Case:
A 32 year old woman with CKD V presented to the ED with 3 days of abdominal pain. She was diagnosed with Pseudomonas aeruginosa peritonitis; her peritoneal dialysis (PD) catheter was removed and Ceftriaxone was started.

PMH:
CKD V secondary to IgA nephropathy on peritoneal dialysis for 12 years; previous hospitalizations for peritonitis; pulmonary embolism; hemorrhagic ovarian cysts.

Hospital Course:
She developed leukocytosis despite treatment with Ceftriaxone and Ciprofloxacin. Despite this she continued to have severe abdominal pain, poor appetite, nausea and vomiting. Lab work showed leukocytosis. CT abdomen and pelvis: multiple focal peritoneal thickening and partial small bowel obstruction due to peritoneal fibrous narrowing and focal thickening with partial small bowel entrapment. Instead of infection, imaging showed evidence of peritoneal thickening and bowel entrapment.

Physical Exam:
Vitals: stable. Significant for diffuse abdominal tenderness to mild palpation. No rebound or guarding, no palpable masses, and bowel sounds positive.

Labs:
peak WBC of 42.7 (Figure 1)

Imaging:
CT abdomen and pelvis: multiple focal sites of narrowing and focal thickening with partial small bowel obstruction due to peritoneal fibrous rind/adesions. Multiple calcifications with an eggshell-coating pattern on multiple loops of small bowel. (Figure 2)

Discussion:
Encapsulating peritoneal sclerosis is a rare syndrome that encompasses radiographic images in the presence of clinical symptoms (such as those that occur with small bowel obstruction), and may also include typical pathology findings and membrane transport characteristics. Rarely diagnosed in the US, it is often associated with long-standing peritoneal dialysis (Figure 3) but can also occur in the absence of PD.

Pathogenesis:
While not yet completely understood, the prevailing theory is the two-hit hypothesis. The initial “hit” describes the changes that occur in the peritoneum. The “second hit” can then lead to progression of the underlying changes to EPS. In this patient, the first hit may have been the long-standing PD with the second hit being one of multiple factors such as peritonitis, PD catheter removal, or C. difficile colitis.

Treatment:
This is somewhat controversial due to limited data. Medical treatment usually consists of immunosuppressants and if necessary, nutritional support. Additionally, surgical options such as enterolysis may be considered. While this patient had initial improvement with steroids, whether this is maintained in the future remains to be seen.

Prognosis:
Patients typically have a poor prognosis with reported mortality rates of up to 50% (Figure 4). Although the data is somewhat variable between studies, morbidity and mortality rates appear to be associated with the length of time patients have been on peritoneal dialysis. Following the diagnosis of EPS, our patient was hospitalized multiple times for abdominal pain, partial small bowel obstruction, and CVA.

References: