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Idiopathic abdominal cocoon syndrome: Preoperative diagnosis with computed tomography

We report the case of a patient with abdominal cocoon syndrome (ACS), an extremely rare cause of intestinal obstruction.

A 52-year-old man with abdominal pain was referred to our radiology department. Written informed consent was obtained from the patient. Contrast-enhanced computed tomography (CECT) of the abdomen showed clustered, distended small intestinal loops within a contrast-enhanced sac-like structure (Figure 1). On laparotomy, the fibrous membrane that encapsulated almost the whole small intestine was carefully dissected and completely excised. Histopathological examination of the excised tissue revealed intense peritoneal fibrosis.

Although ACS was named "abdominal cocoon" by Foo et al. (1) in 1978, it was first described by Owtschinnikow in 1907 as "peritonitis chronica fibrosa incapsulata". This condition may be idiopathic or secondary. The idiopathic form is extremely rare, whereas the secondary form is more common. Underlying causes include extended practolol therapy, continuous ambulatory peritoneal dialysis, tuberculosis, and orthotopic liver transplantation (2,3); none of these were present in our patient. The most frequent clinical presentation of patients with ACS is acute or chronic intermittent small intestinal obstruction (1). Involvement of the large intestine, stomach, or other abdominal organs is not frequent. In our patient however, we noted that in addition to the small intestinal loops, the large intestine and stomach were also encased by the thick fibrous peritoneum. CECT with multiplanar reconstruction images, which clearly shows peritoneal thickening, signs of intestinal obstruction, clustering and fixation of intestinal loops, should be the preferred imaging technique (4). The definite diagnosis of ACS is generally made on the basis of intraoperative and histopathological findings, similar to the present case (5). Treatment should consist of complete surgical excision of the encapsulating peritoneal sac with adhesiolysis.

Figure 1. A coronal reconstruction of contrast-enhanced computed tomography (CECT) image showing a contrast-enhancing sac containing small and large intestinal loops.

Few male cases of idiopathic ACS have been reported in the medical literature, and this condition is difficult to diagnose preoperatively. Clinicians should be aware of the typical radiological findings, and should attempt to arrive at a preoperative diagnosis. This would help to prevent unexpected situations during laparotomy and improve the prognosis of the patients

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