Pseudomyxoma Peritonei - What every radiologist should know

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Learning objectives

After reviewing this exhibit, the learners will:

• Understand the etiopathology of Pseudomyxoma Peritonei (PP)
• Recognize clinical and imaging findings associated with this disease, with emphasis on ultrasound (US) and computed tomography (CT)
• Comprehend the treatment options and prognosis of PP
Background

Definition

Pseudomyxoma peritonei (PP) is a rare condition characterized by the gradual accumulation of large volumes of mucinous ascites. (1)

The term PP was described for the first time by Cruveilhie in 1848 as "maladie gèlatineuse du péritoine", and since that time, this term is more a clinical or radiologic description instead of a disease.(1,2) In fact, the characteristic "Jelly Belly" is applied to describe copious, thick mucinous or gelatinous material on the surfaces of the peritoneal cavity. (1,2)

Epidemiology and Etiopathology

PP is a rare condition with an incidence of one case per million population per year reported on the literature. (3).

The mean age of patients at diagnosis is 49 years (range, 23-83 years), and more frequently affects women than men. (4).

The pathogenesis and classification of PP has been controversial along the years. According to past literature, the PP could be divided in two categories:

**Pseudomyxoma peritonei**

A form that contains benign or borderline-appearing epithelial cells or cells from well-differentiated (low-grade) mucinous carcinomas and was referred to by some as disseminated peritoneal adenomucinosis (1,2,5). This form of pseudomyxoma peritonei does not invade the stroma and appears to spread along the peritoneal surfaces, characteristics that make it amenable to surgical debulking. It tends to have a relatively indolent or protracted clinical course, especially if debulking is successful.(1,2,5)

**Peritoneal mucinous carcinomatosis**

Characterized by invasive, high-grade, moderately or poorly differentiated mucinous carcinoma with large extracellular pools of mucin. This second category originates from a mucinous carcinoma of the gastrointestinal tract, gallbladder, pancreas, or ovary, and its clinical course is fatal.(1,2)

Nevertheless according to more recent literature, it is now accepted that the majority of cases of classic pseudomyxoma peritonei develop from low-grade mucinous carcinomas that arise in the appendix and that penetrate or rupture into the peritoneal cavity (1,2,6,7).
The majority of epithelial tumors of the appendix are mucin rich, demonstrate circumferential mucosal involvement, and have a strong propensity to form mucoceles. (8)

Mucocele of the appendix occurs when obstruction of the appendiceal lumen results in mucus accumulation and consequent abnormal dilatation. (9)

The mucus production maybe a result from benign mucinous neoplasm (cystoadenoma) or malign mucinous neoplasm (cystoadenocarcinoma). (8,9) The rupture of a mucocele with malign mucinous neoplasm may lead to intraperitoneal spread od the cells causing PP. (8,9)

Nevertheless some literature posit that most cases of PP are appendiceal in origin and that associated ovarian lesions usually represent metastatic disease, this is controversial. (1,8,11)

Clinical Features

Patients typically complain of progressive abdominal pain, increasing abdominal girth, and weight loss. (1,2,10,11)

In some cases, patients complain of predominantly right-sided pain or they may have signs and symptoms similar to those of appendicitis when pseudomyxoma peritonei is located predominantly in the right lower quadrant. (1)

With progression of the disease, the intestine becomes encased in mucinous material and bowel obstruction eventually occurs. (1)

Pathologic Features

There are differences between pseudomyxoma peritoneu versus peritoneal mucinous carcinomatosis. The first manifests with gelatinous material covering the peritoneal surface and mucinous ascites. PP tends to spare the peritoneal surfaces of the bowel and to accumulate beneath the right hemidiaphragm, along the omental surfaces, in the gravity-dependent portions of the pelvis, right retrohepatic and subhepatic spaces, and left paracolic gutter. While the second, often has larger soft-tissue and fibrotic component and may be adherent to all peritoneal surfaces including the serosal surfaces of the intestine, pseudomyxoma peritonei. (13,14)

