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Review

An unusual case of extensive peritoneal calcification: A case report

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Abstract

The peritoneum is the largest serous membrane of the body and can be exposed to several injuries that may cause abnormal findings on imaging exams. Linear peritoneal calcification is remarkably rare, usually secondary to long duration peritoneal dialysis.

We report an uncommon case of extensive peritoneal calcification in a 39-year-old female without long exposure to peritoneal dialysis solutions, in which peritoneal calcification could be linked to Alport syndrome and previous adverse reaction to intraperitoneal iodinated contrast.

Radiologist should be aware of this and related imaging findings, know when to search for them as well as understand their clinical value.

Keywords: Peritoneal calcification; Encapsulating Peritoneal Sclerosis; Alport syndrome; Iodinated contrast

Contents

1. Introduction

Cases of linear peritoneal calcification have been reported in literature, though they occur rarely. In Encapsulating Peritoneal Sclerosis (EPS) a peritoneal membrane damage develops an inflammatory cascade that results in sclerosis and eventually calcification [1].

EPS has been can be either primary or secondary, being long exposure to peritoneal dialysis solutions [2,3] the most common cause of the secondary form. The incidence of EPS has only been studied in patients on peritoneal dialysis and is estimated to be 0.54–4.4% [4], although this number can rise considerably with the time on PD.

The diagnosis of EPS combines clinical symptoms with pathological and imaging findings [5]. The symptoms manifest disturbances in intestinal function such as abdominal pain, nausea, vomiting and ultimately anorexia and weight loss [6]. Among the imaging techniques available, CT is the modality of choice in the diagnosis of EPS, demonstrating peritoneal thickening, calcification, bowel wall thickening, bowel tethering, dilation and fluid loculation [5]. Final diagnosis requires direct observation of peritoneum and histology [7].

In a symptomatic patient, the mortality associated with EPS is high, reaching 60% 4 months after the diagnosis [6]

2. Case presentation

In August 2014 a 39-year-old female presented to the emergency department of our hospital. She had Alport disease, end-stage renal disease and secondary hyperparathyroidism.

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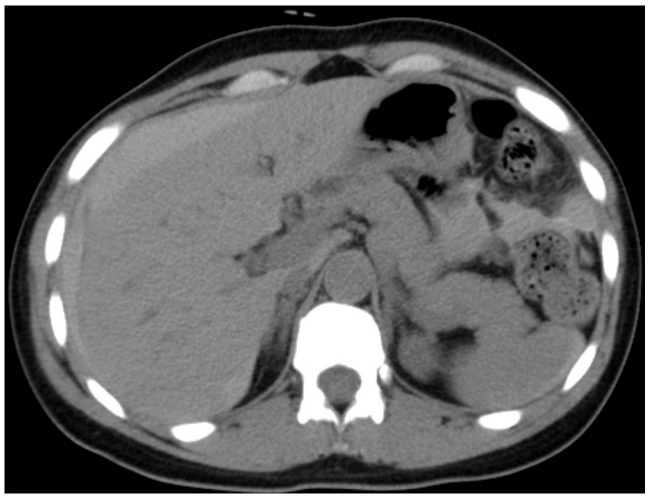

Fig. 1. An abdominal CT was obtained with iodinated contrast injected through the peritoneal catheter (CT peritoneography). The exam revealed a good diffusion of contrast, with no images of leak or abdominal collections.

She referred a past history of one cesarean followed by a laparoscopic tubal ligation. In October 2013 she initiated peritoneal dialysis. The catheter introduced was soon found dysfunctional, with no drain of dialysate. Laparoscopic removal of adhesions and catheter repositioning were performed, but complicated with hemoperitoneum. Peritoneal lavage by the peritoneal catheter was performed, but abdominal wall swelling was noticed and a leak within the laparoscopic port was suspected.

