Encapsulating peritoneal sclerosis in hemodialysis patient: a case report

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ABSTRACT

Background: Encapsulating peritoneal sclerosis is a rare manifestation nowadays.

Case Report: This was a case report of a young female with ESKD on hemodialysis for more than 6 months with a previous history of continuous ambulatory peritoneal dialysis (CAPD) and recurrent peritonitis which currently presented with painful abdominal distension. The CT scan of the abdomen showed a clumping of small bowel loops at the left side encased into a thin fibrocartilaginous membrane. Initially, she was treated for intrabdominal sepsis; however, after recurrent admission, intravenous Methylprednisolone was started, followed by high-dose steroids and tamoxifen as a treatment for encapsulating peritoneal sclerosis (EPS). Surprisingly, she improved and had a good clinical response. She was readmitted again after a few months with abdominal pain due to an abrupt stop in steroid but improved again after the steroid was started.

Conclusion: Abdominal pain in a patient with a history of CAPD peritonitis needs to raise suspicion of EPS even though is rare. Radiological Imaging is helpful in making early diagnosis. The most important thing is steroids as a mainstay of the treatment of EPS besides tamoxifen with a minimal 1-year duration or lifelong as surgical intervention is very risky.

Keywords: Encapsulating peritoneal sclerosis, peritonitis, clumping of bowel loops, tamoxifen, steroid.

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Background

Encapsulating peritoneal sclerosis (EPS) occurs due to the thickening of the fibrocartilagenous peritoneal membrane which encloses the bowel leading to intestinal obstruction and malnutrition.

This is commonly seen in patients with longstanding peritoneal dialysis as dialysate can cause inflammation and thickened fibrosis of the peritoneal membrane. Currently, there are no laboratory tests used to diagnose EPS. The diagnostic tools are mainly CT Imaging.

Steroids with the combination of tamoxifen are used to reduce the degree of inflammation and halt the progression into fibrous tissue.

This case demonstrates a 22-year-old woman with a history of peritoneal dialysis which is complicated by multiple episodes of peritonitis causing her conversion into hemodialysis. She presented with abdominal pain. CT abdomen revealed a clumping of small bowel loops (SBLs) at the left side of the abdomen and encased into a thin fibrocartilaginous membrane with surrounding free fluid. She was treated medically with steroids without surgical intervention as she improved clinically during admission.

Case Presentation

A 22-year-old lady with nephrotic syndrome secondary to focal segmental glomerulosclerosis, diagnosed at the age of 15, presented with generalized body swelling for 3 years treated with prednisolone and mycophenolate mofetil (MMF) for a total of 2 years and 6 months. However, her kidney function continued to deteriorate and she was started on kidney replacement therapy, which was peritoneal dialysis due to a failed immunosuppressant. 2 months after the Tenchkoff catheter was inserted, noted bloody discharge aspirated from the lumen itself. Laparoscopic readjustment was done and Tenchkoff catheter was adhered to the omentum; the tip of Tenchkoff was located at the pelvic region with blood clot obstructing the distal catheter with good inflow and outflow after removal. Peritoneal dialysis was continued, however, complicated with multiple episodes of peritonitis with *Staphylococcus aureus*, *Pseudomonas aeroginosa*, and *Streptococcus beta-hemolytic* Group G including fungal in origin (*Chaetomium globosum*). Subsequently, her Tenckoff catheter was removed due to infection, and she was converted into hemodialysis. 6 months later, while on

hemodialysis with fistula, she presented with sudden onset abdominal pain, pricking in nature, radiating to the back associated with nausea and vomiting causing her poor oral intake. Her pain score was 7/10. Otherwise, she was able to pass flatus and motion before admission.

Upon physical examination, a soft but tender abdomen over the left iliac fossa with a palpable mass measuring 5 × 5 cm, firm but there was no guarding noted. Lung and cardiovascular examinations were unremarkable.

She was not in sepsis, with a blood pressure of 186/108 mmHg and a heart rate of 82 bpm. Afebrile with temperature 36.8°C and oxygen saturation 100% under room air.