In low-grade mucinous tumors that produce classic pseudomyxoma peritonei, mucin pools may have few tumor cells, and, when present, the tumor cells can be cytologically bland rather than frankly malignant. Hyalinized collagenous tissue may be admixed with the mucin or ex- tend through the lobules of omentum. Benign-appearing cells with slight nuclear atypia or those of a low-grade or well-differentiated carcinoma may be found lining the mucin pools. (Fig.1) The cellular
areas may be very sparse or focally distributed. Scattered lymphocytes and histiocytes may be present. Invasion and infiltration through the underlying peritoneal surface is typically not present. (1,15)

**Treatment and Prognosis**

Treatment usually consists of surgical debulking with appendectomy, omentectomy. (8) Repeated surgical intervention to remove the accumulated mucinous material remains the treatment of choice and is the only therapy known to prolong survival. (2,10,11) Individual survival depends significantly on the type of underlying tumor. Patients with adenocarcinoma of the ovary or appendix have a worse prognosis than those with a benign neoplasm. (11,12). An overall 5-year survival rate of 40%-50% is suggested from the literature

The clinical course of diffuse pseudomyxoma peritonei is typically insidious and unrelenting, with a 5-year survival rate of 40%-65% from the literature. (11,16). Intraperitoneal chemotherapy may be of benefit to some patients (17).
Imaging findings OR Procedure details

PP unspecific clinical presentation explains the central role of imaging methods in the study of this so often misdiagnosed condition.

The initial investigation of these patients will usually involve ultrasound (US) or computed tomography (CT), so the recognition of pseudomyxoma peritonei imaging features on these techniques is essential for an early, preoperative diagnosis.

Radiography

This exam usually is not very useful in the diagnosis.

Nonetheless the radiographic findings are similar to those seen in patients with massive ascites. It may appear increased opacity throughout the abdomen, with poor definition of the intraabdominal organs and obliteration of the psoas margins. Focal collections of mucin in the right subhepatic space may obscure the inferior hepatic margin or contribute to medial displacement of the liver tip (Hellmer sign). The ascending and descending colon may be displaced centrally and the pararectal fat stripe laterally when large amounts of mucin are in the paracolic gutters. In the pelvis, mucin in the paravesical spaces will produce symmetric areas of opacity on either side of the bladder.

Ultrasound

US commonly the first evaluation requested for abdominal pain. Therefore is important to recognize and evaluate such features as:

- Evaluate with US abdominal areas of stasis of fluid within the peritoneal cavity, once this will be the locals where mucinous deposits will accumulate. The most common locals are the pouch of Douglas in women or the retrovesical space in men, the right lower quadrant near the ileocecal junction, the right subhepatic space (Morison pouch), and the right subphrenic space. (Fig. 2)
- Echogenic ascites reflects the mucin and the gelatinous nature of the fluid. The echogenic foci inside the abdominal fluid are nonmobile in contrary to echoes in non-pure ascites. (Fig. 3)
- "Starburst" appearance of the bowel loops, because instead of floating freely, are displaced centrally and posteriorly by the surrounding mass. (19)
- The ascites may also have septa that are thought to represent the margins of the mucinous nodules. (Fig. 4)
- Scalloping of the liver and/or spleen margin by adjacent peritoneal masses are characteristic of PP.
• Echogenic masses, that are thought to be due to the presence of numerous tiny cysts, either focal or sheetlike, because of the involvement of the parietal peritoneum and omentum are also observed in these patients. (20)

**Computed Tomography**

CT is commonly the next exam that patients with PP are offered. When performing this evaluation it is important to be aware of the following features:

• Scalloping represents the indentations that occur on the capsular margins of the intraperitoneal organs such as liver and spleen, from the extrinsic pressure of the intraperitoneal mucinous implants (1). Scalloping sign is the most important diagnostic helping in the discrimination of PP from simple ascites. (Fig. 5)

• The mucinous ascites has usually a low attenuation, however areas of soft-tissue attenuation may be present and represent solid tumor elements, fibrosis, or compression of the mesentery (1,20,22).

