CASE REPORT OLGU SUNUMU

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A Case of Sclerosing Encapsulated Peritonitis Secondary to Familial Mediterranean Fever, Presenting with Recurrent Ileus Attacks

Tekrarlayan İleus Atakları ile Gelen Ailesel Akdeniz Ateşine Sekonder Bir Sklerozan Enkapsüle Peritonit Vakası

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ABSTRACT Sclerosing encapsulated peritonitis (SEP) is a rare condition characterized by acute and/or subacute intestinal obstruction caused by the intestines being encased in a fibrocollagenous cocoon-like sac. This condition often arises from either idiopathic or secondary causes, with secondary causes being more prevalent. familial Mediterranean fever (FMF) is one such secondary cause. In SEP, patients may experience symptoms such as acute ileus, while some may remain asymptomatic. This report details a case of SEP associated with FMF, in which the patient experienced recurrent ileus attacks over a six-month period, accompanied by severe abdominal pain, nausea, vomiting, restricted oral intake, and weight loss. The condition responded well to treatment with corticosteroids and colchicine.

Keywords: Ileus; abdominal pain; peritonitis; sclerosis; familial Mediterranean fever

ÖZET Sklerozan enkapsüle peritonit (SEP) bağırsağın fibrokolajenöz koza benzeri bir kese ile kaplanması sonucu, akut ve/veya subakut bağırsak obstrüksiyonu kliniği ile ortaya çıkan nadir görülen bir durumdur. Çoğunlukla idiyopatik ya da sekonder nedenlere bağlı gelişir. Sekonder nedenler daha yaygın görülür ve ailesel Akdeniz ateşi (AAA) bu nedenlerden biridir. SEP'te semptomlar sıklıkla akut ileus şeklinde ortaya çıkabileceği gibi bazı hastalar da asemptomatik olabilir. Bu yazıda; 6 aydır tekrarlayan ileus atakları, şiddetli karın ağrısı, bulantı kusma, oral alım kısıtlılığı ve kilo kaybı semptomları ile başvuran SEP tanısı ile birlikte AAA tanısı alan ve kortikosteroid ve kolşisin tedavisi ile yanıt alınan 41 yaşında erkek bir olgu sunulmaktadır.

Anahtar Kelimeler: İleus; karın ağrısı; peritonit; skleroz; ailesel Akdeniz ateşi

Sclerosing encapsulated peritonitis (SEP) is a rare condition of unknown cause, characterized by the complete or partial encasement of the small intestine in a fibro-collagenous sac resembling a cocoon.[1] SEP can be classified as either primary (idiopathic) or secondary based on its etiopathogenesis and the pathological features of the membrane surrounding the bowel.[1,2] The idiopathic form of SEP is extremely rare, while the secondary form is more common.[3] Several factors may lead to secondary SEP, including continuous ambulatory peritoneal dialysis, the presence of a LeVeen shunt, previous abdominal surgery, retrograde menstruation, systemic lupus erythematosus, sarcoidosis, familial Mediterranean fever (FMF), tuberculosis, and the use of beta-blockers.[4] The primary (idiopathic) form of SEP is also known as abdominal cocoon syndrome. This form is most frequently observed in adolescent fe-

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males and retrograde menstruation accompanied by viral infection is hypothesized as a potential cause. However, this hypothesis has yet to be confirmed since this condition can also occur in male patients.[5]

Clinically, patients may experience recurrent abdominal pain, nausea, vomiting, anorexia, weight loss, and malnutrition due to episodes of acute, subacute, or chronic ileus or subileus. Some patients may remain asymptomatic.[1] The clinical diagnosis of SEP can be challenging in the early stages because its symptoms are not specific. As a result, radiological imaging plays a crucial role in the diagnosis.[1,2] Symptoms often present as acute ileus, which is frequently diagnosed only during surgery.[1,2]

Treatment for SEP is typically conservative in the early stages, although surgical intervention may be necessary in more advanced cases. In this article, we present a case of a patient diagnosed with SEP during evaluations for recurrent abdominal pain and ileus attacks, who was later found to have FMF as the underlying cause of SEP.

