

CASE REPORT

Primary retroperitoneal mucinous cystadenocarcinoma in a male patient

T P Thamboo, R Sim, S-Y Tan, W-M Yap

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Primary retroperitoneal mucinous cystadenocarcinomas (PRMCs) are rare. This is the first reported case in the literature in English of PRMC in a man. The 64-year-old man presented with a large retroperitoneal cystic tumour measuring 24×20×16 cm³, which was removed intact. Areas ranging from a benign mucinous cyst to borderline mucinous tumour to mucinous cystadenocarcinoma were observed on microscopy. Strong patchy staining for cytokeratins 7 and 20 and strong diffuse staining for MUC2 and MUC5AC core peptides, similar to staining patterns in ovarian mucinous tumours, were shown in the benign and atypical epithelium. Staining for CA19.9 and carcinoembryonic antigen was also shown by both components. The theory of its origin from the mucinous metaplasia of peritoneal (mesothelial) inclusion cysts, rather than from ectopic ovarian tissue or ovarian teratomas, is supported by the occurrence of such a tumour in a male patient.

Primary retroperitoneal mucinous cystadenocarcinomas (PRMC) are rare, with less than 20 reported cases in the literature in English, and all occurring in the female sex. We report the case of a PRMC in a male patient, the first such case to our knowledge. We describe the expression of cytokeratins and mucin core peptides, with a review of the literature.

CASE REPORT

A 64-year-old man presented with acute abdominal discomfort. Physical examination showed a palpable non-tender left flank mass. Both testes were descended and were clinically normal. A computed tomography scan showed a 19×15 cm² retroperitoneal cystic mass occupying the left side of the abdominal cavity (fig 1A). It was separate from the pancreas and left kidney, which were normal. Laparotomy showed a large cystic tumour attached to the left psoas muscle; it was excised intact. The patient recovered and had no evidence of recurrent disease 18 months after the operation.

Pathology

The specimen received was an intact cyst measuring 24×20×16 cm³, with a smooth external surface and containing yellow-brown gelatinous material. The cyst wall was thin, with several small firm projections on the inner surface measuring 0.1–0.5 cm in diameter.

When viewed microscopically, the cyst wall was mostly lined by cuboidal to columnar mucinous cells of the intestinal type. In most areas, the cells were single-layered, with no evidence of atypia. Occasional broad projections with fibrous cores covered by similar mucinous epithelium were seen. In foci, the cells were stratified (3–4 cells high), with mild to moderate atypia and mitotic activity. Stroma-free papillae

(fig 1B) and foci with signet-ring-type cells were also observed (fig 1C). The fibrous cyst wall showed the presence of spaces containing mucin and lined by atypical cells.

There was strong but patchy immunohistochemical staining for cytokeratin 7 (CK7) (fig 2A) and cytokeratin 20 (CK20) in the benign and atypical epithelium. Both components also showed diffuse strong staining for the MUC2 and MUC5AC gene products (figs 2B and C). No staining was identified for the MUC1 gene product. Moderate staining for CA19.9 was evident in the benign epithelium, with slightly weaker staining in the atypical epithelium. Weak apical membranous staining for carcinoembryonic antigen was found in the epithelial cells of the benign and atypical components. (The antibodies were from Dako, Carpinteria, California, USA (CK7, CK20), Novocastra, Newcastle-upon-Tyne, UK (MUC1, MUC2, MUC5AC, CA19.9) and Neomarkers, Fremont, California, USA (carcinoembryonic antigen). The dilutions used were 1:1000 for CK7, 1:700 for CK20, 1:200 for MUC1, 1:100 for MUC2 and MUC5AC, 1:400 for CA19.9 and 1:800 for carcinoembryonic antigen. An avidin–biotin complex method (IVIEW Detection System, Ventana, Arizona, USA) or a polymer-bound enzyme–antibody system (Envision ChemMate Detection System, Dako, USA) was used for detection. Appropriate positive and negative controls were used)

The final diagnosis was PRMC.

DISCUSSION

This is the first case of a PRMC in a male subject reported in the literature in English. Less than 20 cases of PRMC have been reported, all of which occurred in the female sex.^{1–8} A smaller number of benign and borderline mucinous tumours have been reported. The reported PRMCs have ranged in size from 5 to 24 cm in diameter and have tended to be solitary and unilocular, with smooth internal surfaces showing occasional small papillary projections, similar to the present case. Histological examination of the epithelium encountered in these tumours has been largely described using the terminology applied to ovarian mucinous tumours.

Only a few reports have studied the immunohistochemical profile of primary retroperitoneal mucinous tumours. Motoyama *et al*⁵ found that the benign and borderline mucinous epithelium in their two cases showed apical membranous staining for carcinoembryonic antigen, whereas the areas of mucinous cystadenocarcinoma showed more extensive, cytoplasmic staining with this marker. Tenti *et al*,⁹ on studying two cases of PRMC, found that positive staining for intestinal cell markers (M3SI, CAR-5) was present mainly in the malignant areas. Expression of markers of gastric differentiation (M1, cathepsin E, concanavalin A and pepsinogen II), however, was found mostly in the endocervical-type benign and borderline areas. These staining