• With disease progression the gelatinous ascites advances and incarcerates the abdominal organs. (Fig. 1,20,22)

• Severe abdominal ascites maybe accompanied with pleural effusion. Fig (10)

• Amorphous, curvilinear, faint calcifications may be present (1)

• All CT scans should evaluate appendiceal region searching for neoplasm or mucocelo. Also the ovaries should be evaluated.

**Differential Diagnosis**

Differentiating the classical pseudomyxoma peritonei from mucinous carcinomatosis is difficult because their CT findings overlap despite their different pathologic features and clinical courses.

However there are some different characteristics to retain that are schematized on table 1.

<table>
<thead>
<tr>
<th>Pseudomyxoma Peritonei</th>
<th>Peritoneal Mucinous Carcinomatosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Origin</strong></td>
<td>Appendiceal mucinous neoplasm ; Ovaries?</td>
</tr>
<tr>
<td><strong>Pathology</strong></td>
<td>Epithelial cells or cells from well-differentiated</td>
</tr>
<tr>
<td><strong>US</strong></td>
<td><strong>CT</strong></td>
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<td>-----------------</td>
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<tr>
<td>Echogenic foci inside the abdominal fluid are nonmobile</td>
<td>Scallopning of the liver and/or spleen margin</td>
</tr>
<tr>
<td>Echogenic foci inside the abdominal fluid are mobile</td>
<td>No intra-abdominal organ scalloping</td>
</tr>
<tr>
<td>No intra-abdominal organ scalloping</td>
<td>Tends to involve the chest more frequently with effusions or pleural masses and may also be accompanied by mesenteric or retroperitoneal lymphadenopathy, omental caking, and invasion into parenchymal organs (23)</td>
</tr>
</tbody>
</table>

*(low-grade) mucinous carcinoma with large extracellular pools of mucin*
Conclusion

During the last decade, there has been an effort to understand and differentiate PP from mucinous carcinomatosis, and to comprehend the etiopathology of the classical PP.

Indeed the distinction between these two conditions is important clinically because patients with pseudomyxoma peritonei from an appendiceal low-grade mucinous neoplasm have a 5-year survival rate of 50% and those who have mucinous carcinomatosis have a 5-year survival rate of less than 10%.(1)

Consequently, regarding Pseudomyxoma peritonei is an unspecific diagnosis, and knowing that many patients will undergo US or CT as an initial investigation, it's important to recognize its radiological features so that the diagnosis can be made in a fast, easy and preoperative way.
Fig. 1: Photomicrograph (original magnification, x10, HE stain) neoplastic adenocarcinoma cells (mucin production). The cells with mucinous cytoplasm are the source of the extracellular mucin.

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Fig. 2: Echogenic and heterogeneous mucinous deposits of pseudomyxoma peritonei that accumulate in areas of stasis in the abdomino-pelvic cavity.

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Fig. 3: Gelatinous fluid, non-pure with echogenic deposits of mucine and echoes.

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**Fig. 4:** Non-pure echogenic ascites with septa.

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Fig. 5: Mucinous ascites (low CT attenuation) and septas. Liver and spleen scalloping.

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Fig. 9: Coronal CT multiplanar imaging; Large volume of mucinous ascites, with encasement of the abdominal organs and slight liver scalloping. Notice the small bowel loops with a "starbust" appearance.

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Fig. 8: The same male patient two years later; Low density mucinous ascites with septa, causing encasement of the bowel loops.

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Fig. 7: The same male patient six months later

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Fig. 6: Male patient with mucinous ascites at the time of the diagnosis

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Fig. 10: Pleural effusion and left lower lobe atelectasis associated with pseudomyxoma.

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