A CT peritoneography (Fig. 1) was obtained to search for an abdominal leak. Iodinated contrast was injected through the peritoneal catheter and the patient was encouraged to walk, allowing a good diffusion of contrast through the peritoneal cavity. Shortly after contrast injection the patient developed intense abdominal pain and hypotension that were attributed to contrast adverse reaction. The CT was obtanied 30 min after contrast injection, revealing a good diffusion of contrast, with no images of leak or abdominal collections. The contrast was then drained through the catheter. Ten days later the patient was seen for abdominal pain and elevated inflammatory markers (C-reactive protein = 40) and the Tenckhoff catheter was removed for a suspected infection. Cultures of peritoneal fluid were negative. Antibiotics for twelve days were given and the patient improved clinically and analytically. The patient chose then to start hemodialysis.

On the day she visited our emergency department, she reported complaints of lumbar pain and dysuria for the previous few days. She had no nausea nor vomiting. She had no fever. On physical examination the abdomen was soft and nontender. The bowel sounds were normal. Laboratory data were normal, except for: hemoglobin = 8.9 (normal 12–16 g/dL), leukocytosis = 14,700 (4000–10,000), leukocyturia > 200/field (<5/field); C-reactive protein = 23.5 (<1.0), urea = 22.4 (2.4–6.4 mmol/L), creatinine = 316 (46–92 μmol/L), PTH = 324 (16–87 pg/mL), calcium = 2.40 (2.10–2.55 mmol/L) and phosphorus 1.47 (0.41–1.45 mmol/L).

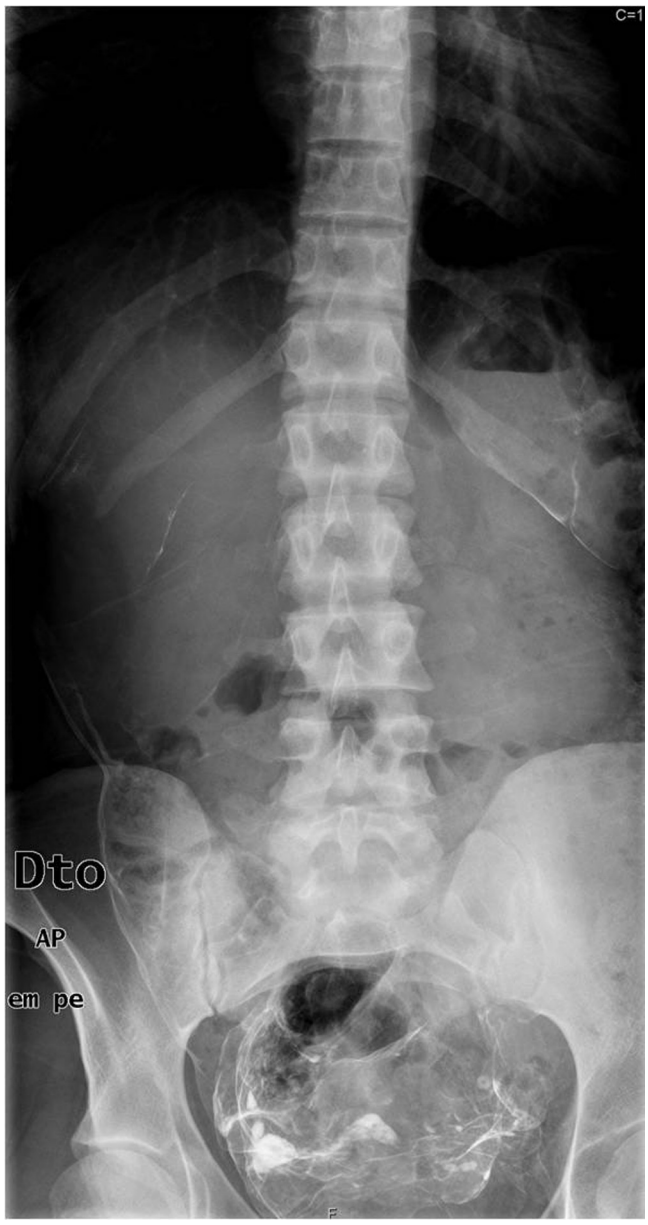

Fig. 2. A lumbar radiography showed diffuse peritoneal calcification, most evident in the lower abdomen.

A plain lumbar radiography (Fig. 2) showed diffuse peritoneal calcification, most evident in the lower abdomen. There were no abnormalities of the lumbar spine. Renal ultrasound was normal, with no signs of renal obstruction.

The abdominal CT (Fig. 3) revealed extensive visceral (arrows in a) and parietal peritoneal calcification (arrowheads in a) with areas of focal thickening in the pelvic peritoneum (arrowheads in c).

Antibiotics to the urinary infection were given and the patient was hospitalized. Blood cultures were negative, while *Escherichia coli* was isolated in the urine culture. She improved clinically, inflammatory markers decreased and she was then discharged from the hospital with no symptoms.

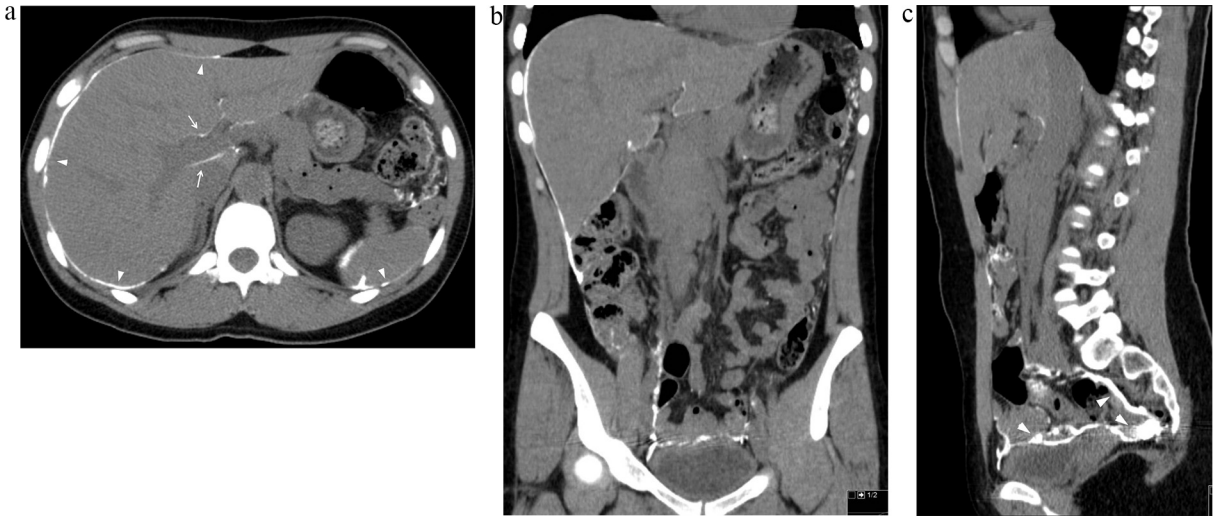

Fig. 3. The abdominal CT without contrast revealed extensive visceral (arrows in a) and parietal peritoneal calcification (arrowheads in a) with areas of focal thickening in the pelvic peritoneum (arrowheads in c).

3. Discussion

Cases of linear peritoneal calcification have been reported in patients on long-standing peritoneal dialysis and have previously been classified as "Calcifying Peritonitis" [8,9]. This term is not currently used but is now included in the spectrum of EPS [8].

EPS has been described as primary or, more frequently, secondary. Long exposure to peritoneal dialysis solutions is the most cited caused, although it can also develop after renal-transplant [10], with hyperparathyroidism [8,11], with the use of certain beta-blockers [3] or after recurrent episodes of peritonitis [8,12], among others. Sampimon [13] demonstrated an increased susceptibility for patients with Alport disease to develop EPS.

The "two-hit" model has been used to explain the pathophysiology of EPS, hypothesizing that a predisposing factor (injury) and an initiating factor (such as an inflammatory stimuli) are required for EPS to develop [14]. From this model we can assume that, in a susceptible patient, it can develop after any peritoneal inflammatory stimuli. Eisenberg and colleagues [15] reported the development of peritoneal inflammation secondary to iodinated contrast agents in Guinea-pigs.

To the best of our knowledge no cases of EPS secondary to intraperitoneal contrast have been reported. This can be explained by the low frequency of its use. Nevertheless we believe that it could have been the cause of our imaging findings.

Clinically, patients with EPS can be asymptomatic or may present with symptoms caused by modifications of gastrointestinal transit. Nakamoto [16] divided EPS into four clinical and pathological progressive stages, ranging from lack of symptoms in stage 1 to complete bowel obstruction and anorexia characterizing the stage 4.

Imaging findings will translate the pathological progression of the disease, with peritoneal calcification predominating in the early stages and the most advanced cases presenting with findings of bowel obstruction such as air-fluid levels, dilated loops or even bowel clustering. On the different imaging techniques, abdominal radiography shows a low accuracy in this diagnosis, being either normal or revealing peritoneal calcification, air-fluid levels or loop dilations [5]. Contrast studies can reveal proximal small bowel dilatation, delayed transit time and, in advanced cases, "cauliflower sign" attributable to lower quadrant agglomeration of bowel loops [17]. Abdominal ultrasound can show peritoneal thickening, loculated peritoneal fluid, peritoneal calcification, adherences, dilated or diminished peristaltic intestinal loops [5]. Contrast CT is regarded as the imaging modality of choice [5]. Diagnostic criteria have been developed by Tarzi [18] and Vlijm [19]. Tarzi created a 22-point score, considering peritoneal thickening, peritoneal calcification, bowel wall thickening, bowel tethering (0–4 points each), loculation and bowel dilation (0–3 points). The mean score of EPS patients was 9 (2–16), comparing to an average score of 1 (0–3) in controls patients on peritoneal dialysis. Magnetic Resonance has been proposed, although less studied in these patients [20]. Positron emission tomography can show an increased peritoneal uptake but is not able to distinguish from acute peritonitis [5].

In our case, the diagnosis was made by CT, although it could already be suspected by the radiography. No intravenous contrast was given due to the previous reaction to contrast. Our patient CT score is 5, inferior to the average obtained by Tarzi, but matching an early stage of disease that we expected due to the lack of symptoms (stage 1). The focal peritoneal thickening observed in the most dependent portions of the abdomen might be explained by the longer time exposure of these areas to the peritoneal contrast.

Definitive diagnosis is made from direct observation of peritoneum and histology. Laparotomy is indicated in later stages of the disease and shows peritoneal thickening, adhesions, tethering, fibrosis or bowel retraction [7]. Histological changes in the peritoneum include interstitial fibrosis, capillary proliferation and calcification [7].

No specific treatment has been developed for EPS, being proposed the use of total parenteral nutrition [3,21], corticoids

[3,21], tamoxifen [22,23] and surgery with enterolysis of intestinal adhesions for advanced cases [3,21]. When presenting with symptoms EPS has a high mortality, usually as a result of bowel obstruction, malnutrition and sepsis [6].

We demonstrated an unusual case of peritoneal calcification, in which there was no history of long exposure to peritoneal dialysis solutions. We believe that intraperitoneal iodinated contrast exposure combined with Alport syndrome have caused the abnormalities seen. Long standing hyperparathyroidism might also have played a role explaining the increased susceptibility of our patient for the development of EPS.

Conflict of interest

The authors wish to confirm that there are no known conflicts of interest associated with this publication and there has been no financial support for this work that could have influenced its outcome.

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