Pseudomyxoma peritonei originating from urachus—case report and review of the literature

A.K. Agraval MD PhD,* P. Bobiński MD,†
Z. Grzebieniak MD PhD,* J. Rudnicki MD PhD,‡
G. Marek MD PhD,* P. Kobielak MD,* M. Kazanowski MD,*
S. Agrawal,‡ and A. Halon MD PhD§

ABSTRACT

Pseudomyxoma peritonei (PMP) is a rare clinical condition defined as extensive intraperitoneal spread of mucus associated with a variety of mucinous tumours of varying biologic behavior. Although appendix or ovaries have usually been implicated as the primary site, cases have been reported in association with neoplastic lesions of other sites. Pseudomyxoma peritonei originating from urachal remnants is a unique entity, reported only 18 times in the English literature thus far. Considering the rarity of the lesion, we report the case of a 50-year-old man surgically treated for PMP associated with a low-grade mucinous urachal neoplasm. Unique aspects of case are the low histologic aggressiveness of the causative lesion (reported only twice worldwide) and the early stage of the disease, with a relatively small amount of intraperitoneal free mucin. Review of the literature about PMP in general and a collation of previously reported cases of PMP originating from the urachus are presented and discussed.

KEY WORDS

Pseudomyxoma peritonei, urachus, mucinous neoplasms, urachal adenocarcinoma

1. INTRODUCTION

Pseudomyxoma peritonei (PMP) is a rare clinical condition defined as extensive intraperitoneal spread of mucus associated with a variety of mucinous tumours of varying biologic behavior. The incidence of PMP is about 1–2 per million population. Low incidence forecloses the possibility of large-scale studies, and as a result there is broad diversity in the definitions, pathology, site-of-origin theories, treatment protocols, and prognosis for this disease.

The clinical entity of PMP embraces a wide spectrum of neoplasms from the benign and borderline to malignant lesions with invasive growth capacity.

Ronnett and colleagues introduced 3 diagnostic categories within this heterogeneous group: disseminated peritoneal adenomucinosis (DPAM), including histologically benign peritoneal lesions; peritoneal mucinous carcinomatosis (PMCA), with pathologic features of adenocarcinoma; and disease with features intermediate between DPAM and PMCA. Distinctive for PMP is a slow but relentless accumulation of abundant gelatinous fluid within the peritoneal cavity. Within this mucinous material, neoplastic cells may be bland and very scanty.

Although the appendix has been implicated as the primary site in most cases, incidences have been reported in association with neoplastic lesions of other sites, including ovary, fallopian tube, colorectum, colorectal tissue dislocated after previous surgical interventions, small bowel, stomach, gallbladder, lung, breast, pancreas, mucinous cysts of spleen, and extraperitoneal tumours. The primary site of invasion can also remain unknown. In unusual cases, the urinary tract can also be the primary source of PMP. To our knowledge, PMP originating from urachal remnants, as in the case we report here, has been reported only 18 times in the English literature thus far.

Clinically, PMP is a slowly progressive disease. Primary cystic tumour gradually grows, filled with a large amount of gelatinous fluid. After wall rupture because of intraluminal pressure, mucus-containing neoplastic cells leak and spread through the abdominal cavity, transported passively by physiologic peritoneal fluid flow. The tumour implants are usually distributed at sites of peritoneal fluid absorption (omentum, subphrenic space, Treitz ligament) and gravity sinks (pelvis, left abdominal gutter). Continuous peristaltic movement of intact small bowel usually protects the intestine wall from neoplastic invasion. Nevertheless, tumour cells can adhere to surfaces damaged during previous operations and to ovaries. This pattern of tumour dissemination, distinctive for PMP, is called “the redistribution phenomenon.”
During the prolonged clinical course of the disease, massive gelatinous ascites causes compression of the intra-abdominal organs, mechanical and functional gastrointestinal obstructions, and dyspnea leading to cachexia and, potentially, death. Pseudomyxoma peritonei often relapses, each time after a shorter interval, and local tumour recurrence can be expected in 60%–91% of the operated population (depending on the report), with a median disease-free interval of only 24 months, often leading to multiple operations.

Repeated intervention is an important aspect of PMP management. Interventions are associated with an increasing rate of mortality (ranging from 0% to 18%), perioperative morbidity (major complications requiring re-operation or intensive care, or causing chronic disability, are reported in 27%–70% of cases), and long-term treatment complications, of which aggregating adhesive disease is central to the debilitated condition of patients.

To summarize, although tumour growth usually tends to remain confined to the abdomen for many years, and blood-borne and lymph node metastases are uncommon, the disease has the potential for aggressive clinical behavior, including local tumour recurrence, and is always lethal unless radically treated.

2. CASE SUMMARY

A 50-year-old white man with no significant past medical history was admitted to the Second Department of General and Oncological Surgery of Wrocław Medical University with a tumour of the lower abdomen. The patient reported a 4-month history of intermittent lower abdominal pain radiating to the back. He denied other ailments, recent weight changes, or abdominal distention.

On admission, the patient’s general condition was good. His physical exam was notable only for a palpable tender mass in the central lower abdomen. Laboratory tests revealed mild monocytosis, with 11.4% white blood cells, and an elevated level of fibrinogen. Tumour marker studies were insignificant (carcinoembryonic antigen 0.5 ng/mL, and carbohydrate antigen 19-9 3.72 U/mL). All other blood and urine analyses, and chest radiography completed before surgery, showed no abnormalities.

Ultrasoundography revealed a unilocular, solid, heterogenous, hypoechoic, well-circumscribed tumour with fibrous partitions in the central lower abdomen, measuring 42×44×78 mm and extending from the anterosuperior dome of the urinary bladder to the anterior abdominal wall. Computed tomography imaging demonstrated the presence in the central lower abdomen of a heterogenous, hypodense, irregular polycystic mass with minute peripheral calcifications and without contrast enhancement, compressing and modelling intestine (Figure 1); mesenteric cyst was a primary consideration in the differential diagnosis. Evaluation for distant metastases was negative.

The patient subsequently underwent laparotomy. Revision of the abdominal cavity disclosed a polycystic mass emanating from the urachal remnant and mucinous material confined to the rectovesical excavation (Figure 2). The appendix was explored and considered to be normal-appearing. Evaluation for distant metastases and lymph node involvement was negative. The tumour was entirely removed within macroscopically healthy margins, including the extended aspect of parietal peritoneum and umbilical ligament. Partial cystectomy was performed. Approximately 150 mL of thick translucent yellow mucinous material was collected from the rectovesical excavation, and thorough lavage was performed. This procedure resulted in a macroscopically complete cytoreduction [Sugarbaker completeness of cytoreduction (cc) score 43 of 0].

The postoperative course was uneventful. At the time of article submission, our patient had been referred to long-term follow-up, but considering the low histologic aggressiveness (low-grade mucinous neoplasm, disseminated peritoneal adenomucinosis), the macroscopically complete cytoreduction, and the microscopically negative margins, no adjuvant chemotherapy was administered.

2.1 Pathology Findings

Macroscopic examination of the specimen revealed a well-circumscribed, gelatinous, polycystic mass emanating from the urachal remnant and measuring 8×7×4.5 cm, with abundant mucin within the lumen (Figure 3). In cross-section, the tumour was composed of smooth-walled cysts containing mucin.
pools. The mass did not include any papillary or solid lesions and did not communicate with the urinary bladder lumen.

Histologically, the cysts were lined by a tall columnar epithelium, with slight cellular crowding and stratification. The epithelium showed features of simple benign columnar epithelium without areas of dysplasia. Cytologically, the lining columnar cells had abundant mucin in the cytoplasm and showed a slight degree of loss of polarity, but no pleomorphism or mitotic figures were present. No stromal invasion of the tumour cells was observed, although mucin was extravasated into the cyst wall. The association of this tumour with the urachal remnant and its location just anterior to the dome of the urinary bladder confirmed the primary urachal origin. The mucinous material collected from the rectovesical excavation was extensively histologically sampled, but no malignant cells were detected in the specimens.

Histopathologic interpretation was consistent with PMP potentially originating from the urachal remnant (World Health Organization classification: low-grade mucinous neoplasm, disseminated peritoneal adenomucinosis). The specimen margins were free of infiltration. No malignant penetration or presence of mucus within the urinary bladder wall was detected. Figure 4 presents selected images from the pathology evaluation.

