

## ORIGINAL ARTICLES

# Imaging of uncommon peritoneal diseases

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## Abstract

Among peritoneal diseases, a well recognized lesion is peritoneal carcinomatosis which is the most frequently found neoplastic disease that involves the peritoneal cavity. However, there are many uncommon peritoneal diseases including malignant peritoneal mesothelioma, primary sclerosing encapsulating peritonitis, intraperitoneal loose body, peritoneal echinococcosis, primary serous papillary carcinoma of peritoneum, extrapelvic endometriosis, ruptured ovarian teratoma with peritoneal dissemination and peritoneal inclusion cyst. Knowledge of those imaging findings is helpful in making diagnosis or narrowing differential diagnosis.

## Keywords

Uncommon peritoneal diseases, Peritoneal diseases, Imaging of peritoneum

## 1 Introduction

Many peritoneal lesions involve peritoneal lining but peritoneal carcinomatosis is frequently encountered condition. However primary neoplastic and nonneoplastic process though uncommon also arise from common site of origin and imaging findings often mimic each other and that of peritoneal carcinomatosis. This article summarizes the CT findings of uncommon peritoneal diseases including malignant peritoneal mesothelioma, primary sclerosing encapsulating peritonitis, intraperitoneal loose body, peritoneal echinococcosis, primary serous papillary carcinoma of peritoneum, extrapelvic endometriosis, ruptured ovarian teratoma with peritoneal dissemination, and peritoneal inclusion cyst. Familiarity of those uncommon peritoneal diseases is paramount important to make a diagnosis or appropriate differential diagnosis.

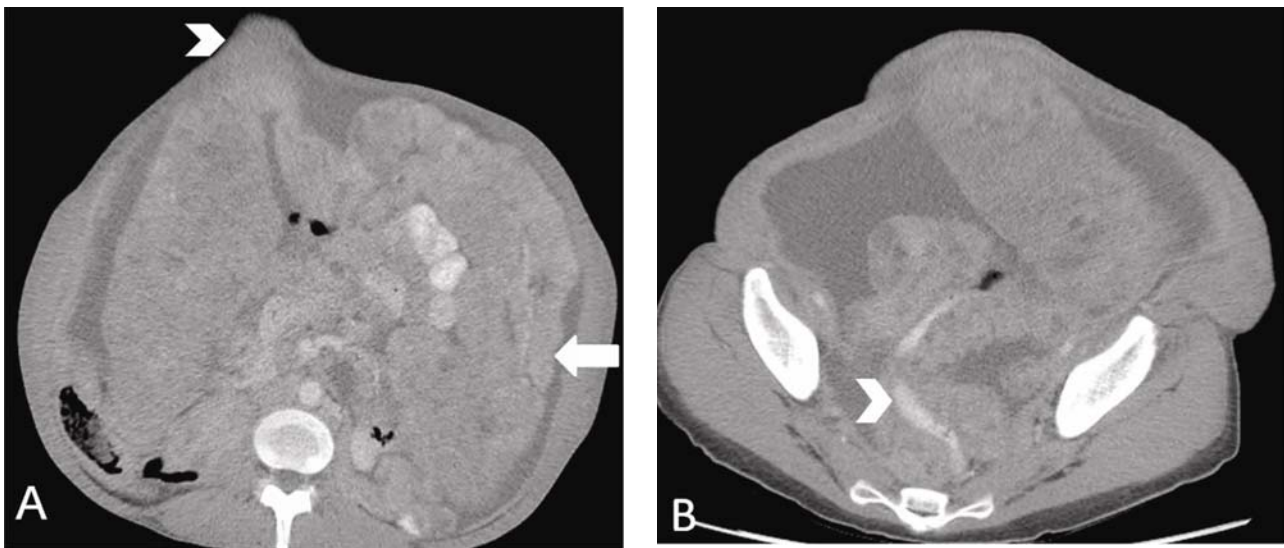
## Objective

The purpose of this article is to review the imaging findings of various uncommon peritoneal diseases and to emphasize the imaging hallmark to make a specific diagnosis or narrow differential diagnosis

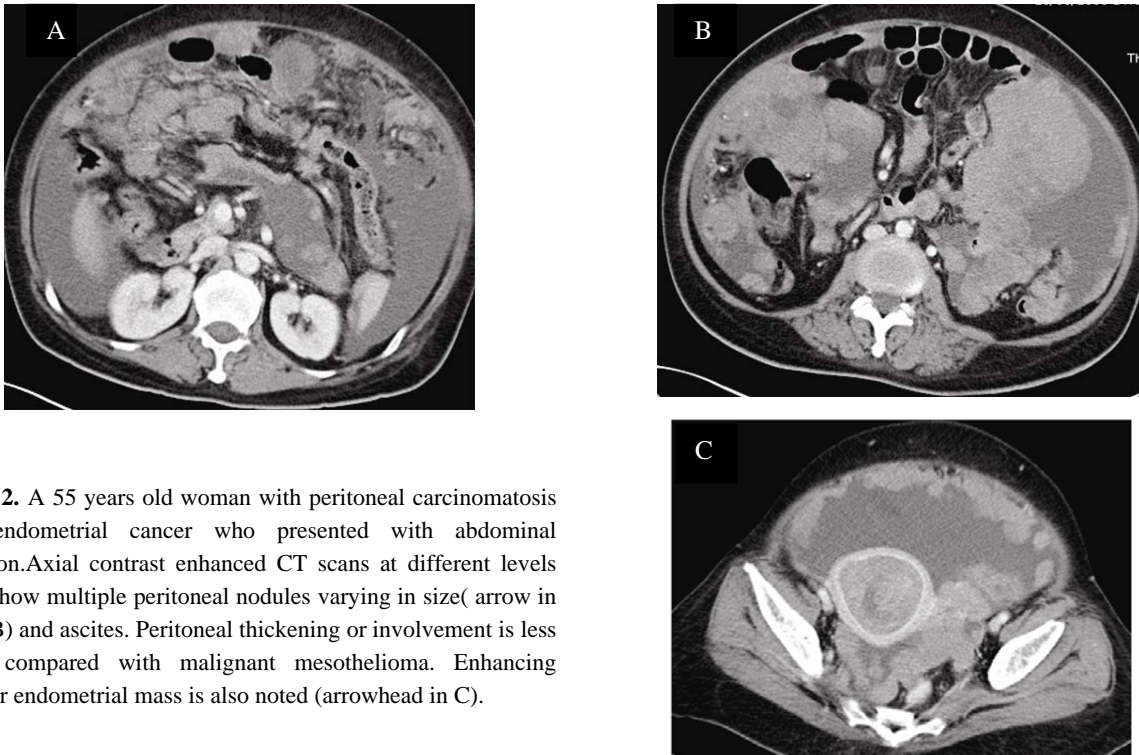
## 2 Malignant peritoneal mesothelioma

Malignant mesothelioma is a rare malignant neoplasm that arises from mesothelial cells lining the serosal membranes of body cavities such as pleura, peritoneum and pericardium. Second only to pleura, malignant mesothelioma commonly involve the peritoneal cavity. Malignant peritoneal mesothelioma accounts for 6-10% of malignant mesothelioma <sup>[1]</sup> and

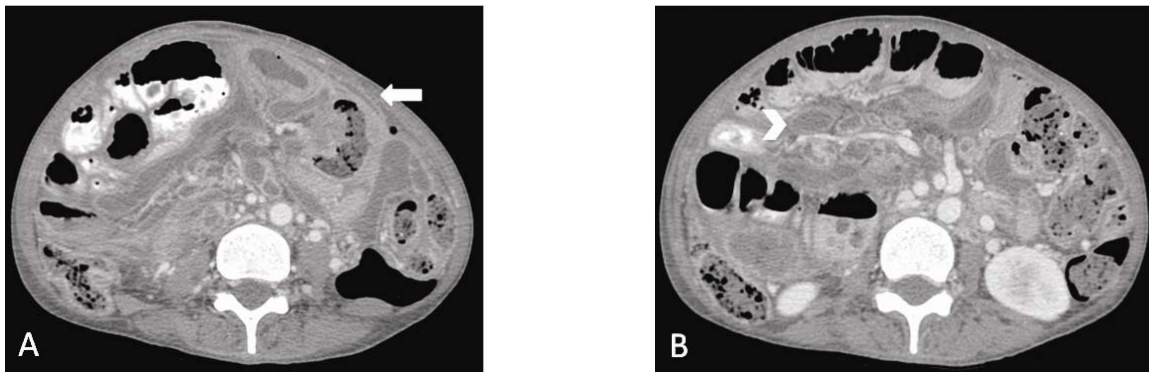
tends to be occurred in older patients during the fifth and sixth decades, men affected more than woman. Approximately half of the reported cases, the patient has a history of asbestos exposure [2]. The clinical presentation are nonspecific. Malignant peritoneal mesothelioma can be disseminated in sheets of tissue over the visceral and parietal peritoneal surfaces, then coalesce to form the masses, thereby encasing the abdominal organs or hollow viscus in the peritoneal cavity. The small bowel may become fixed, rigid, immobile and encased causing small bowel obstruction which are the sign of advanced disease. At cross sectional imaging, computed tomography (CT) and magnetic resonance imaging (MRI) are not difference in term of detection abnormality. Malignant peritoneal mesothelioma are divided into two distinct forms including diffuse and focal or localized forms reflecting the findings on the gross pathology. In diffuse form which infiltrate and irregularly or nodularly thicken peritoneum in a sheetlike fashion. As the disease progresses, the nodular thickenings become plaque like masses or omental caking (see Figure 1). Calcifications within diffuse form peritoneal involvement is considered rare in contrast to pleural counterpart [3]. Focal or localized form is characterized by dominant, moderate to large sized or localized irregular border masses with associated peritoneal studding. The attenuation is homogeneous or heterogeneous. Cystic degeneration can be seen. Calcifications within this focal or localized form is also rare. On MR, demonstrates intermediate to low signal intensity on T1 weighted image and intermediate to high signal intensity on T2 weighted image. Other shared findings include ascitic fluid, direct invasion of both solid abdominal organs and hollow viscus, and remote metastases. The amount of ascites is quite variable ranging from massive, diffuse ascites to focal, small, loculated collections of fluid. Several studies have reported that the amount of ascites is disproportionately small in relation to degree of tumor involvement in malignant peritoneal mesothelioma compared with peritoneal carcinomatosis. Nodal metastases are not the common findings. Whereas pleural involvement of malignant mesothelioma may be the coexisted feature as the disease combination. Peritoneal carcinomatosis is one of the top differential diagnosis. Evidence of primary malignant neoplasm, nodal metastases and occasionally masses with dystrophic calcification are the helpful key roles to support peritoneal carcinomatosis than that of malignant peritoneal mesothelioma (see Figure 2). Peritoneal lymphocytosis is rare with nodal involvement in peritoneum or retroperitoneum but lack of omental involvement compared with malignant peritoneal mesothelioma. Tuberculous peritonitis typically associates with lymphadenopathy probably necrosis within peritoneum and small bowel mesentery and smooth peritoneal thickening raising this concern (see Figure 3). Primary peritoneal serous carcinoma, unlike malignant peritoneal mesothelioma is likely discovered in woman with advanced age or postmenopausal woman with no history of asbestos exposure.



**Figure 1.** A 62 years-old man with malignant peritoneal mesothelioma. Axial contrast enhanced CT scans (A obtained at upper level than B) show massive thickening of peritoneum with heterogeneous enhancement (arrow). Due to its rigidity, it causes bulging of anterior abdominal wall (arrowhead in A). The sigmoid colon is encased by peritoneal mass (arrowhead in B).



**Figure 2.** A 55 years old woman with peritoneal carcinomatosis from endometrial cancer who presented with abdominal distention. Axial contrast enhanced CT scans at different levels (A-C) show multiple peritoneal nodules varying in size (arrow in A and B) and ascites. Peritoneal thickening or involvement is less severe compared with malignant mesothelioma. Enhancing irregular endometrial mass is also noted (arrowhead in C).

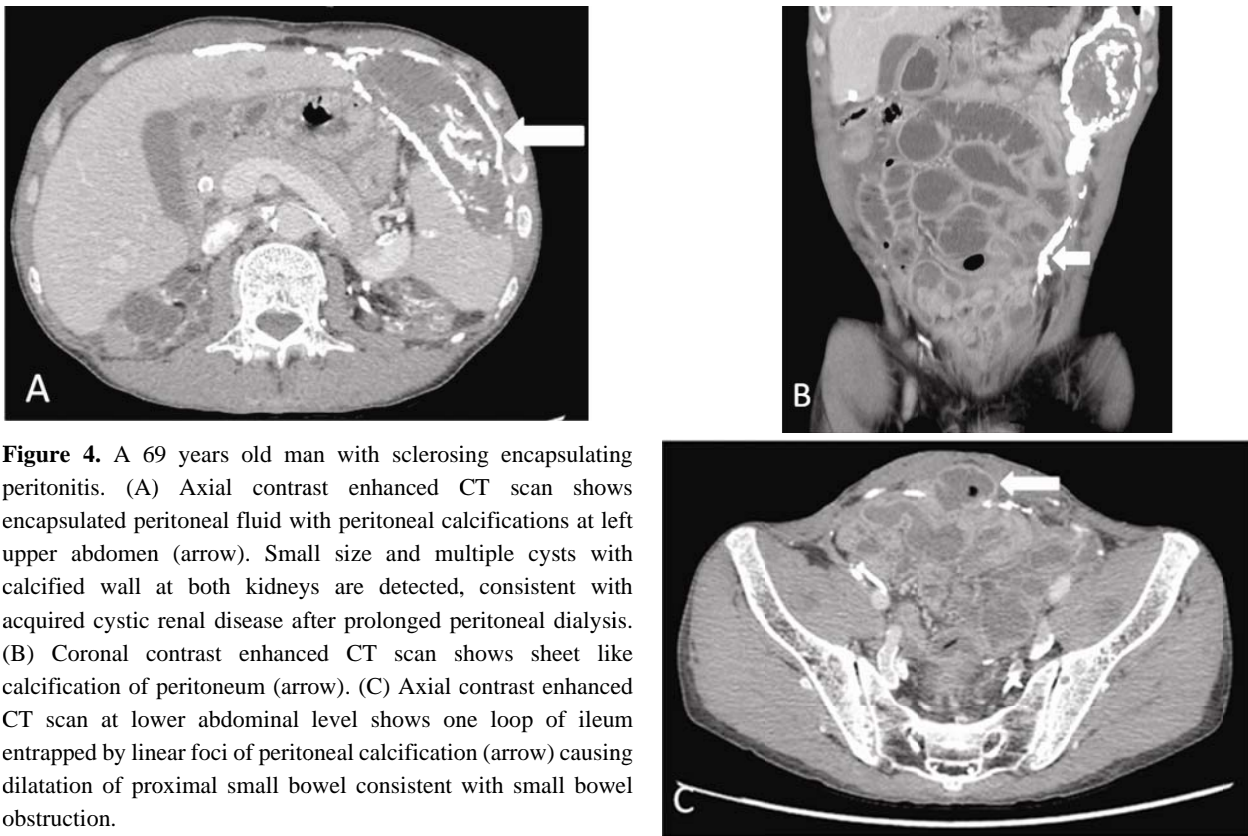


**Figure 3.** A 33 years old man with tuberculous peritonitis. Axial contrast enhanced CT scans (A and B) show smooth thickening of peritoneum (arrow), ascites and multiple mesenteric nodes with central necrosis (arrowhead)

### 3 Primary sclerosing encapsulating peritonitis

Primary sclerosing encapsulating peritonitis was first described as a complication of intermittent peritoneal dialysis in 1980<sup>[4]</sup> but now a well recognized but uncommon serious complication of continuous ambulatory peritoneal dialysis (CAPD), and the frequency of occurrence is related to duration of CAPD. In a large study by Rigly and Hawley<sup>[5]</sup>, the overall prevalence of sclerosing encapsulating peritonitis was 0.7%. It is inflammatory process of peritoneum characterized by thickening which can progress to encapsulate some or all the small bowel loops resulting in partial or complete small bowel obstruction. The bowel wall itself is thickened or calcified. Loculated fluid collection, visible peritoneal membranes and peritoneal thickening may be identified. Making diagnosis is not difficult but often delayed due to nonspecific insidious symptoms. The clinical history of CAPD especially in long duration and radiological findings are the mainstay to establish diagnosis. Clinical manifestations include abdominal pain, bloody dialysis content, sign and symptom of small bowel obstruction. Radiological CT findings of primary sclerosing encapsulating peritonitis can be simplify divided into peritoneal abnormal findings, small bowel abnormalities and loculated fluid collections<sup>[6]</sup>. Peritoneal

abnormalities show enhancement of peritoneal thickening which is seen in all cases, that can be smooth or irregular and nodular. When it progresses, it can form peritoneal encapsulation by sheath of fibrous or sclerosed peritoneum namely cocooning (see Figure 4), to involve small bowel loops. Peritoneal calcification usually begins as linear fashion involving both visceral and parietal peritoneum but extensive conglomerate calcification may be seen in advanced diseases. The visceral peritoneal calcification has been described as bowel wall serosal or mural calcification. Small bowel abnormalities include small bowel obstruction and dilatation resulting from sclerosing thickened peritoneum cocoon surrounding small bowel loops. Fibrotic thickening firstly involves outer aspect of bowel wall leading to mural fibrosis and calcification, thickening of small bowel wall, adherent bowel loops, narrowing of the bowel lumen, and proximal bowel obstruction. The fibrotic process may manifest as a mass of small bowel loops tethered together. Ultimately, small bowel necrosis with perforation may occur. Loculated fluid collection causing by fibrotic thickened peritoneal membranes are seen up to 90% of cases. It may be small or large and single or multiple <sup>[7]</sup>.



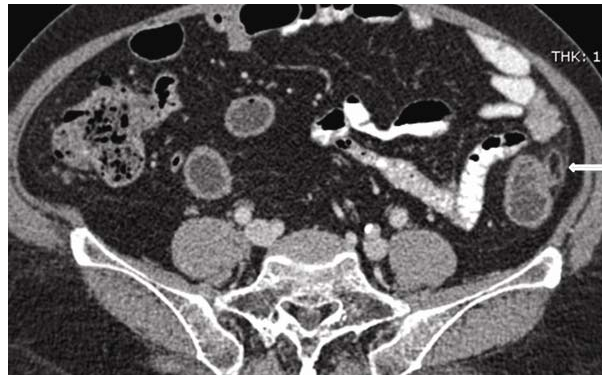
**Figure 4.** A 69 years old man with sclerosing encapsulating peritonitis. (A) Axial contrast enhanced CT scan shows encapsulated peritoneal fluid with peritoneal calcifications at left upper abdomen (arrow). Small size and multiple cysts with calcified wall at both kidneys are detected, consistent with acquired cystic renal disease after prolonged peritoneal dialysis. (B) Coronal contrast enhanced CT scan shows sheet like calcification of peritoneum (arrow). (C) Axial contrast enhanced CT scan at lower abdominal level shows one loop of ileum entrapped by linear foci of peritoneal calcification (arrow) causing dilatation of proximal small bowel consistent with small bowel obstruction.

#### 4 Intraperitoneal loose body

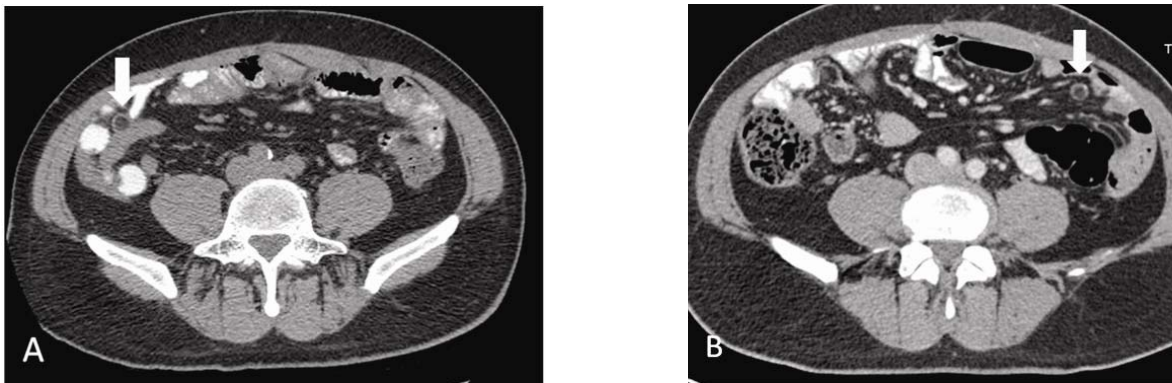
Peritoneal loose body or peritoneal mice occasionally found at laparotomy are calcified concretions that grow around nidus of necrotic tissue. However, it may also be detected at imaging for unrelated conditions. At pathology, peritoneal loose body is a laminated calcification around a core of necrotic adipose tissue and in most cases they are small in size ranging 0.5-2.5 cm and not produce any symptom. Most common origin of peritoneal loose body is appendices epiploicae. When epiploic appendages twist or torse upon themselves resulting in acute appendagitis enable to be treated conservatively (see Figure 5). In this case, the patient experienced abdominal pain and low grade fever. On the contrary, chronic torsion may not be recognized because of few or nonspecific symptoms which subsequently leading to saponification, calcification, fibrosis and eventually autoamputation, which can be freely move into peritoneal cavity. It frequently identified in a variable location at serial imaging mostly in the pelvis because it gravitate to most dependent part of the abdominal cavity. Typical CT findings include well defined soft tissue nodule with coarse calcification in the



peritoneal cavity (see Figure 6). Differential diagnosis includes retained sponge, calcified pedunculated myoma uteri and calcified mesenteric nodes. However mobility and gravity dependent is the hallmark in making diagnosis. Surgical removal is the treatment of choice.



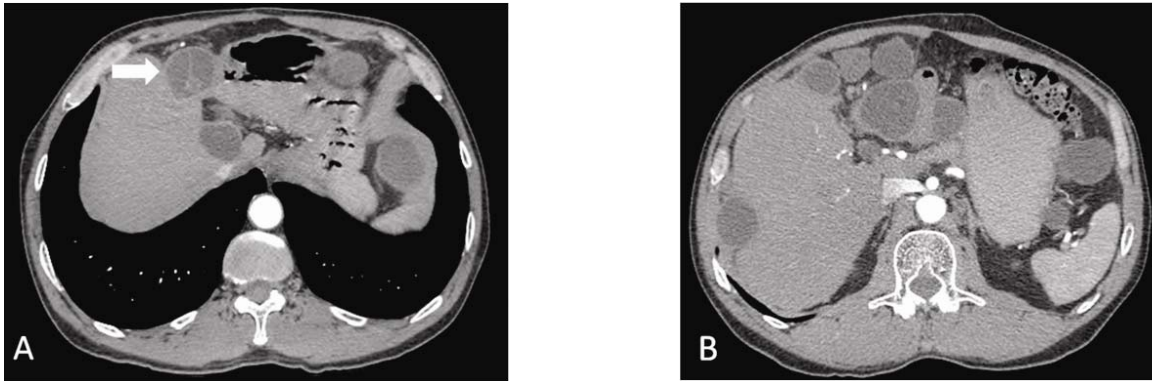
**Figure 5.** A 60 years old man with acute epiploic appendagitis who presented with left lower abdominal pain and fever. Axial contrast enhanced CT scan shows oval fatty mass near descending colon with mild thickening of adjacent peritoneum (arrow) and mild inflammatory stranding, representing inflamed torsed epiploic appendage.



**Figure 6.** A 58 years old man with peritoneal loose body. (A) Axial contrast enhanced CT scan shows well defined oval fatty nodule with partial calcification at right lower abdomen (arrow). (B) Axial contrast enhanced CT scan obtained 3 months earlier shows variable location of this fatty nodule at serial images as it was previously identified at left lower abdomen (arrow).

## 5 Peritoneal echinococcosis

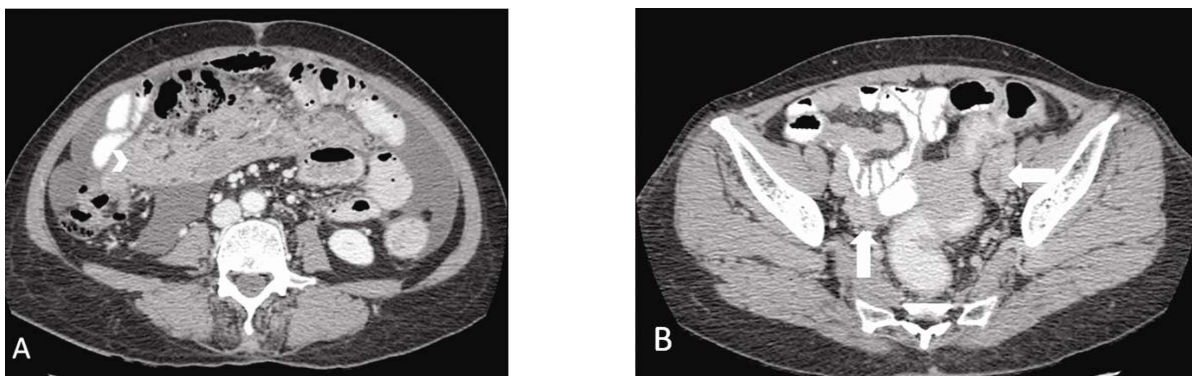
Peritoneal echinococcosis or hydatidosis is uncommon findings caused by *Echinococcus granulosus*, a parasite or helminth of cestode group. It is categorized into primary and secondary peritoneal echinococcosis. Primary peritoneal echinococcosis is a rare disease and only a few cases have been reported in which the mechanism of primary parasitic infestation is not clear. The possible explanation to produce primary peritoneal echinococcosis outside liver or lungs is dissemination via lymphatic or systemic circulation. Secondary peritoneal echinococcosis is more common, related to hepatic infection counterpart. Microrupture of hepatic hydatid cyst, mostly asymptomatic or leakage of echinococcal fluid upon time of surgery result in peritoneal contamination. Occasionally, post traumatic rupture of hepatic hydatid cyst may also cause peritoneal seeding. Intraperitoneal rupture of hepatic hydatid cysts result in release of brood's capsule and scolices and daughter cyst into peritoneal cavity. Hydatid cysts are classified into four types on the basis of appearance including type I simple cyst with no internal architecture, type II cyst with daughter cyst(s) and matrix, type III calcified cyst and type IV complicated hydatid cysts include rupture and superinfection. At imaging, CT is the imaging modality of choice to delineate the extent of peritoneal echinococcosis. CT findings include cysts usually multiple arise anywhere in peritoneal cavity with characteristic appearance of daughter cysts, thickening of cyst wall, calcification of the wall, presence of internal septations and hydatid sand (see Figure 7).



**Figure 7.** A 35 years old man with peritoneal echinococcosis. (A-B) Axial contrast enhanced CT scan through upper abdomen show multiple cystic lesions with daughter cysts scattering in peritoneal cavity. Ruptured hepatic echinococcosis is likely (arrow).

## 6 Primary serous papillary carcinoma of peritoneum

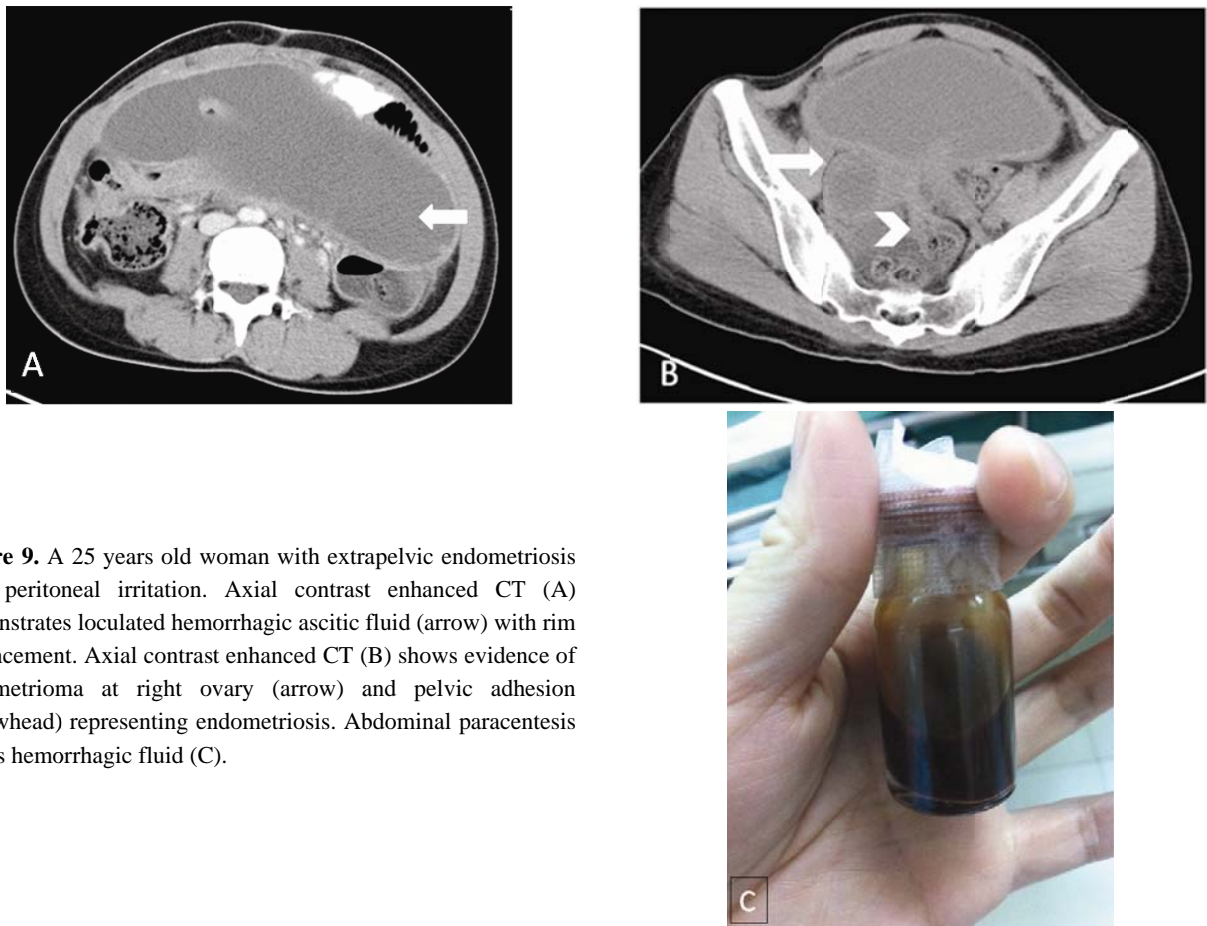
Primary serous papillary carcinoma of peritoneum is a rare malignant epithelial tumor arises from either peritoneal mesothelial cells with mullerian differentiation or nests of ovarian tissue remnants within peritoneum. It is not distinguishable primary serous papillary carcinoma of peritoneum from peritoneal carcinomatosis. Zissin et al<sup>[8]</sup> reported that 6 of 30 patients with primary serous papillary carcinoma of peritoneum had adnexal masses or ovarian enlargement on CT but no evidence of tumor involvement was seen in both ovaries on pathology. Primary serous papillary carcinoma of peritoneum is diagnosed when these following criterias have been met, including normal both ovaries, greater involvement of extraovarian sites than the involvement on the surface of ovary or limited involvement at ovarian surface epithelium but no stromal invasion or involving cortical stroma with tumor size less than 5 mm × 5 mm. Imaging findings of ascites, enhancing peritoneal nodules, peritoneal thickening, and enhancing omental masses on CT or MRI, similar to those of peritoneal carcinomatosis but usually no identifiable adnexal mass (see Figure 8). However some patients may have no ascites or only small quantity at time of presentation. Calcification within peritoneal or omental mass represent psammoma bodies histopathologically. The size of ovaries are usually normal, despite tumor implants may be presented on the surface of ovary. Therefore the diagnosis of primary serous papillary carcinoma of peritoneum should be awared in elderly woman with diffuse peritoneal lesions mimicking peritoneal carcinomatosis and normal appearance of both ovaries without demonstrable primary malignancy. Besides peritoneal carcinomatosis, malignant peritoneal mesothelioma and TB peritonitis may mimic primary serous papillary carcinoma of peritoneum. But malignant peritoneal mesothelioma is likely to occur in patient with asbestos exposure and pleural involvement may be occurred. In TB peritonitis, low density center or necrotic lymph nodes or calcified lymph nodes and abnormal chest findings are seen. Laparotomy should be performed to make final diagnosis in the case of diagnostic dilemma.



**Figure 8.** A 45 years old woman with primary serous papillary carcinoma of peritoneum. (A and B) Axial contrast enhanced CT shows ascites, peritoneal stranding (arrowhead in A) and enhancement, mimicking peritoneal carcinomatosis but normal both ovaries (arrow in B).

## 7 Extrapelvic endometriosis with peritoneal irritation

Endometriosis is defined as the presence of endometrial tissue outside the uterine cavity generally involve ovaries and rectovaginal septum. The ascitic fluid is likely to be the consequence of peritoneal irritation due to ruptured endometriotic cysts. The ascites is exudative resembling gynecologic malignancy. Only 30-40 cases of endometriosis with ascites have been reported in the literature, reflecting the rarity of this condition. However, hemorrhagic ascites secondary to endometriosis is an unusual occurrence. The clinical presentations include abdominal distention, weight loss or abdominal pain. Treatment with GnRH analog to suppress ovarian function is the effective conservative treatment. However long term follow up is recommended because of the high risk of recurrence. Therefore, extrapelvic endometriosis with peritoneal irritation should be considered in premenopausal women who develop bloody ascites (see Figure 9).



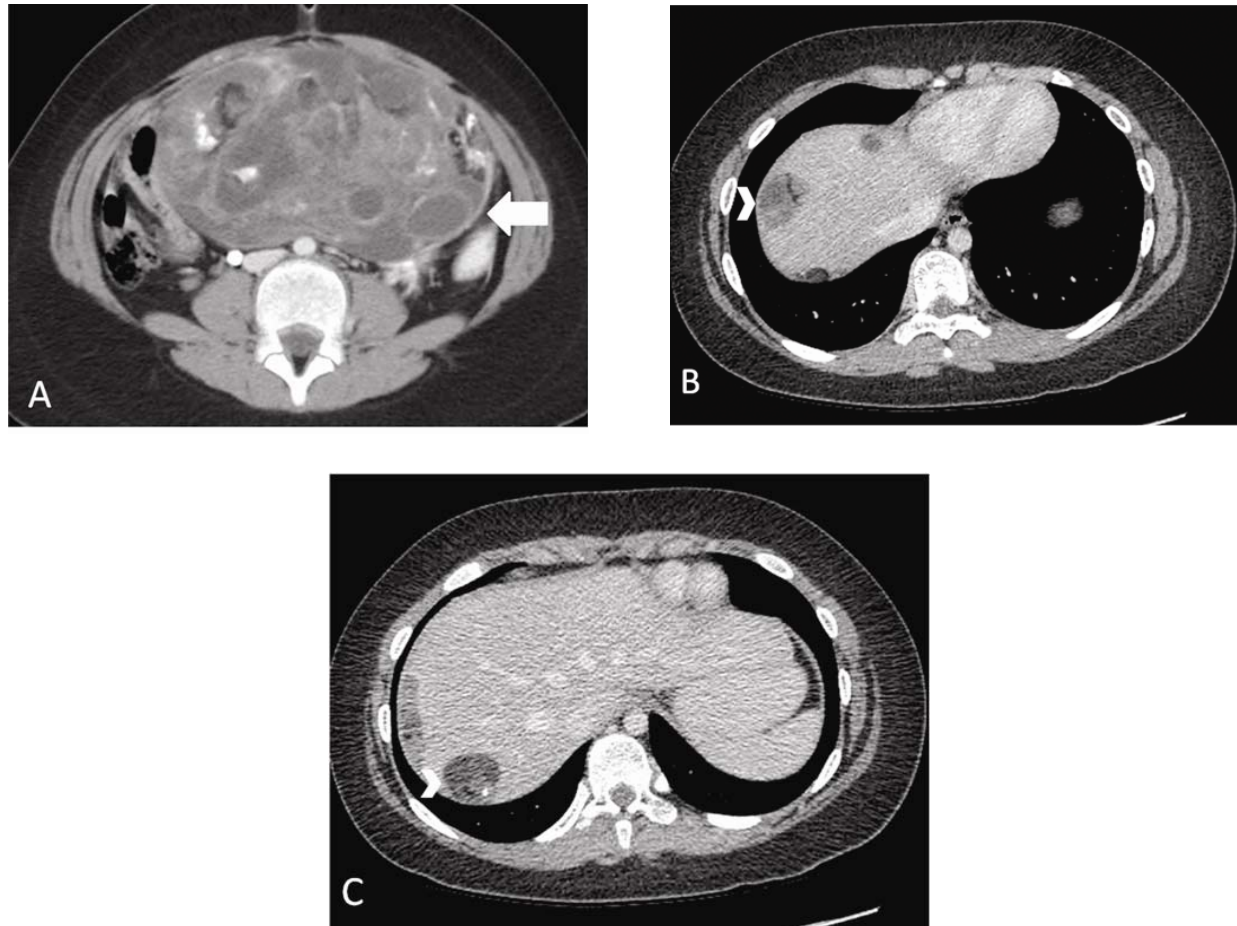
**Figure 9.** A 25 years old woman with extrapelvic endometriosis with peritoneal irritation. Axial contrast enhanced CT (A) demonstrates loculated hemorrhagic ascitic fluid (arrow) with rim enhancement. Axial contrast enhanced CT (B) shows evidence of endometrioma at right ovary (arrow) and pelvic adhesion (arrowhead) representing endometriosis. Abdominal paracentesis shows hemorrhagic fluid (C).

## 8 Ruptured ovarian teratoma with peritoneal dissemination

Ovarian teratoma is a common ovarian tumor accounting for 20% of adult ovarian tumors. It is composed of all three germ cell layers including ectoderm, mesoderm and endoderm. The clinical manifestation of ovarian teratoma are varied. Most patients present with incidentally discovered asymptomatic adnexal mass on radiological imaging. It is not difficult to obtain the diagnosis on radiologic examination. Typical imaging findings of ovarian teratoma are cystic ovarian mass with intratumoral fat component and calcification. Ovarian teratoma can be associated with various complications such as torsion, rupture, malignant transformation, infection and autoimmune hemolytic anemia. Spontaneous rupture of ovarian teratoma is an uncommon entity due to the presence of thick capsule. Rupture occurs in 1-4% of ovarian teratoma. When it ruptures, the peritoneum is irritated by liquefied sebaceous content leading to acute or chronic peritoneal inflammation. In



acute peritonitis, it causes by sudden rupture of tumor contents that may result on shock or hemorrhage and usually associate with torsion, trauma, infection or labor. In more common chronic peritonitis, the recurrent or chronic leakage of sebaceous material irritating peritoneal lining causing chronic granulomatous peritonitis. At U/S,CT or MRI, variable amount of ascites with fat fluid level, diffuse or focal omental infiltration, multiple fatty peritoneal implants and dense adhesion may be presented in both acute and chronic peritoneal inflammation (see Figure 10). Peritoneal carcinomatosis and TB peritonitis may simulate this entity but ascites with fat fluid level and fatty peritoneal masses are strongly support this diagnosis.



**Figure 10.** A 38 years old woman with ruptured ovarian teratoma with peritoneal dissemination. (A-C) Axial contrast enhanced CT shows abdominal mass with fat and calcifications consistent with teratoma (arrow in A). Noted multiple fatty peritoneal implants at perihepatic space (arrowhead in B and C).

## 9 Peritoneal inclusion cysts

Peritoneal inclusion cysts also known as peritoneal pseudocyst is a fluid-filled cysts that conform to the shape of peritoneal cavity results from nonneoplastic reactive mesothelial proliferation. Most patients with peritoneal inclusion cysts present with pelvic pain or pelvic mass. It occurs almost exclusively in premenopausal woman with active ovaries, pelvic adhesions, history of previous abdominal or pelvic surgery, trauma and impaired absorption of peritoneal fluid. It ranges in size from small to large that may fill entire pelvis and abdomen but tend to grow slowly. Ultrasound, CT or MRI typically show cystic masses with septations or loculated fluid collection within the pelvis but no enhancing solid component. The ovaries are usually normal or entrapped by but clearly separate from cystic locules



## 10 Conclusion

Diverse uncommon diseases of peritoneum including malignant peritoneal mesothelioma, primary sclerosing encapsulating peritonitis, intraperitoneal loose body, peritoneal echinococcosis, primary serous papillary carcinoma of peritoneum, extrapelvic endometriosis, ruptured ovarian teratoma with peritoneal dissemination, and peritoneal inclusion cyst may have overlapping imaging appearances with those of peritoneal carcinomatosis. But some distinctive imaging features that described in this article is helpful to allow the specific diagnosis to be documented.

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