British Journal of Medicine & Medical Research
15(1): 1-3, 2016, Article no.BJMMR.24552
ISSN: 2231-0614, NLM ID: 101570965

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Idiopathic Sclerosing Encapsulating Peritonitis Revealed by Intestinal Obstruction

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Authors' contributions

This work was carried out in collaboration between all authors. Author HZ gave concept of the study and designed. Author EA helped to acquisition of data. Authors HZ and AL analyzed and interpreted the data. Authors HZ and DH drafted the manuscript. Authors AB and ABM approved the final version of manuscript.

Article Information

DOI: 10.9734/BJMMR/2016/24552
Editor(s):
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Complete Peer review History: http://sciencedomain.org/review-history/14154

Case Study

Received 25th January 2016
Accepted 7th March 2016
Published 14th April 2016

ABSTRACT

Sclerosing encapsulating peritonitis (SEP) is a very rare entity characterized by encasement of small intestine by a fibrocollagenous membrane. It is divided into primary (idiopathic) which is named as abdominal cocoon and secondary forms. The preoperative diagnosis is difficult and most cases are diagnosed intraoperatively. A conservative treatment approach is the most suitable management strategy in asymptomatic idiopathic SEP.
In this paper, we aimed to present a case of idiopathic SEP revealed by intestinal obstruction.

Keywords: Sclerosing encapsulating peritonitis; abdominal cocoon syndrome.

1. INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) is a very rare entity characterized by encasement of small intestine by a fibrocollagenous membrane. It is divided into primary (idiopathic) which is named as abdominal cocoon and secondary forms [1]. The preoperative diagnosis is difficult

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and most cases are diagnosed intraoperatively. A conservative treatment approach is the most suitable management strategy in asymptomatic idiopathic SEP [2].

In this paper, we aimed to present a case of idiopathic SEP revealed by intestinal obstruction.

1.1 Observation

A 63-year-old male patient, with no medical history complained about abdominal pain and nausea evolving for three months. The patient also lost at 5-6 kg during the same period. He presented in October 2015 to our emergency department with vomiting. On physical examination, he had a good general state. There was a mobile mass measuring 20 cm occupying the upper abdomen. There were also marked rebound tenderness and guarding in all quadrants. His laboratory tests were as follows: WBC 6.50, Hemoglobin 13 gr/d and normal hepatic and renal functions. There were localized air-fluid levels at midline on abdominal X-Rays. Abdominal computed tomography was in favor of an intestinal obstruction secondary to a volvulus of the jejunum with no signs of suffering (Fig. 1).

Fig. 1. Abdominal CT in frontal plane, showing agglomerate small intestine encased by a dense wall (blue arrow)

Because of the clinical and radiological discordance an exploratory laparotomy is needed. The patient had been informed in detail about the disease, the planned and executed procedures and a consent form had been obtained.

A laparotomy was performed with the presumptive diagnosis of tuberculous peritonitis-carcinomatosis peritonei. Abdominal exploration revealed encapsulation of all intestinal segments by a fibrous membrane that encased whole small intestine and caused dense adhesions (Fig. 2). There is no volvulus.

There were no signs of intestinal suffering so we did not perform membrane excision and adhesiolysis, and peritoneal biopsies were performed. The patient had a favorable evolution and leaved hospital after five days.

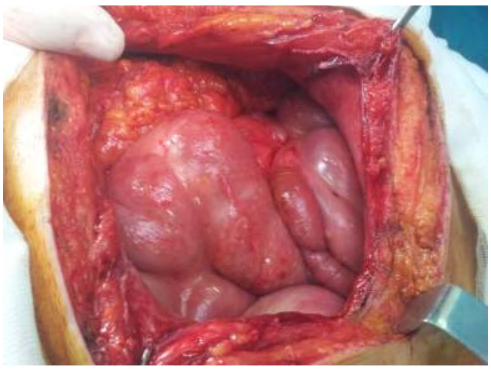

Fig. 2. From: Idiopathic sclerosing encapsulating peritonitis (abdominal cocoon)

Histologic exam showed sclerosis tissue with no identifiable structure. The peritoneum was replaced by a thick fibrous tissue rich in collagen, with connective tissue richly vascularized and infiltrated.

2. DISCUSSION

Depending on the underlying causes, SEP is divided into primary (idiopathic) and secondary forms. The idiopathic form of the disease was named as ‘abdominal cocoon syndrome’ in 1978. Idiopathic SEP is mostly observed in female children living in tropical and subtropical countries, although may be seen in adults of advanced age [3].

The exact etiology is unclear. In women, retrograde menstruation, retrograde extension of gynecological infections has been implicated as probable causes of disease [4]. In contrast, the exact mechanism has remained unclear in men. The most common known cause of secondary SEP worldwide is peritoneal dialysis. Abdominal tuberculosis is responsible for a sizeable part of secondary SEP in underdeveloped countries. Previous surgeries, sarcoidosis, gastrointestinal tumors, fibrogenic foreign body, beta-blocker use, ventriculo peritoneal and peritoneo venous shunts and recurrent peritonitis are among other rare causes [5].

However, the patient presented here had no apparent cause, neither in medical history nor in the microbiological tests. Thus, we considered the case as idiopathic SEP.

While some cases of SEP have an asymptomatic course, most exhibit symptoms characterized by recurrent acute or sub acute attacks of gastrointestinal obstruction [6]. Among the symptoms of the disease, abdominal pain, nausea and vomiting are the primary symptoms. Occasionally, a soft and painless mass lesion can be palpated in some patients. This was the case for our patient.

Abdominal computed tomography (CT) is currently the most useful radiological method for diagnosis of SEP. The characteristic CT sign is the appearance of loops of small intestine that conglomerate at midline and are encased by a dense mantle.

But preoperative diagnosis is still difficult in symptomatic cases with SEP and most patients are diagnosed by intra operative signs and histo-pathological findings.

Conservative management is the most suitable therapy in mildly symptomatic cases. Such cases are best managed by intestinal repose, nasogastric decompression and nutritional support [7].

Surgical therapy is used in markedly symptomatic cases. Different options such as membrane excision with adhesiolysis or resection and anastomosis can be used alone or in combination, depending on the condition of patient [7].

Intestinal resection should be avoided owing to certain complications including anastomosis leaks and short bowel syndrome that increase morbidity and mortality. The role of laparoscopy in the management of SEP is unclear [7].

3. CONCLUSION

SEP forms a minority of unusual conditions that lead to intestinal obstruction. Preoperative diagnosisis a true challenge and most reported cases have been incidentally diagnosed during laparotomy. Conservative approach should be the norm to avoid complications such as anastomosis leaks and short bowel syndrome.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
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