Demystifying appendiceal and ovarian pseudomyxoma

Clinical History:

Postmenopausal woman with unremarkable medical history apart from resection of benign ovarian cyst decades earlier. Currently suffering from malaise, weight loss and painless increased abdominal girth over 6 weeks despite diuretics prescribed by general practitioner. Mild anaemia without significant abnormalities of remaining routine laboratory tests.

Imaging Findings:

Preoperative CT (Fig.1, 2) panoramically showed presence of higher-than-water attenuation perihepatic, perisplenic and mesenterial effusion, and a huge, mostly hypoattenuating mass with fine enhancing septations, which occupied the entire pelvis surrounding the uterus, displaced the right colon cranially and extended above the umbilicus. Additionally, a well-demarcated ovoid structure measuring 3.5x2.5 cm was seen abutting the caecum, with internal hypoattenuation similar to the huge surrounding mass, and thin enhancing periphery similar to septations. Laparotomic surgery (postoperative status shown in Fig. 3) confirmed mucinous ascites, and required resection of a large (23x19x8 cm) mucoid mass encasing adnexa and dilated cystic-appearing appendix, and required right hemicolecotomy and hysteroadnexectomy. Histopathology diagnosed pseudomyxoma peritonei from low-grade (single row of enlarged cells without atypias) primary mucinous tumour of appendix, extending to the ovaries.

Discussion:

A rare (estimated incidence 1-2 cases/1.000.000 people/year) but insidious condition mostly encountered in females (4:1 predominance), pseudomyxoma peritonei (PMP) refers to the presence of either free or organised mucin (with or without neoplastic cells) in the peritoneal cavity. Patients are mostly 50 to 70 years old, often present with palpable mass and abdominal distension, and may suffer from bowel obstruction [1, 2]. The vast majority of PMP result from mucinous appendiceal tumours (MATs), which are categorised as either low-grade (78%) or frankly malignant (22%) according to mucin, cytological and architectural features. Being the commonest form of uncommon appendiceal tumours, MATs have a characteristic slowly progressive natural history with peritoneal (rather than lymphatic and haematogenous) spread, and highly variable clinical behaviour. Roughly 20% of MATs ultimately develop PMP, via intraluminal accumulation of mucin, mural perforation, spread and
circulation of mucin and cells into the peritoneal cavity, ultimately leading to formation of gelatinous implants on serosal surfaces and omentum. The rare (<10%) extra-appendiceal PMP result from ovarian, gallbladder, gastric and colorectal neoplasms [2-7].

Cross-sectional CT or MR imaging typically depict MTA as well-capsulated, peripherally enhancing tubular or cyst-like structures with smooth or thickened walls. The differential diagnosis is a simple mucocele (macroscopic description of grossly distended appendix) secondary to a non-neoplastic cause such as an appendicolith. In PMP, fluid- or proteinaceous attenuation ascites coexists with peritoneal nodules or bulky deposits in the omentum, pelvis, right hemidiaphragm and retrohepatic space, which may compress or scallop solid organs; the bowel serosal surface are typically spared. Additionally, the adnexa are involved in 65-70% of females: in these occurrences, the appendix is considered the primary site of disease. Therefore, when imaging suggests a cystic ovarian tumour with PMP, radiologists should search for a coexistent, usually unsuspected MTA [1, 4, 6, 8, 9].

Treatment of PMP requires right hemicolectomy and lymph node dissection, sometimes aggressive surgical debulking and hyperthermic intraperitoneal chemotherapy. The prognosis is worse for high-grade (5-year survival rates below 50%) compared to low-grade (62-100%) tumours [2, 4, 5].

**Differential Diagnosis List:** Low-grade appendiceal mucinous tumour with pseudomyxoma peritonei and adnexal involvement, Pseudomyxoma peritonei from primary ovarian mucinous tumour, Peritoneal carcinomatosis, Peritoneal lymphoma, Appendiceal mesenchymal tumour/carcinoid

**Final Diagnosis:** Low-grade appendiceal mucinous tumour with pseudomyxoma peritonei and adnexal involvement

**References:**


Description: Coronal (a) and axial (b...d in craniocaudal order) showed postoperative status after resection of pseudomyxoma peritonei, right hemicolecetomy and hysteroadnexectomy. Note disappearance of ascites. Origin: Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
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Description: Precontrast images showed higher-than-water nonhaemorrhagic (18-20 Hounsfield units) ascites (*), a huge hypoattenuating pelvic mass containing some septations (thin arrows), which extended cranially above the umbilicus. Note uterus (thick arrow). Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The huge hypoattenuating abdominopelvic mass contained some septations (thin arrows), extended cranially above the umbilicus, and abutted the right colon (+). Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Additionally, a well-demarcated ovoid structure (arrows) measuring approx 3.5x2.5 cm showed thin peripheral rim isoattenuating with septa and internal hypoattenuation similar to the huge abdominopelvic mass. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** The well-demarcated ovoid structure (arrows) was located just postero-inferiorly to the caecum, within the huge abdominopelvic mass, and showed thin peripheral rim isoattenuating with septa and internal hypoattenuation. Note higher-than-water nonhaemorrhagic ascites (*). **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: After iv contrast, enhancing septations (thin arrows) were better perceptible within the huge hypoattenuating mass, which extended cranially above the umbilicus. Note uterus (thick arrow), higher-than-water nonhaemorrhagic (18-20 Hounsfield units) ascites (*). Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** After iv contrast, enhancing septations (thin arrows) were better perceptible within the huge hypotennuating mass. Note surrounded, displaced right colon (+) and higher-than-water nonhaemorrhagic (18-20 Hounsfield units) ascites (‘). **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
**Description:** After iv contrast, enhancing septations (thin arrows) were better perceptible within the huge hypoattenuating mass. Note uterus (thick arrow), higher-than-water nonhaemorrhagic (18-20 Hounsfield units) ascites (*). **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** Detail images confirmed well-demarcated ovoid structure (arrows) abutting the caecum(+) with internal hypoattenuation similar to huge surrounding mass, thin enhancing periphery similar to septations. **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
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