3. DISCUSSION

The urachus is a tubular structure extending medially from the apex of the bladder to the allantoid during fetal development; normally obliterating after birth, it converts into the median umbilical ligament. Its lumen is lined with a transitional epithelium that has the potential for focal glandular metaplasia, possibly followed by malignant transformation. Vestigial persistence of this duct is revealed in 30%–70% of autopsies. Urachal...
remnants can cause diverse abnormalities, including congenital anomalies (patent urachus, umbilical–urachal sinus, vesicourachal diverticulum, urachal cyst) and acquired diseases (infection, neoplasia). Tumours are uncommon and may be benign (adenoma, fibroma, fibroadenoma, fibromyoma, hamartoma) or malignant (adenocarcinoma in 90% of cases, usually with intestinal-type histology)\textsuperscript{41}. Urachal carcinoma accounts for 0.01% of all malignancies\textsuperscript{27,38} and 0.17%–1% of all bladder cancers\textsuperscript{27,38,41,44}.

Pseudomyxoma peritonei originating from mucinous neoplasm of the urachus is an extremely rare condition. We identified 18 previous cases in the English literature. Table 1 presents their distinctive features.

The collected cases present a variety of clinical pictures, stages at time of surgery, histologic types, and outcomes within a single clinical entity. The low number of reported cases and the differences in management and reported follow-up do not permit any generalizations or statistical assessments. However, we agree with the authors of earlier reports, that PMP originating from the urachus probably shares the pathophysiology, clinical behaviour, and prognosis of PMP with an appendiceal origin. Because peritoneal spread and local invasion dominate the clinical picture and present a major therapeutic problem, PMP originating in the urachus requires the treatment approach recommended for PMP arising from appendix rather than that for urachal adenocarcinoma\textsuperscript{2,34,35,41}.

Reported symptoms (Table 1) are diverse, develop slowly, and usually remain unspecific: lower abdominal pain, distention, palpable mass, weight loss, intermittent bowel obstruction, hematuria, and urinary retention. Mucinuria because of persistent connection between the urachal remnant and the urinary bladder is highly distinctive for this disease\textsuperscript{35}, but occurs relatively rarely. Physical examination (abdominal distention with so-called “jelly belly” ascites, and less often fever and peritoneal irritation) and laboratory tests (leukocytosis, anemia) are also not discriminative\textsuperscript{33}. In an important proportion of cases (42%), elevated serum carcinoembryonic antigen and carbohydrate antigen 19-9 are present. Both markers are not disease-specific, but remain useful and recommended for primal diagnosis, evaluation of cytoreduction, and postoperative follow-up\textsuperscript{28,35,45}.

Ultrasonography and computed tomography or magnetic resonance imaging, followed by histopathologic verification of extensively sampled tumour, are the preferred way to establish a diagnosis of PMP\textsuperscript{37,46}. Ultrasonographic findings supporting a diagnosis of PMP of urachal origin are a large collection of intraperitoneal fluid and a midline-situated cystic tumour attached to the dome of the urinary bladder, often of heterogeneous echogenicity, with multi-septation and intramural or septal calcification. Computed tomography and magnetic resonance images (especially T2-weighted sagittal images) of linear structure continuing from the tumour toward the umbilicus may complement other data.

Scallopings of the visceral surfaces and intestinal loop displacement evidence a large quantity of ascites and is discriminative with PMP\textsuperscript{31,35,40}. Cytology examination of aspirated mucus has little diagnostic value, because acellular mucin often spreads further than do malignant cells\textsuperscript{1}. Transurethral biopsies containing limited diagnostic material can provide false results because the tumour is often of heterogeneous architecture with focal invasive malignancy\textsuperscript{47}.

The described lack of pathognomonic symptoms in combination with the slow and occult clinical course of this tumour usually results in disease being locally advanced at the time of diagnosis\textsuperscript{34,41}.

To the best of our knowledge, no generally accepted staging system has been applied in PMP. Among the 18 previously described cases of urachus-origin PMP (Table 1), most were at an advanced stage at clinical presentation. Six reports (including that of the present case) identified the presence of a solitary cystic tumour with various amounts of mucinous material containing sparse neoplastic cells, but without invasion into the urinary bladder. In 3 cases, the bladder wall was additionally infiltrated. A single synchronous extra-urachal locus of invasion was reported once. Multiple intra-abdominal tumour implants or peritoneal carcinomatosis was already present in 9 cases at the time of diagnosis.

In contrast to the natural history of urachal adenocarcinoma, PMP hardly ever gives rise to lymphatic and blood-borne distant metastases. However, it does show a strong tendency for local recurrence\textsuperscript{2,33,41}. Consequently, the stage of local invasion throughout the peritoneal cavity is the first major prognostic factor\textsuperscript{48}.