Abbreviation: PRMC, primary retroperitoneal mucinous cystadenocarcinoma

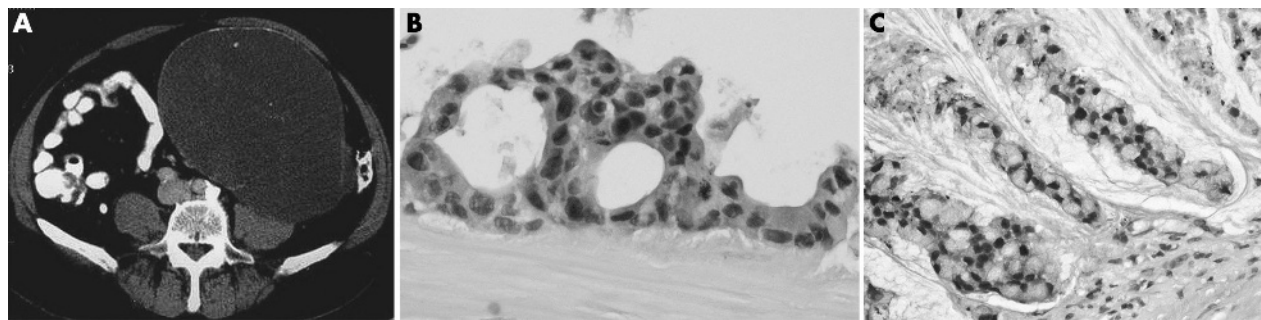


Figure 1 (A) Computed tomography image showing a large left retroperitoneal cyst adjacent to, but separate from, the left kidney. (B) Areas of the cyst wall lined by atypical mucinous epithelium showing stratification and stroma-free papillae (haematoxylin and eosin (H&E), original magnification $\times 400$). (C) Other areas of the cyst wall with signet-ring-type cells (H&E, original magnification $\times 200$).

patterns were similar to those found in ovarian mucinous tumours. The staining pattern for both CK7 and CK20 in the present case is also similar to the pattern seen in ovarian mucinous tumours of the intestinal type.¹⁰

The pattern of expression of the mucin peptide core antigens in this case is interesting, with strong diffuse expression of both gastric-type (MUC5AC) and intestinal-type (MUC2) antigens. This is consistent with the expression of the MUC5AC and MUC2 genes in ovarian mucinous tumours.^{10–13} MUC5AC gene expression has been found in 98–100% of primary ovarian mucinous tumours by immunohistochemistry,^{10–11} whereas MUC2 gene expression has been noted in 70–100% of ovarian mucinous carcinomas.^{11–12} Lower levels of MUC2 expression were observed in borderline and benign ovarian mucinous tumours.^{11–13}

The expression of CA19.9 seen in the present case was also noted in another case of PRMC in a Japanese case report,¹⁴ and the expression of the pancreatic marker DU-PAN-2 was noted in two cases by Tenti *et al.*⁹ The significance of the expression of these pancreatic markers is not clear.

The most popular theory of the pathogenesis of PRMC is that invaginations of the peritoneum (mesothelium) result in inclusion cysts that undergo mucinous metaplasia, thus resulting in a mucinous cystadenoma.¹⁵ These may then progress on to borderline and malignant mucinous tumours. Other main theories include an origin from ectopic ovarian tissue and an origin from an ovarian teratoma.⁸ Despite the similarities in morphological and immunohistochemical profiles of ovarian mucinous tumours and primary retroperitoneal mucinous tumours, the two theories of an origin from ectopic ovarian tissue and an origin from an ovarian

teratoma, however, cannot account for a tumour arising in a male patient, as in this case. A possible source of a retroperitoneal mucinous tumour in the male sex could be an undescended testis, which was excluded in this case.

The differential diagnoses in PRMC include metastatic mucinous tumours from sites such as the ovaries, the intestines (including the appendix) and the pancreas, and these have to be excluded by careful history-taking, clinical examination and diagnostic imaging.

In most cases of PRMC, the prognosis seems to be good. Most case reports indicate that the patients remain free of recurrence or metastases on short-term follow-up (3 months to 3 years). Experience with PRMC, especially in men, however, remains limited because of the rarity of these tumours and their biological behaviour will be better defined only when more cases are encountered.

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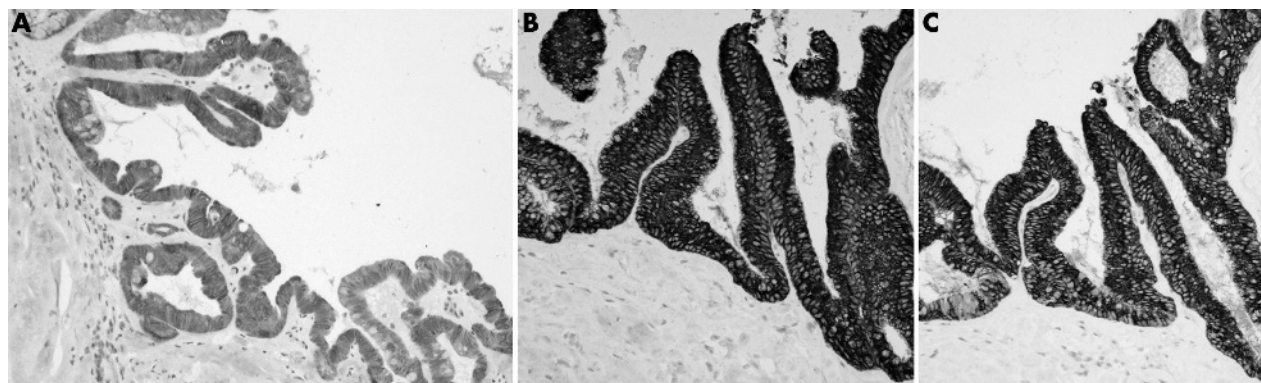


Figure 2 (A) Atypical mucinous epithelium with patchy cytoplasmic staining for CK7 (CK7 immunostain, original magnification $\times 40$). Atypical mucinous epithelium with diffuse strong staining for the MUC2 (B) and MUC5AC (C) gene products (MUC2 and MUC5AC immunostains, original magnification $\times 100$).

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