A 50-year-old male was admitted to Firoozgar Hospital, affiliated with Tehran University of Medical Sciences, in October 2010 because of abdominal pain and vomiting for the previous three months. His pains usually begin one to two hours following a meal, and occasionally were followed by vomiting. Vomitus consisted of undigested food. The patient had an eight kilogram weight loss over the past two months. His past medical history was remarkable for diabetes mellitus and scrotal hernia surgery. Vital signs were normal and physical examination revealed an extended guarding. No organomegaly was detected. Laboratory tests were unremarkable. Abdominal sonography revealed a large mass in the abdominal cavity with echogenic septa. An upper GI endoscopy showed the presence of a small sliding hiatal hernia with grade II esophagitis. An abdominal computed tomography (CT) scan was performed (Figure 1).

What is your diagnosis?

See the next page for diagnosis.
Photoclinic Diagnosis:  
**Idiopathic Peritoneal Sclerosis or Abdominal Cocoon**

The CT scan (Figure 1) showed aggregation of the small bowel loops in the mid-abdomen along with increased density of the surrounding mesentery, which has formed an enhancing capsule around the aggregated bowel loops. There was also an apparent increase in small bowel wall thickness or its serosal coating. Possible diagnoses included either sclerosing encapsulated peritonitis (SEP) or an internal hernia with an obstructed venous return. The patient underwent abdominal surgery. An encased intestine in a membrane with multiple adhesions was found and released (Figure 2). Pathologic examination of the excised membrane (Figure 3) showed extensive collagen deposition and fibrosis which led to hemorrhage and infarction, however, no significant inflammation was observed.

Primary idiopathic peritoneal sclerosis (fibrosis) is a rare cause for small bowel obstruction. Its pathogenesis is poorly understood, but the most rational pathophysiologic explanation is peritonitis leading to sclerosis, membrane formation, and consequently to a cocoon abdomen. Abdominal cocoon (AC) of unknown cause has been described, mostly in young girls from tropical and subtropical regions. For these patients, suggested explanations include primary transvaginal peritonitis, retrograde menstruation, or an immunologic response to gynecologic infection, but it does not explain the occasional occurrence of AC in males. Additionally, no organisms were isolated in any of the previously reported cases, with the exception of one case associated with tuberculosis. As mentioned, secondary causes of SEP are included prolonged treatment with beta blockers, particularly practolol, sarcoidosis, and systemic lupus erythematosus. The above mentioned causes were not noted in our patient. Therefore, he most likely represented the idiopathic variety of this disease. He had experienced recurrent episodes of intestinal obstruction caused by compression of the intestines within the contracting cocoon, which has lasted for at least three months. Abdominal distension and weight loss were also prominent problems.

The treatment of choice is a simple release of the adhesions and the entrapped intestine. In some complicated cases bowel resection is mandatory. The long-term prognosis of cocoon abdomen after lyses of the adhesions is usually excellent. Thus despite its rarity, this disease should be kept in mind, recognized and treated in patients presenting with recurrent abdominal pain or intestinal obstruction. The importance of correct diagnosis lies in the fact that, in most cases, a simple release of the adhesions is all that is needed to treat this disease.

**References**