The second independent prognostic factor is the histologic grade of the causative malignant lesion\textsuperscript{2,7,35,41,43,49}. According to data published by Ronnett and colleagues\textsuperscript{7}, the 5-year survival rate in patients with PMP of appendiceal origin is 84% for patients with DPAM, 37.6% for patients whose lesions have intermediate features, and only 6.7% for patients with PmCA. Sugarbaker\textsuperscript{43}, in presenting treatment results for 385 patients with PMP, reported 5-year survival rates after complete cytoreduction of 86% for patients with DPAM and 50% for patients with lesions having intermediate histopathologic features. Miner and colleagues\textsuperscript{42}, referring to surgical treatment in 97 patients, observed that 90% of 10-year survivors had low-grade pathologic features. Table 1 shows great histologic variation in patients with PMP of urachal origin, from mucinous urachal tumour of low malignant potential and mucinous borderline tumour of low malignant potential; to well-, moderately-, and poorly-differentiated urachal adenocarcinoma. According to the classification system proposed by Ronnett and colleagues\textsuperscript{7}, 3 of those cases (including the present case) represent benign disseminated peritoneal adenomucinosis (DPAM), 5 represent intermediate disease, and 11 demonstrate histologically aggressive PmCA.
<table>
<thead>
<tr>
<th>Reference</th>
<th>Sex (M/F)</th>
<th>Age (years)</th>
<th>Symptoms</th>
<th>CEA (ng/mL)</th>
<th>CA19-9 (U/mL)</th>
<th>Findings</th>
<th>Surgical procedure</th>
<th>Chemotherapy</th>
<th>Follow-up (months)</th>
<th>Outcome</th>
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<tr>
<td>Faulkner et al., 1954</td>
<td>M</td>
<td>65</td>
<td>Abdominal pain, weight loss</td>
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<td>49</td>
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<td>Loggie et al., 1997</td>
<td>M</td>
<td>35</td>
<td>Gross painless hematuria</td>
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<td>Preoperative systemic cyclophosphamide, doxorubicin, cisplatin, HIPEC with mitomycin C and leucovorin;</td>
<td>22, 31</td>
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<td>M</td>
<td>45</td>
<td>Abdominal distension</td>
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<td>M</td>
<td>34</td>
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<td>Extensive peritoneectomy, sigmoidectomy, small-bowel segmental resection, omentectomy, partial cystectomy</td>
<td>Preoperative systemic with 5FU and leucovorin; HIPEC with mitomycin C</td>
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<td>Stenhouse et al., 2003</td>
<td>M</td>
<td>54</td>
<td>Abdominal pain, rectal bleeding</td>
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<td>6</td>
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<td>Symptoms</td>
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<td>CA19-9 (U/mL)</td>
<td>Findings</td>
<td>Surgical procedure</td>
<td>Chemotherapy</td>
<td>Follow-up</td>
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<td>M</td>
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<td>Left inguinal hernia</td>
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<td>Suprapubic ruptured cyst, 9×7×5.5 cm</td>
<td>Mucinous urachal cystic tumour of low malignant potential</td>
<td>Tumourectomy, extensive peritoneectomy, partial cystectomy, appendectomy</td>
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<td>M</td>
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<td>Macroscopic hematuria</td>
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<td>Mucinous urachal adenocarcinoma</td>
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<td></td>
<td>M</td>
<td>47</td>
<td>Mucinuria, abdominal pain and distention</td>
<td>14</td>
<td>594</td>
<td>Multiple intra-abdominal tumours</td>
<td>Tumourectomy, extensive peritoneectomy, right hemicolecctomy, rectosigmoid colectomy, omentectomy, appendectomy</td>
<td>HIPEC with mitomycin C and doxorubicin;</td>
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<td>Free of disease</td>
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<td>Age</td>
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<td>CA19-9 (U/mL)</td>
<td>Findings</td>
<td>Surgical procedure</td>
<td>Chemotherapy</td>
<td>Follow-up (months)</td>
<td>Outcome</td>
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<td>Yan et al., 2006</td>
<td>F</td>
<td>34</td>
<td>Infertility</td>
<td></td>
<td></td>
<td>Intraoperative</td>
<td>Early postoperative intraperitoneal 5FU; adjuvant systemic 5FU</td>
<td></td>
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<td>Sugarbaker et al., 2008</td>
<td>F</td>
<td>32</td>
<td>Infertility</td>
<td></td>
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<td>Intraoperative</td>
<td>Early postoperative intraperitoneal 5FU, leucovorin, 5FU</td>
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<td>Recurrence, died of disease</td>
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<td>Khalid et al., 2008</td>
<td>F</td>
<td>14</td>
<td>Weight loss, recurrent subacute</td>
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<td>Tumourectomy, extensive peritoneotomy, rectosigmoid colectomy, omentectomy, splenectomy</td>
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<td>Sugiyama and Ito, 2009</td>
<td>M</td>
<td>58</td>
<td>Abdominal fullness</td>
<td>41.5</td>
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<td>Suprapubic cyst, 11×10×10 cm; multiple intra-abdominal implants</td>
<td>Tumourectomy, extensive debulking of diffuse lesions, omentectomy</td>
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<td>Free of disease</td>
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<td>Lamb et al., 2010</td>
<td>M</td>
<td>63</td>
<td>Chronic urinary retention</td>
<td>Not stated</td>
<td></td>
<td>Not stated</td>
<td>Cystic mass in dome of bladder, omental metastases</td>
<td>Tumourectomy, debulking with limited peritoneotomy, omentectomy, partial cystectomy</td>
<td>No</td>
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</tr>
<tr>
<td>Reference</td>
<td>Sex (M/F)</td>
<td>Age (years)</td>
<td>Symptoms</td>
<td>CEA (ng/mL)</td>
<td>CA19-9 (U/mL)</td>
<td>Findings</td>
<td>Surgical procedure</td>
<td>Chemotherapy</td>
<td>Follow-up (months)</td>
<td>Outcome</td>
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<td>Nozaki et al., 2011</td>
<td>M</td>
<td>37</td>
<td>Abdominal pain</td>
<td>Not stated</td>
<td></td>
<td>Cystic tumour beneath umbilicus</td>
<td>Tumourectomy, extensive peritonectomy</td>
<td>No</td>
<td>54</td>
<td>Free of disease</td>
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<tr>
<td>Kebapçı et al., 2012</td>
<td>M</td>
<td>59</td>
<td>Abdominal turgidity and discomfort</td>
<td>Not stated</td>
<td></td>
<td>Suprapubic ruptured cyst, 10×8×5 cm</td>
<td>Tumourectomy, partial omentectomy</td>
<td>After recurrence: systemic 5FU and mitomycin C</td>
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<td>Recurrence, died of disease</td>
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<td>Martinez et al., 2012</td>
<td>M</td>
<td>32</td>
<td>Mucinuria</td>
<td>Not stated</td>
<td></td>
<td>Multiple intra-abdominal implants, residual after previous operation</td>
<td>Extensive peritonectomy, tumour debulking, right hemicolectomy, omentectomy, partial cystectomy, appendectomy</td>
<td>HIPEC with oxaliplatin</td>
<td>24</td>
<td>Free of disease</td>
</tr>
<tr>
<td>Agrawal et al. (present case)</td>
<td>M</td>
<td>50</td>
<td>Abdominal pain</td>
<td>0.5</td>
<td>3.72</td>
<td>Suprapubic polycystic tumour, 7.8×4.4×4 cm</td>
<td>Tumourectomy, partial cystectomy, extended parietal peritonectomy</td>
<td>No</td>
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</table>

M/F = male or female; CEA = carcinoembryonic antigen; CA19-9 = carbohydrate antigen 19-9; HIPEC = hyperthermic intraperitoneal chemotherapy; 5FU = 5-flourouracil.
The third main prognostic factor is completeness of cytoreduction\textsuperscript{42,43,48}. Table II presents the \textit{cc} score published by Sugarbaker\textsuperscript{43}. In PMP, the aim of surgical procedures is to excise all visible tumour, or if full excision is not achievable, to leave residual nodules less than 0.25 cm. The 0.25 cm diameter is considered the maximal achievable penetration distance for locally applied chemotherapy\textsuperscript{41}.

Sugarbaker\textsuperscript{43} describes 5-year survival rates of only 20\% and no patient surviving 10 years after incomplete cytoreduction (\textit{cc}-2 and \textit{cc}-3). No significant differences in survival were observed for patients with \textit{cc}-2 and \textit{cc}-3 cytoreductions. Miner and colleagues\textsuperscript{42} observed median survivals of 12.8 and 4.2 years after complete cytoreduction and incomplete cytoreduction respectively.

Abdominal distension, weight loss, invasion of major organs\textsuperscript{1}, and (if prior surgery has been performed) a large extent of prior surgical intervention\textsuperscript{43} have also been assumed to be poor prognostic factors.