CASE REPORT

A 41-year-old male patient had been experiencing recurrent abdominal pain for 10 years. He was hospitalized with a preliminary diagnosis of ileus four times in the last six months, each time being discharged after conservative treatment. The general surgery clinic performed a diagnostic laparoscopy, suspecting ileocecal valve stenosis and ileus. During the operation, abundant ascites fluid, multiple adhesions and thickening of the peritoneum were observed in the abdomen. During the intraoperative frozen section analysis, an increase in connective tissue was noted in the peritoneum. The patient was admitted to our gastroenterology clinic for investigation of his condition. He had no known comorbidities but presented with a history of recurrent ileus attacks over the past six months. The patient reported experiencing severe abdominal pain that significantly impacted his quality of life and frequently disturbed his sleep. Additionally, he experienced nausea, vomiting, and a weight loss exceeding 10 kg during this period.

During the physical examination, there was widespread tenderness and abdominal distension. However, no defence or rebound was noted. A soft, palpable mass approximately 15 cm in diameter was found around the umbilicus near the left lower quadrant. Laboratory tests, including biochemistry and complete blood count, indicated normal values. During a colonoscopy conducted due to suspicion of Crohn's disease, the proximal part of the sigmoid colon could not be traversed because of altered anatomy likely caused by intra-abdominal fibrotic changes. The mucosa of the rectum and sigmoid colon appeared normal. A faecal calprotectin test showed a significantly elevated level of 894 mcg/g. Possible infectious processes and malignancy were ruled out. A computed tomography (CT) scan of the abdomen and pelvis revealed diffuse free fluid in the peritoneal cavity, thickening of the peritoneal surfaces, and wall thickening with enlargement in the distal jejunum and ileal intestinal loops. Cytological examination of the ascitic fluid sample collected during the procedure indicated nonspecific inflammation and mesothelial proliferation. A peritoneal biopsy showed evidence of fibrosis and chronic inflammation, with no amyloid staining present.

The following findings were observed in the MR enterography conducted on the patient: widespread ascites with a tendency to encapsulate, diffuse thickening and contrast enhancement in the peritoneum, and a peritoneal band compressing the terminal ileum. There was also widening of up to 5 cm in the bowel loops proximal to the compression, diffuse wall thickening in the small bowel loops, and findings compatible with SEP (Figure 1). It is also pertinent to note that two of the patient's siblings have been diagnosed with FMF. FMF was also included in the preliminary diagnosis of the patient with a family history. And heterozygous positivity for the M680I and V726A mutations was detected in the FMF gene.

Our patient had peritonitis as a major criteria for FMF and abdominal pain as a minor criteria. Additional supporting criteria included a family history of the condition, severe pain that required bed rest, episodes of spontaneous remission, asymptomatic intervals between attacks, negative results from a laparotomy and a history of consanguineous marriage. Based on the clinical and laboratory findings, the patient was evaluated as FMF and diagnosed with SEP

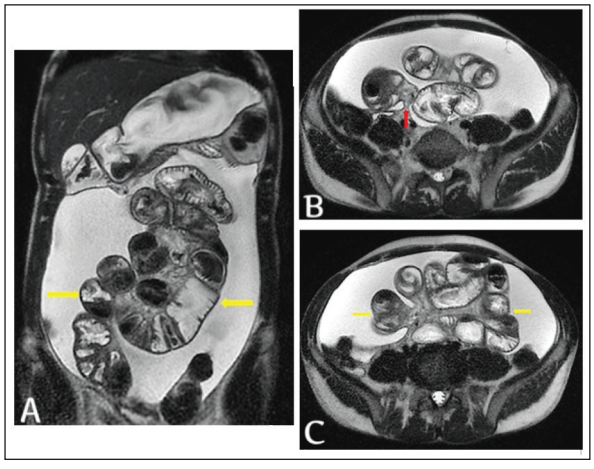

FIGURE 1: Coronal **(A)** T2 and axial **(B, C)** T2 weighted magnetic resonance imaging images: it shows enlarged small bowel rings collected in a sac-like structure compatible with peritoneal encapsulation in the center of the abdominal cavity (yellow arrows). The possible cause of the blockage is a thickened peritoneal band, which causes narrowing at the level of the terminal ileum and enlargement of the small intestine ans proximal to this level (red arrows). Ascites is also present in the abdomen.

due to a delayed diagnosis and lack of treatment for FMF. The patient was started on colchicine treatment. Surgical intervention was not considered because no therapeutic procedures could be performed in the previous surgery.

Methylprednisolone was initiated at a dose of 0.5 mg/kg/day for the first month. The dosage was planned to be reduced by 8 mg each month while continuing steroid treatment. During monthly follow-up visits at the outpatient clinic, the patient's abdominal pain, nausea and vomiting significantly decreased. Additionally, their bowel habits returned to normal and improvements in oral intake and weight gain were observed.

Informed consent for this study was obtained from the patient.

DISCUSSION

SEP was first described in 1907 by Owtschinnikow, who referred to it as "encapsulation of peritonitis chronic fibrosis".[6] SEP is primarily reported in the literature as a condition occurring in young women.[7] However, a review of 118 cases indicated that SEP is actually twice as common in men as in women.[8]

Detecting and diagnosing SEP can be challenging without the use of radiological imaging techniques. Flat abdominal radiography, ultrasonography, and CT may reveal dilated segments of the intestines. A characteristic finding of SEP is the presence of a thin sac-like membrane surrounding the intestines, which can be identified using CT and magnetic resonance imaging.

The treatment for SEP should be tailored to each patient based on factors such as the prevalence, whether it is a primary or secondary occurrence and the stage of the disease. A conservative approach is recommended for patients with mild gastrointestinal symptoms as the first-line treatment. In these cases, providing nutritional support and addressing any underlying diseases (in secondary cases) are important components of care.

Immunosuppressants and antifibrotics that target the underlying disease mechanism may be used for patients who do not respond to conservative treatment. The beneficial effects of immunosuppression on SEP were first observed in kidney transplant patients, who reported improvements in SEP symptoms following immunosuppressive therapy.[9] A variety of medications have been tested in this context, including corticosteroids, colchicine, azathioprine, cyclosporine, mycophenolate mofetil, and rapamycin (an mTOR inhibitor).[10,11]

Corticosteroids work by inhibiting collagen synthesis and maturation, thus suppressing inflammation within the peritoneal membrane. This action can reduce the thickening of the peritoneal membrane associated with the condition.[12] However, in patients with advanced peritoneal fibrosis, immunosuppressive therapy alone may not suffice.[13]

Tamoxifen, utilized as an antifibrotic in treating SEP, exerts its effects by inhibiting transforming growth factor β 1, a crucial cytokine involved in the fibrosis process.[13] Surgical intervention for SEP is advised only for patients experiencing severe ileus symptoms.[14]

The patient discussed in this study was initiated on corticosteroid and colchicine treatment, to which they had a very positive response. During follow-up, the patient's abdominal pain decreased, and weight gain was observed as their oral intake improved. No-

tably, the patient did not require any surgical procedures during the follow-up period.

In conclusion, SEP is a rare disease that manifests with nonspecific symptoms and can lead to disruption of the intestinal passage in advanced stages, making it difficult to diagnose. There are various treatment approaches available, including both medical and surgical options. However, this disease can be effectively controlled with early diagnosis and appropriate management.

Source of Finance

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Hatun Kaboğlu Bilgin, Emine Selin Yıldırım;* ***Design:*** *Hatun Kaboğlu Bilgin, Emine Selin Yıldırım;* ***Control/Supervision:*** *Ramazan Erdem Er;* ***Data Collection and/or Processing:*** *Ramazan Erdem Er, Hatun Kaboğlu Bilgin, Emine Selin Yıldırım;* ***Analysis and/or Interpretation:*** *Ramazan Erdem Er, Hatun Kaboğlu Bilgin, Emine Selin Yıldırım;* ***Literature Review:*** *Hatun Kaboğlu Bilgin, Emine Selin Yıldırım;* ***Writing the Article:*** *Emine Selin Yıldırım, Hatun Kaboğlu Bilgin;* ***Critical Review:*** *Ramazan Erdem Er;* ***References and Fundings:*** *Ramazan Erdem Er;* ***Materials:*** *Ramazan Erdem Er.*

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