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In-Depth Clinical Review

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Imaging in encapsulating peritoneal sclerosis

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Abstract
Encapsulating peritoneal sclerosis (EPS) is a rare but very severe complication of long-term peritoneal dialysis (PD). Since the first reports on this disease in the eighties, several imaging techniques have been used for its diagnosis. Because of the rarity of this condition, uniformity in modality and protocols for abdominal imaging for diagnosis has been lacking overtime. Nowadays, computed tomography (CT) is most often used. In this review, we provide an overview of all imaging modalities that have been used overtime to diagnose EPS as a late complication of PD. Imaging features characteristic for EPS and advantages as well as shortcomings of all modalities are discussed. We believe that when EPS is suspected, CT with contrast enhancement should be the modality of first choice in clinical practice.

Keywords: abdominal imaging; encapsulating peritoneal sclerosis; imaging modalities

Introduction

Encapsulating peritoneal sclerosis (EPS), a condition in which a fibrous cocoon has surrounded the bowel loops [1], is an uncommon but devastating complication of chronic peritoneal dialysis (PD). Long PD duration and chronic exposure to dialysis solutions are considered risk factors for its development [2–4]. Clinically, patients can present with symptoms of abdominal pain, nausea, vomiting, repeated bowel obstruction, blood-stained effluent and loss of ultrafiltration capacity. The diagnosis of EPS is based on clinical symptoms in combination with pathological findings and abdominal imaging [5].

Recently, Stuart *et al.* [6] described all imaging techniques applied in characterizing various complications of PD. Several imaging techniques have been used over the past decades to diagnose EPS. Case reports, case series and some larger studies have been published over the years. An increased awareness of computed tomography (CT) as imaging modality for diagnosing EPS has developed [7]. In a recent paper that reviewed the clinical significance and implications of EPS, imaging modalities were described in short and CT scanning was suggested as the investigation of choice in patients with established EPS [8].

The present review focusses on all imaging modalities specifically used to diagnose EPS, nowadays and in the past, and discusses their features, qualities and shortcomings. It is limited to diagnostic techniques for EPS secondary to PD. The Medline database was searched for relevant reports and studies on imaging modalities to diagnose EPS. The separate imaging modalities were entered in combination with 'peritoneal sclerosis', 'EPS', 'sclerosing peritonitis' and 'PD' as search terms. We restricted the language of our search to English.

Abdominal radiography

Plain abdominal radiography can show air–fluid levels and signs of bowel dilation, indicating obstruction [9–11]. Another common feature is the presence of peritoneal calcification [6, 12–15]. However, plain abdominal X-ray films can appear normal even though EPS is present [12, 16]. No data on sensitivity and specificity of plain abdominal films are available. Although it is readily available and helpful in establishing bowel obstruction and peritoneal calcifications, it does not provide conclusive or sometimes not even additional information on the presence or absence of EPS; therefore, we conclude that when EPS is suspected, an abdominal X-ray has no additional diagnostic value in diagnosing EPS.

Ultrasonography

Ultrosonography (US) has been used in the past when EPS was suspected. US characteristics of EPS are best appreciated with peritoneal fluid *in situ*. In one study, US findings of 14 EPS patients were reviewed [16]. Abnormal small bowel activity was present in 12 patients, tethering of bowel to the posterior abdominal wall in 10, intraperitoneal echogenic strands in 7 and membrane formation in 5. In another study by Krestin *et al.* [11] disturbed motility during real time, US was observed in all 13 patients, signs of intestinal obstruction

in 9 and bowel wall thickening in 5. Campbell *et al.* reviewed US images of five patients that died from EPS, four patients with EPS suspicion and six patients considered to be at an increased risk for EPS due to prolonged PD therapy. They found a characteristic appearance in several patients consisting of an echogenic membrane in the bowel wall [12]. Calcifications can also be detected with US [11, 15]. US is non-invasive and has no radiation burden. A major limitation is that the interpretation of the images is very dependent on the radiologist. There are no data on sensitivity, specificity and reproducibility.

Computed tomography

The use of CT in EPS diagnostics was introduced in 1988 [17]. Abdominal CT scans of two patients with a clinical suspicion of EPS revealed loculated ascites, adherent bowel loops, narrowing of bowel lumen and a thickened peritoneum. Several other case reports described similar CT findings and other features such as bowel dilation [9, 18] and the presence of peritoneal calcification [13–15, 18, 19]. Krestin *et al.* described CT findings in nine EPS patients. In all cases, signs of disturbed motility indicated by dilated bowel loops and air–fluid levels were seen and in half of the cases, loculated fluid and contrast-enhanced thickening of the peritoneum were present [11]. Campbell *et al.* [12] also reviewed CT scans of five EPS patients, four patients with EPS suspicion and six patients considered to be at an increased risk for EPS and found peritoneal thickening and calcifications in some cases.

Three studies compared CT scans of EPS patients to those of other PD patients. In the first study, CT findings of 10 EPS patients were compared to those of 71 control PD patients [20]. Peritoneal calcifications, peritoneal thickening, fluid loculation and tethering of small bowel loops were considered diagnostic for EPS. In the second study, abdominopelvic CT scans of 27 patients with EPS were compared to CT scans of 15 hemodialysis and 20 PD patients by using a severity scoring system [21]. Scoring parameters included peritoneal calcification and thickening, bowel wall thickening, bowel tethering and dilation and fluid loculation. A highly significant difference was found between total CT scan scores of EPS patients and scores of controls. The clinical outcome of EPS patients varied and the total CT scan score did not show a correlation with this outcome, making this score unsuited for predicting the clinical course. The authors also showed that CT scans could not be used for screening purposes because EPS patients had only mild abnormalities in 9 of 13 cases on CT scans that were performed >4 months before the diagnosis.

In the third study, performed by our own group, CT findings characteristic for EPS were investigated. We studied 15 EPS patients and 16 long-term PD control patients [22]. We found that contrast-enhanced CT had a sensitivity of 100% and a specificity of 94% for diagnosing EPS when experienced radiologists applied a combination of specific CT findings. A cut-off point for a positive test was set at positively scoring three of the six following items: peritoneal enhancement, thickening and calcifications; adhesions of bowel loops; signs of bowel obstruction and fluid loculation/septation. A representative example of a CT scan of an EPS patient is shown in Figure 1. Diagnosis of EPS is based on clinical features of intestinal obstruction accompanied by radiological imaging of bowel encapsulation [23]. This means that only these patients are labelled as having EPS in the described studies and that subsequently, less severe cases are not taken into account. The value of CT scanning in this last group of patients has not been evaluated in these studies and one could speculate that its value is much less.

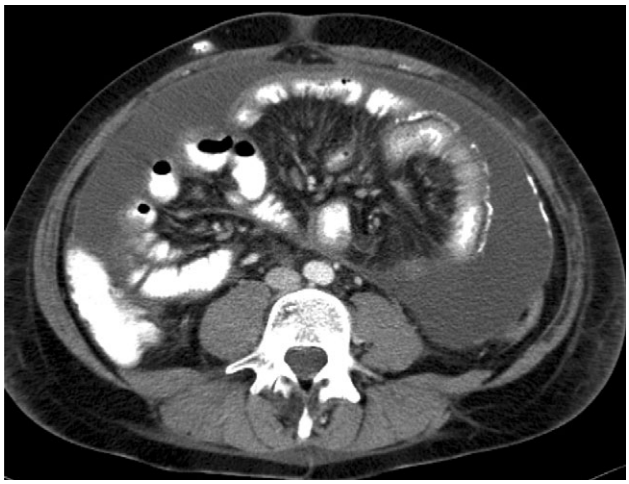

Fig. 1. An example of a computed tomographic scan of a patient with EPS. It shows ascites and bowel loops that are drawn into the centre of the abdominal cavity indicating adhesions and an enhanced thickened peritoneum with calcifications both visceral and parietal.

CT peritoneography, a technique in which a CT scan is combined with peritoneal contrast medium inserted through the peritoneal catheter, can demonstrate scar tissue and pathological peritoneal recesses [24]. However, calcifications can be overlooked because they can be obscured by high attenuation of contrast medium [25]. It might be valuable to evaluate the presence of EPS with this technique but to our knowledge, no studies have been published.

Major advantages of CT are that it is well tolerated by patients and readily available in most hospitals. Shortcomings of CT are radiation burden and risk of loss of residual renal function due to contrast-induced nephropathy. Despite these shortcomings, it is considered a safe technique. When used in the right clinical setting in symptomatic patients, danger of radiation exposure of CT in general is outweighed by the medical need and beneficial effect of an accurate diagnosis [26]. To prevent contrast-induced nephropathy, patients should be well hydrated before, during and after the procedure. Although the incidence is relatively low in well-hydrated patients, the risk is increased in patients with a severely decreased kidney function. In a recent study, 7 of 58 patients with a residual renal function of <30 mL/min developed contrast-induced nephropathy [27]. If a long-term PD patient has no residual renal function anymore, it is of no concern. In any other case, CT without contrast enhancement should be considered.

Magnetic resonance Imaging

Two case reports described magnetic resonance imaging (MRI) findings in EPS patients. Small bowel distension

and circumscribed focal wall thickening were described in one patient [28] and massive lobulated ascites in the omentum with wall enhancement of the lobulated ascites and compression of the bowel in another [29]. An advantage of MRI is that there is no radiation burden. Magnetic resonance (MR) peritoneograhy has been used to detect complications of PD [30, 31] but to our knowledge, it has never been used for the purpose of diagnosing EPS. Gadolinium-containing MR contrast media are associated with nephrogenic systemic fibrosis and should therefore be avoided in patients with renal failure [32–34]. Also, MRI is a time consuming and rather costly technique, which is not yet as available as CT, making widespread use less appealing.

Colon transit studies

Follow-through examinations of small and large bowels have been performed in EPS patients. In one case, a small bowel follow-through revealed bowel wall thickening of a distal jejunal loop followed by a 'cauliflower-like' formation of ileum loops [18]. In another case, small bowel follow-through with barium showed bowel dilation and encapsulated loops [10]. In the study by Krestin *et al.* [11], an upper gastrointestinal follow-through examination was performed with barium in three patients and water-soluble Diatrizoate in five before surgical intervention took place. All cases demonstrated a delayed transit time but no clear evidence of compressing intraperitoneal bands was present. In the study by Campbell *et al.* [12], all 10 living patients underwent a colon transit study. They swallowed capsules containing radiopaque markers on three successive days. On Day 4, a plain abdominal film was made and the amount of markers was counted. Four patients had increased numbers of colonic markers indicating significantly slowed colonic motility.

Follow-through examinations can provide information on bowel function and may be helpful in locating the obstruction site. However, they are invasive, time consuming and require preparations that could interfere with fluid restrictions of dialysis patients. Nowadays, they are less frequently used in clinical practice.

Imaging techniques using radioactivity

The usefulness of fluordeoxyglucose positron emission tomography (PET) in diagnosing EPS was studied in three EPS patients and five asymptomatic long-term PD patients [35]. For this technique, radioactively labelled tracer was administered intravenously; thereafter, a PET scan was done. The authors showed that this technique detects the inflammatory phase, if present, of 'sclerosing peritonitis' because of an increased tracer uptake in the peritoneum. However, a positive scan could also occur as a result of an acute peritonitis; therefore, the clinical presentation should be taken into account in its interpretation. Recently, a case report was described in which radiolabeled dialysate was inserted in the peritoneal cavity after which a peritoneal scintigraphy was performed because peritoneal adhesions were suspected [36]. Non-uniform distribution of the dialysate in combination with loculated tracer accumulation confirmed the presence of adhesions. It might be possible that that this technique could be effective in detecting EPS but no studies have been published. An obvious disadvantage is the use of radioactive material for these modalities.

Conclusion

EPS is a rare but life-threatening complication of long-term PD. Biomarkers in peritoneal effluent have a potential role in early diagnosing EPS [37] but, until now, no imaging screening methods are available. Accurate imaging techniques for diagnosing this severe disease are of great importance. A variety of imaging techniques, invasive as well as and non-invasive, have been used and studied to diagnose EPS. In this review, we have provided an overview of these modalities and discussed their specific findings, advantages and limitations.

CT is the most frequently studied imaging technique for diagnosing EPS. It is the only technique that has been investigated in case–control designs [20–22] and for which data on sensitivity and specificity are available [22]. Although we have discussed several shortcomings, CT has been shown to accurately diagnose EPS. We advocate that CT with contrast enhancement should be the modality of first choice when EPS is suspected. Evaluation of the CT scans should preferably be performed by experienced radiologists with knowledge of PD and EPS. In conclusion, CT is the definitive imaging modality for EPS at the present time. However, it should be noted that data of other imaging techniques, such as MRI, are lacking. Due to the shortage of publications, drawing certain conclusions remains difficult. Studies comparing different imaging modalities with one and other in patients with and without EPS should be conducted to solve this issue.

Conflict of interest statement. None declared.

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