The optimal treatment for patients with PMP remains controversial. As mentioned earlier, peritoneal spread and local invasiveness are essential and defining for PMP, and surgical management (radical or partial cystectomy with \textit{en bloc} resection of tumour, urachal remnant, umbilicus, and posterior rectus abdominis fascia) might be insufficient for urachal adenocarcinoma\textsuperscript{49}.

Most publications recommend, at least at the first operation, an aggressive approach resulting in complete cytoreduction (\textit{cc}-0 or \textit{cc}-1) with radical peritonectomy and resection of involved major organs, followed by perioperative intraperitoneal chemotherapy\textsuperscript{2,5,27,32–35,39,41,43}. That procedure has demonstrated prolonged survival at the cost of associated severe acute toxicity and elevated morbidity. Conversely, given that aggressive procedures reduce quality of life for patients and are usually not curative (high rates of recurrence are reported among long-term survivors), some authors prefer selective debulking. Miner and colleagues\textsuperscript{42} suggest, that in case of advanced intra-abdominal spread, an aggressive surgical procedure does not ensure completeness of cytoreduction. In that group of patients, the authors advise a treatment strategy based on symptom management and preservation of function.

In the treatment protocol for PMP established by the Basingstoke and North Hampshire Foundation Trust Hospital (British National Tertiary Pseudomyxoma Referral and Treatment Centre), the cytoreductive procedure must include radical peritonectomy and removal of involved viscera to extirpate all visible tumour implants. Palliative debulking is performed if complete cytoreductive surgery is not achievable. Additional appendectomy and revision of ovaries to exclude the two main origins of PMP are advised\textsuperscript{24}. Because radical cystectomy does not improve outcomes, a conservative partial cystectomy is usually recommended\textsuperscript{2,34,37}. Thorough lavage of the abdominal cavity with mucolytic agents (5\% dextrose in water or low molecular weight dextran solution) has been suggested to be beneficial\textsuperscript{32}.

The surgical procedure must routinely be followed by perioperative intraperitoneal chemotherapy\textsuperscript{2,5,27,32–35,39,41,43}. Most recent studies suggest an essential advantage for application of hyperthermic intraperitoneal chemotherapy\textsuperscript{24,34,35,43,48,50,51}. In cases of early-stage PMP with low malignant histology, some authors recommend not to dispense with intraperitoneal chemotherapy\textsuperscript{39}. Only a few authors have described some benefit with systemic chemotherapy and radiotherapy. Nevertheless, such treatments can be applicable and useful for palliation in fast-deteriorating patients with recurrent high-grade malignant disease\textsuperscript{2,3}.

<table>
<thead>
<tr>
<th>Score</th>
<th>Cytoreduction</th>
<th>Size of largest residual tumour nodule</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>\textit{En bloc} resection</td>
<td>Nodules not visible</td>
</tr>
<tr>
<td>1</td>
<td>Complete cytoreduction</td>
<td>Nodules &lt; 0.25 cm</td>
</tr>
<tr>
<td>2</td>
<td>Incomplete cytoreduction, moderate residual disease</td>
<td>Nodules 0.25–2.5 cm</td>
</tr>
<tr>
<td>3</td>
<td>Incomplete cytoreduction, gross residual disease</td>
<td>Nodules &gt; 2.5 cm</td>
</tr>
</tbody>
</table>

4. SUMMARY

Although increasing numbers of cases of PMP have been reported, this neoplasm remains a diagnostic challenge because of its rarity and lack of discriminative symptoms. Pseudomyxoma peritonei should be considered in the differential diagnosis for abdominal and retroperitoneal tumours, especially with intercurrent ascites, because misdiagnosis delays effective treatment and affects survival.

5. CONFLICT OF INTEREST DISCLOSURES

We certify that there is no financial conflict of interest with any organization concerning the material discussed in this manuscript.

6. REFERENCES


AGR AWA L et al.


**Correspondence to:** Anil Kumar Agrawal, 2nd Department of General and Oncological Surgery, Wroclaw Medical University, 213 Borowska Street, Wroclaw 50-556 Poland.  
**E-mail:** dranilpreeti@gmail.com

* Second Department of General and Oncological Surgery, Wroclaw Medical University, Wroclaw, Poland.  
† Department of General Surgery II, Lower-Silesian Specialised Hospital, Wroclaw, Poland.  
‡ Wroclaw Medical University, Wroclaw, Poland.  
§ Division of Pathomorphology and Oncological Cytology, Wroclaw Medical University, Wroclaw, Poland.