the appendix with pseudomyxoma peritonei presenting as total uterine prolapse: a case report. *J Reprod Med* 1992;37:103–6.
- 45 Radosavljevic G, Nedeljkovic B, Kacar V. Pseudomyxoma of the pleural and peritoneal cavities. *Thorax* 1993;48:94–5.
- 46 Ben-Hur H, Schachter M, Mashiah A, *et al.* Recurrent mucinous adenocarcinoma of the ovary presenting as an inguino-labial hernia. *Eur J Gynecol Oncol* 1996;17:299–302.
- 47 Baker WC, Goldman LB, deVere , *et al.* Pseudomyxoma peritonei presenting as a scrotal mass. *J Urol* 1988;139:821–3.
- 48 Peek DF, Beets GL. Pseudomyxoma peritonei in the pleural cavity; report of a case. *Dis Colon Rectum* 1999;42:113–15.
- 49 Koizumi J, Noiguchi H. Pseudomyxoma retroperitonei with spontaneous skin fistula. *Abdom Imaging* 1999;24:193–5.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow