Sclerosing encapsulating peritonitis: Imaging Findings

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Learning objectives

Our objective is to describe the most characteristic imaging findings in the sclerosing peritonitis, by reviewing different imaging techniques, with emphasis on computed tomography (CT).
Background

Sclerosing peritonitis is a condition characterized by chronic fibrotic thickening of the peritoneum, with a broad spectrum of gastrointestinal symptoms, which in severe cases causes encapsulation of the small bowel loops, known as sclerosing encapsulating peritonitis or abdominal cocoon.

It is a rare entity, with a high morbidity and mortality. It is important to consider this pathology in patients at risk, to detect the earliest signs, and to be able to carry out the appropriate treatment.

Of uncertain etiology, it is believed that chronic irritation of the peritoneum is its cause.

It can be classified idiopathically or secondarily. Within the first, it may be found in young women from tropical or subtropical countries [1]. The reason for this prevalence remains unknown.

Among secondary causes, it can be related to continuous ambulatory peritoneal dialysis (CAPD), fungal infections, tuberculosis, prolonged use of beta-blocker practolol, ventriculoperitoneal and peritoneovenous shunts, orthotopic liver transplant, abdominal surgery, sarcoidosis, systemic lupus erythematosus, familial Mediterranean fever, deficiency of the S protein and luteinized-ovarian tecoma [2-3].

CAPD is one of the most frequent causes, with prevalence between 0.4% and 8.9%, and the risk of development after 5 years of peritoneal dialysis varies between 0.6% and 6.6% [4]. It can also show up many years after cessation, so it is of the utmost importance to be clinically aware.

In the histology of the membranes, we find fibrocollagenous tissue with nonspecific inflammatory component. These changes that are characteristic of sclerosing peritonitis, are not specific and overlap with membrane changes that occur with ultrafiltration failure and infectious peritonitis in long-term peritoneal dialysis [4].

Clinically, its presentation can be asymptomatic, or with nonspecific symptoms such as abdominal pain and distension, nausea, vomiting, palpable mass, anorexia, weight loss and recurrent episodes of small-bowel obstruction. These can lead to malnutrition, sepsis and death. Loss of ultrafiltration and bloody dialysis effluent is seen in dialyzed patients [4]. Elevated C-reactive protein, anemia and hypoalbuminemia may be found [1], but there are neither specific blood markers nor reliable screening methods.

Diagnosis is based on clinical suspicion and confirmed by imaging methods or surgically.

The differential diagnosis related to peritoneal calcifications includes tuberculosis, amyloidosis, hyperparathyroidism, pseudomyxoma peritonei and peritoneal
carcinomatosis. Among small bowel obstruction, the internal hernia must be considered as it can also produce abnormal clustering of small bowel loops [3].

The treatment is related to its cause, including adhesiolysis surgery or immunosuppression with corticosteroids, tamoxifen or sirolimus, and an adequate nutrition. No surgical treatment is required if it is asymptomatic [5]. If it is associated with CAPD, it must be suspended and the patient must be transferred to hemodialysis [4].
Findings and procedure details

Although CT is the best method to study this entity, other techniques can be helpful in its diagnosis. Findings with different imaging methods are detailed below:

**Radiography (X-ray):**

Radiographic findings are nonspecific and include centralize and curve calcifications corresponding to small-bowel or visceral peritoneum, or peripheral calcifications which can correspond to parietal peritoneum calcifications (Fig. 1 on page 7). In small-bowel encapsulation, centrally located gas-filled and dilated loops of bowel may be seen [6].

**Ultrasound (US):**

In early stages there is an increase in peristalsis and peritoneal collections, and fine echogenic bands in the ascitic fluid, especially perihepatic (Fig. 2 on page 7). Then, in more advanced stages, thickening of the intestinal wall and hypomotility of the small intestine, and thickening of the peritoneum, which can calcify. When progressing, it can be observed as an echogenic mass of dilated small intestine due to the piling up of them (Fig. 3 on page 8), encircled by a thick hypoechoic fibrous membrane [7].

**Computed Tomography (CT):**

Imaging findings are mainly based on CT, the best method to study this entity. It can be divided into peritoneal and small-bowel abnormalities, and loculated fluid collections, which may be observed singly or in combination.

The peritoneum is a thin line, hardly seen normally, and its thickening is seen in all cases [6]. The diffuse fibrosing inflammatory process that affects the peritoneum generates its thickening, which is enhanced after the administration of intravenous (IV) contrast (Fig. 4 on page 9 and Fig. 5 on page 10), however this finding can also be observed in infectious pathology. This enhancement is better seen when the peritoneum is next to low attenuation structures, such as fluid or fat [6]. The presence of initially linear calcification (Fig. 6 on page 11) that can affect both peritoneal sheets is also observed, but as the disease progresses, conglomerates can form (Fig. 7 on page 12), even dystrophic calcification due to chronic kidney disease. Increase in calcification is associated with severing clinical symptoms.

The small-bowel is encapsulated, with an entangled appearance, by the thickened peritoneum, which can generate intestinal obstruction. The fibrotic process of the peritoneum can progress to affect the walls of the intestinal loops, generating mural
fibrosis with calcifications (Fig. 8 on page 13), thickening of the wall in advanced phases (Fig. 7 on page 12), adherence of the loops with decrease in light and, finally, obstruction [8].

In 90% of the cases, the presence of liquid collections stands out, being able to be single or multiple (Fig. 9 on page 14). They are due to the presence of loculated ascites, between the peritoneal leaves, and increase in size as the disease progresses. The fluid collections contribute to the central location of the tethered small-bowel loops by way of mass effect [8].

**Magnetic Resonance (MR):**

Its findings similar to those of a CT (Fig. 10 on page 15), although its convenience has not been investigated. Although there is no use of radiation, its higher cost, and the potential risk of nephrotoxicity and nephrogenic systemic fibrosis due to the use of gadolinium, restrains the use of this method [9]. An alternative can be a MR without gadolinium, using dialysate as a contrast medium, but there is no updated experience using this technique to evaluate patients with sclerosing peritonitis [10].

**Positron emission tomography (PET):**

It could be useful for diagnosis during the inflammatory phase, in which an increase in uptake of the tracer by the peritoneum and small-bowel loops can be seen (Fig. 11 on page 16), but its usefulness is limited by the lack of specificity of these findings, which can also be observed in cases of acute peritonitis [9]. There is little experience on this method.
Fig. 1: Abdominal radiograph (AP). Multiple linear calcifications corresponding to small-bowel wall (arrow) and peritoneum (arrowhead). Signs of osteodystrophy are also seen. Same patient as in Figure 8.

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**Fig. 2:** Abdominal US. Transverse gray-scale image of the left flank shows a loculated and septated fluid collection (arrow).

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**Fig. 3:** US Doppler. (A) Transverse image of the right lower quadrant of the abdomen, showing a mass of tethered small-bowel loops (arrows) with wall thickening and hyperflow signs. (B) The superior mesenteric artery shows decreased resistive index (RI: 0.80) and hyperflow signs (500 ml/min).

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Fig. 4: Axial contrast-enhanced CT image of the upper abdomen. Parietal peritoneum thickening and enhancement (arrows).

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Fig. 5: Axial CT with IV contrast of the mid abdomen. Visceral peritoneal thickening (arrow) and a mass of dilate small-bowel loops (arrowheads).

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**Fig. 6:** Axial CT with IV contrast of the mid abdomen. Linear calcifications of the peritoneum (arrows) and dilated small-bowel loops and thickening of its wall (arrowhead). A small subhepatic collection is seen (black arrow).

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Fig. 7: Axial CT with IV contrast of the mid abdomen. Conglomerate calcifications of the peritoneum (arrow) and dilated small-bowel loops and thickening of its wall (arrowhead).

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Fig. 8: Coronal CT post-IV contrast portal venous phase of the abdomen and pelvis. Dilated small-bowel loops, with mural thickening and calcifications (arrows). Also conglomerate calcifications of the peritoneum (arrowhead). Same patient as in Figure 1.
Fig. 9: Axial CT post-IV contrast portal venous phase of the mid abdomen. Large loculated fluid collection (arrows) with mass effect tethering centrally the small-bowel loops.

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**Fig. 10:** 55-year-old man with chronic abdominal pain. (A-B) MR, T1 Fat Sat with IV contrast (portal venous phase) of the abdomen: (A) Axial and (B) Coronal showing enhanced thickened peritoneal membrane (arrows) covering congregated and dilated small-bowel loops with wall thickening (arrowheads). (C) Intraoperative photograph shows a thick membrane encapsulating the small bowel (arrows). (D-E) Microscopic view of the peritoneum with hematoxylin and eosin (H&E): (D) 40x high power showing fibrosis with dense collagen fibers and chronic inflammation (arrowhead); (E) 100x high power showing perivascular chronic inflammatory cells (arrowheads), infiltrate composed of lymphocytes and plasma cells.

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**Fig. 11:** Axial PET-CT fusion. Increased uptake of the radiotracer (18F-FDG) in small-bowel (arrows).

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Conclusion

Sclerosing encapsulating peritonitis is mainly a rare complication of peritoneal dialysis, with nonspecific symptoms. It has a high morbidity and mortality due to small-bowel obstruction. It is important to know the findings of this entity and keep them in mind, when studying patients at risk like in CAPD or, related to the idiopathic form, with subacute or chronic abdominal pain with no clinical history of peritoneal dialysis or other diseases. CT is needed to confirm this entity, and its diagnosis should not be delayed in order to treat the patient.
Personal information

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References