Differential diagnosis

1. Spontaneous bacterial peritonitis (SBP)-in view she has a background history of peritoneal dialysis, and SBP is very unlikely.
2. Intestinal obstruction secondary to bowel malignancy: absence of risk factor for gastrointestinal malignancy and her previous peritoneal dialysis renders this unlikely.
3. Irritable bowel syndrome
4. Twisted ovarian cyst

Investigation

Her blood investigation normocytic normochromic anemia with Hb 8.2 g/dl (NR: F 11.6-15.1, M 13.5-17.4) with normal white cell count and platelet 6.7 × 10^9/l (NR: F 3.4-10.0, M 3.8-9.7) and 247 × 10^9/l (NR 158-410), respectively.

Her renal function showed derangement consistent with her background of ESKD with Na 138 mmol/l (NR 135-145), K 4.5 mmol/l (NR 3.5-5.0), urea 17 umol/l (1.7-8.3), creatinine 726 umol/l (NR 70-130), Se Albumin 46 g/l (NR 38-44), Se Calcium 2.56 mmol/l (NR 2.15-2.55), and Se Phosphate 2.46 mmol/l (NR 0.87-1.45). Inflammatory markers were raised with erythrocyte sedimentation rate 91 and C-reactive protein 42.

Her liver function was normal. Blood culture showed no growth.

The chest radiograph (Figure 1) showed no air under the diaphragm.

Serial abdominal X-rays were done throughout her admission (Figure 2). Noted large, impacted stool (Figure 2a and b), however, reducing once the patient is able to pass out during admission inward (Figure 2c).

Bedside ultrasonography was done upon her presentation revealing a mass measuring 13 × 7 cm with peristaltic movement within the mass. Their left ovary size is 2.5 × 1.7 cm and her right ovary is 3 × 4 cm with a follicular cyst seen measuring 2 × 2 cm and free fluid was present.

Figure 3 CT abdomen showed clumping of SBLs at the left side of the abdomen and encased in a thin fibrocartilaginous membrane. Short segment dilatation of the small bowel measures up to 3.1 cm in diameter with small bowel feces signs (yellow arrow) noted at the left lower quadrant of the abdomen. The clumped bowels are causing displacement of the descending colon posterolaterally and it also causing mass effect to the left psoas muscle posteriorly. The findings likely represent sclerosing encapsulating peritonitis.

Treatment

She was admitted to the ward by the surgical team and kept nil by mouth to monitor her progress. All her electrolyte imbalance was corrected to minimize her symptoms. Initially, she was treated for Intraabdominal sepsis and started with intravenous (IV) Cefoperazone 1 g BD and IV Metronidazole 500 mg TDS for 4 days followed by oral antibiotic at home. However, 3 days later she presented again with similar complaints but more severe ones and was admitted to the general medical ward. She required a painkiller controlled with PCA Fentanyl. This time antibiotic was upgraded to IV Meropenem and she was started with IV Methylprednisolone 500 mg OD and planned for tamoxifen 10 mg OD. The tamoxifen was served late after she was able to tolerate it orally because there was no IV tamoxifen available at our center. She was closely managed and monitored along with the nephrologist and surgical team. She was treated conservatively for her acute abdomen as she improved throughout admission.

Outcome and follow-up

She was markedly improved after starting with IV Methylprednisolone 500 mg OD for 3 days followed by oral high-dose prednisolone (1 mg/kg/day). No more usage of painkillers, able to tolerate orally without vomiting or abdominal pain. She was discharged well and continued with prednisolone and tamoxifen.

5 months later, she presented again with abdominal pain and vomiting and required IV Fentanyl as a painkiller. Further history revealed that she was not on prednisolone

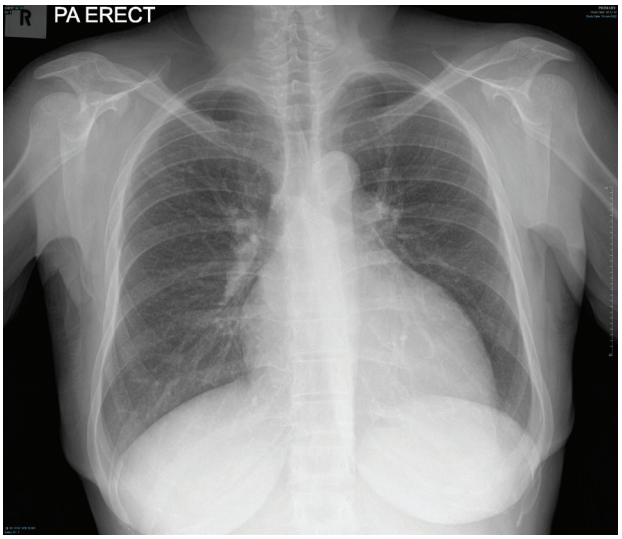
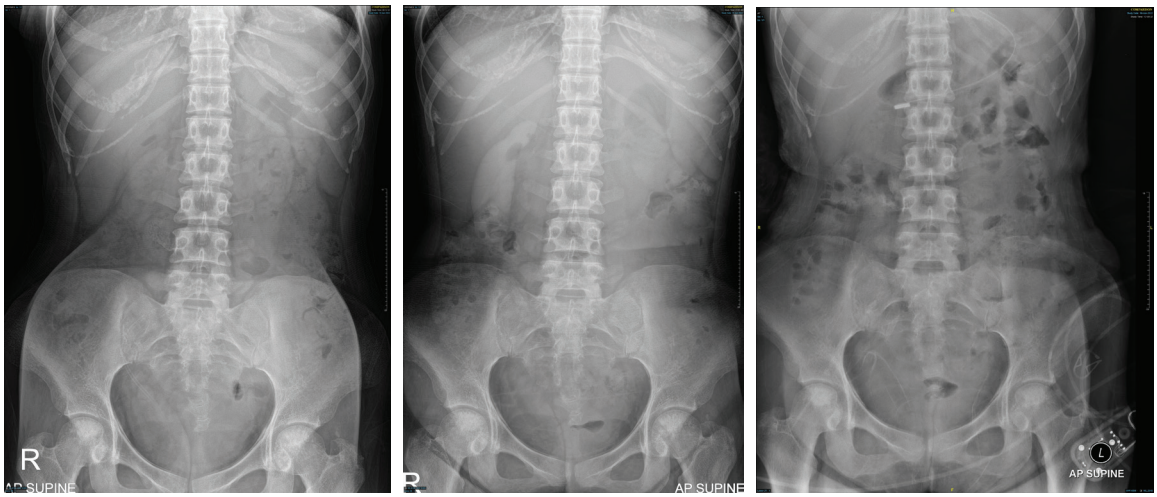
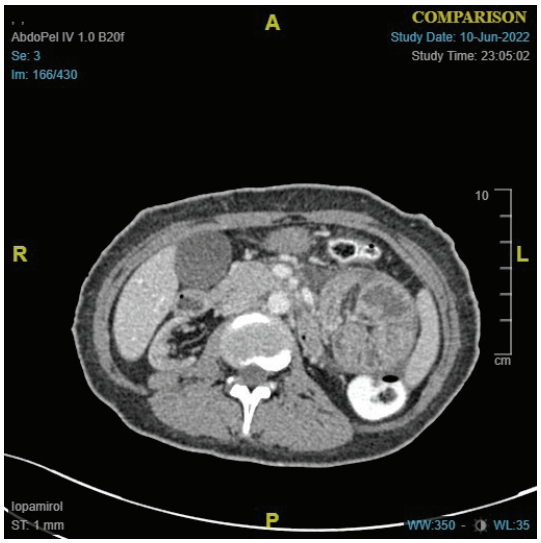


Figure 1. *Chest-ray. Chest radiograph showed no air under the diaphragm.*

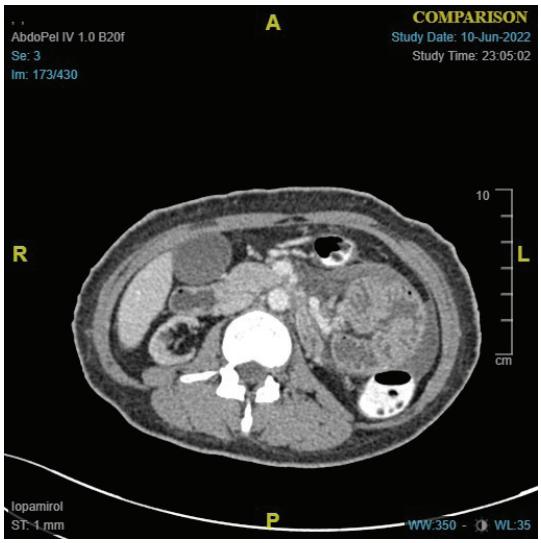


(a) (b) (c)

Figure 2. *Abdomen X-ray. Serial abdominal X-ray were done throughout her admission. No evidence of any dilated bowel to give a clue for a dilated bowel. Noted large impacted stool (a and b), however, reducing once patient able to pass motion during admission inward (c).*



(a)



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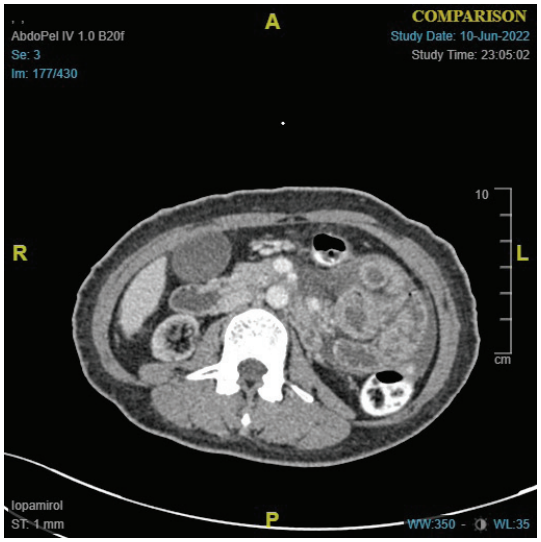

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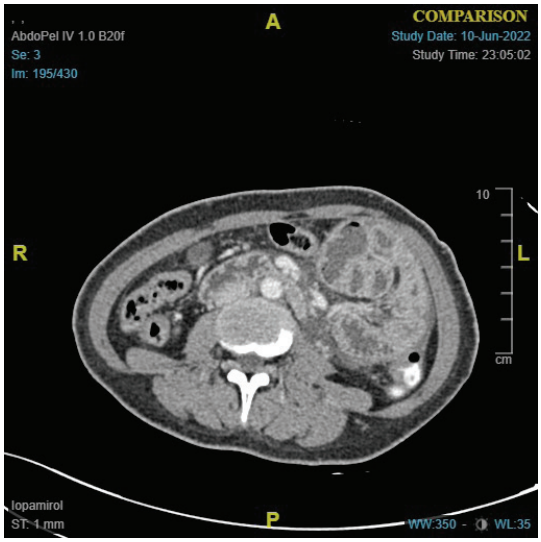

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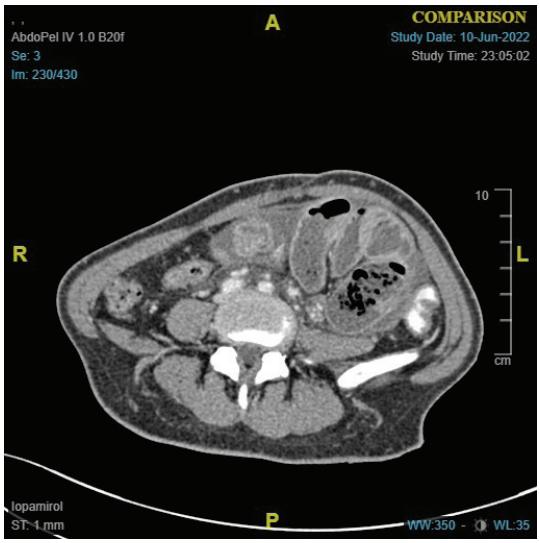

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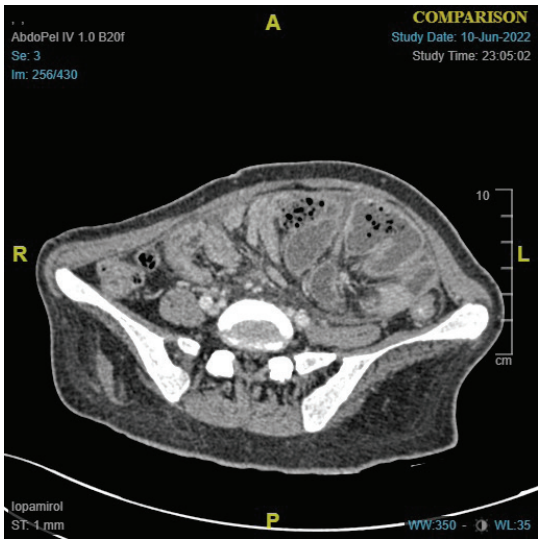

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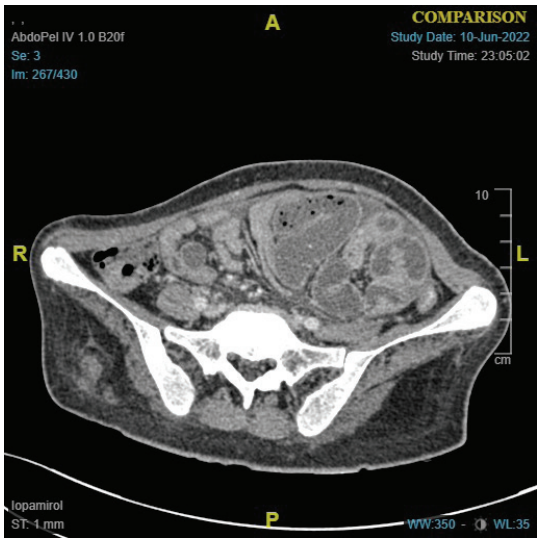

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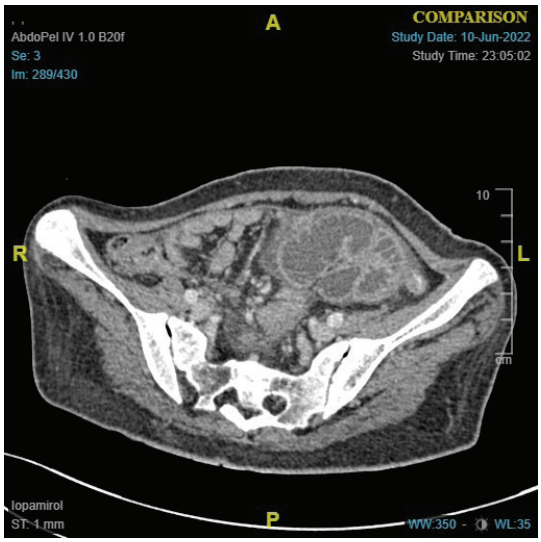

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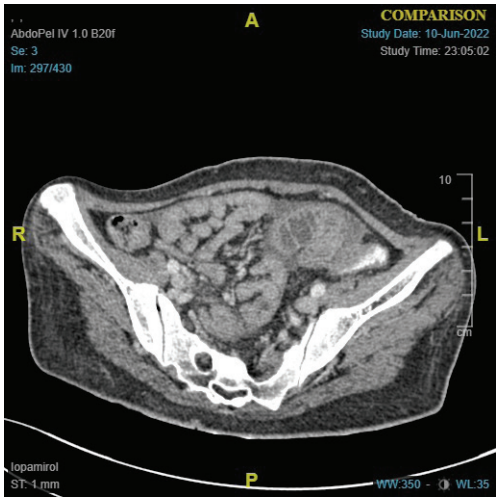

(i)

Figure 3. *CT abdomen. CT abdomen showed clumping of SBLs (a-i) at the left side of the abdomen and encased in a thin fibrocartilaginous membrane short segment dilatation of small bowel measures up to 3 cm in diameter (e) with small bowel feces signs (yellow arrow) (e-g) noted at the left lower quadrant of the abdomen. The small bowel feces sign indicates that there is a small bowel obstruction. The clumped bowels are causing displacement of the descending colon posterolaterally and it also causes the mass effect to the left psoas muscle posteriorly. The findings likely represent sclerosing encapsulating peritonitis with short segments. SBL: small bowel loops; L: liver; S:spleen; Yellow arrow : small faeces sign.*

for the past 1 month but was still on tamoxifen. As usual, after starting IV hydrocortisone, her symptoms resolved, and she was discharged with high dose prednisolone 1 mg/kg/day and planned to taper down the dose and complete treatment at least for 1 year.

Currently, her symptoms are improving but occasionally has episodes of abdominal pain but only minimal, and has good bowel habits. It does not cause her to take painkillers or interrupt her dialysis session with no intradialytic complication. Even though initially on a high dose of steroid, she did not develop the side effects from it.

Discussion

EPS is commonly seen as a complication associated with peritoneal dialysis. EPS is characterized by a thickened fibrocartilaginous peritoneal membrane enclosing the bowel leading to bowel obstruction [1]. EPS carries high mortality rates of approximately 50% after 1 year of diagnosis [2].

EPS is commonly seen in those with peritoneal dialysis. The risk of EPS is directly proportionate with the duration of peritoneal dialysis [2]. Peritoneal dialysate produces glucose degradation product which incites inflammation in the peritoneum. For end-stage renal disease patients, chronic inflammatory conditions increase endothelial permeability, and induce fibrogenesis leading to fibrin deposition and fibrocollagenous membrane formation causing acute obstruction as well as malnutrition [1].

EPS also occurs due to other inflammation conditions such as autoimmune disease, sarcoidosis, peritoneal and bowel malignancies, chronic ascites and gynecology causes such as endometriosis.

Drugs that can potentially cause EPS include calcineurin inhibitors and also beta blockers [1].

Diagnosis of EPS is based on clinical presentation with imaging and laparotomy findings. The most sensitive and specific in the diagnosis of EPS is the identification of peritoneal thickening enclosing the bowel via laparotomy or laparoscopic method. CT scan is the method of choice, which may show a SBL encased in a thick fibrotic membrane. Loculated ascites, increased mesenteric fat density and localized or widespread peritoneal calcification are other radiographic findings that may present in EPS [1].

Treatment of EPS includes pharmacological therapy with immunosuppressants, and surgical intervention is only recommended in the event of failure of medical therapy. Surgery is not preferable except at the center with experience as technically difficult and carries the risk of bowel injury and high output stoma [3].

To date, steroids and tamoxifen are the only agents used and clinically proven agents in EPS treatment. There was a lack of data regarding the use of MMF and Azathioprine in EPS [4]. Glucocorticoids are used in EPS to prevent the progression of peritoneal thickening and fibrosis via anti-inflammatory and immunosuppressive effects. Tamoxifen is a selective estrogen receptor modulator an anti-fibrotic agent that inhibits transforming growth factor beta, thus inhibiting the fibrosis process.

Conclusion

Diagnosis of EPS should be considered in dialysis patients with intestinal obstruction symptoms. Multidisciplinary management is essential in managing a team. There are no large RCT trials available regarding immunosuppressant use in EPS. Therefore, RCT is needed.

What is new?

Regarding EPS, it is known that the risk factors are young age, history of peritonitis, and diagnosis by CT scan, but the difference is that it is developed after this patient already has 6 months of hemodialysis after changing dialysis modalities; second, her first presentation is same as that of with intraabdominal sepsis features but lead to recurrent admission which later improved after starting steroid and tamoxifen. Third, not to taper down steroids very quickly and find the EPS features in CT scans which can be varied in other patients.

List of Abbreviations

EPS	Encapsulating peritoneal sclerosis
MMF	Mycophenolate mofetil
SBP	Spontaneous bacterial peritonitis

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Funding

None.

Consent for publication

A written informed consent to publish this case was obtained from the patient herself.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient**	A 22-year-old woman
2	**Final diagnosis**	EPS
3	**Symptoms**	Abdominal pain
4	**Medications**	IV Methylprednisolone, High dose steroid, tamoxifen
5	**Clinical procedure**	Chest radiograph, abdominal X-ray, CT abdomen
6	**Specialty**	Nephrology, Radiology