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Karewsky syndrome: A case report and review of the literature



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ABSTRACT

INTRODUCTION: Gallstone ileus can be a lethal disease, rarely suspected in the clinical scenario. It represents about 25% of all bowel obstruction cases in patients older than 65. There is a classification of gallstone ileus based on the onset time: acute, subacute and chronic (Karewsky syndrome). We describe the first reported case of chronic gallstone ileus.

CASE PRESENTATION: A 78-year-old female was admitted to the ER with a 15-day case of consistent bowel obstruction. The subject reported a five-year history of recurrent hospital admissions that resolved spontaneously after non-surgical management. Karewsky syndrome was diagnosed and managed with enterolithotomy. After five days of postoperative evolution the patient was discharged, and at six months follow up, no other hospital admission or relapse has been registered.

DISCUSSION: The gallstone ileus diagnosis demands a higher clinical suspicion, there is no biochemical marker, and an abdominal CT is ideal for imaging-based diagnosis. There is no consensus on the optimal surgical approach.

CONCLUSION: We describe the first case of Karewsky syndrome and a gastro-jejune and gastric-choledochus double fistula. We emphasize the importance of higher clinical suspicion for patients with bowel obstruction older than 65 years old and make evident that although there are not evidence-based guidelines for this treatment, enterolithotomy is a recommended approach.

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1. Introduction

Gallstone ileus (GSI) is a clinical entity of mechanical bowel obstruction [1–4]. The most frequent mechanism by which this occurs is by migration of a gallstone through a cholecystoduodenal fistula that gets impacted in the distal ileum lumen [3].

GSI has a female:male ratio of 3.5–6:1, affecting older patients predominantly [1,3] as it represents 4% of all bowel obstruction in patients under 65 years-old and approximately 25% in patients over 65 [5,6]. With regards to the general population, GSI represents 0.3–0.5% of all gallstone complications with 35 per million hospital admissions [2,7].

Clinical signs and symptoms of GSI are non-specific [7], Beuran et al. proposed three clinical presentations: acute, which corresponds to the classical gallstone ileus, subacute, with a partial bowel obstruction, and chronic (Karewsky syndrome). Karewsky

syndrome is characterized with recurrent episodes of abdominal pain due to the passage of a gallstone lodged in the bowel lumen with interspersed asymptomatic long lapses.

This paper is the first to report a patient with Karewsky syndrome [8] comorbid with choledochus-gastric gastro-jejunal fistula.

2. Case presentation

A 78-year-old female was admitted to the emergency department with abdominal distention, diminished bowel movements and rectal tenesmus with 15 day's progression, self-treated with over-the-counter drugs. Five days before admission she experienced complete absence of bowel movement and flatus accompanied by vomiting, progressing two days from gastric to fecaloid content. Immediate relatives described a five-year history of recurrent admissions for partial bowel obstruction and two admissions for complete bowel obstruction resolved with non-surgical management and no apparent etiology identified. Previous surgical events and other comorbidities were denied.

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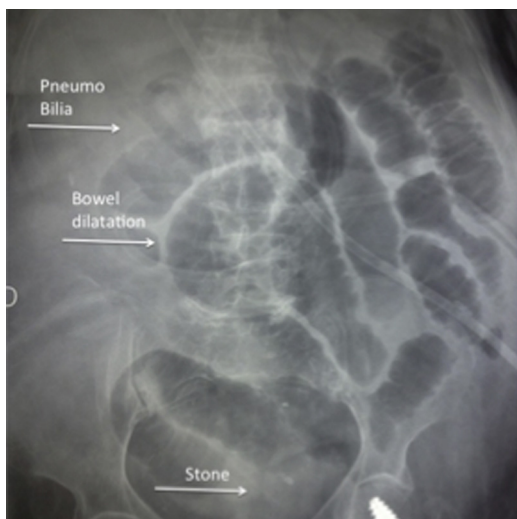


Fig. 1. Rigler's triad.

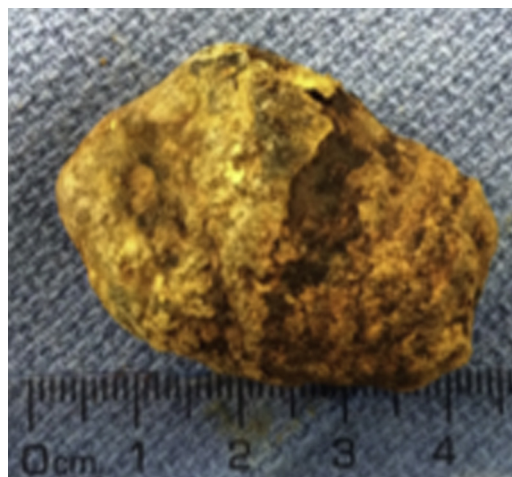


Fig. 3. Gallstone.

Physical exploration revealed a slightly distended and tender abdomen without any surgical scar or a palpable hernia, diminished peristalsis, negative Murphy and Blumberg sign. Laboratory tests were normal (not reported in present paper).

We initiated management protocol for bowel obstruction, with a partial positive response. The abdominal X-ray revealed the presence of Rigler's triad (Fig. 1) confirmed in the abdominal CT (Fig. 2). We then scheduled an emergency laparotomy, finding the site of obstruction at the rectosigmoid junction and extracting the stone with manual extrinsic pressure through the anus (Fig. 3). We performed a Noble maneuver to discard the possibility of other intraluminal stones, and did not perform a cholecystectomy or fistula repair.

To appropriately clarify the tract of the fistula, we performed a water-soluble contrast-enhance GI tract X-ray on the third postoperative day, which showed a gastro-choledochus and gastro-jejunal fistula (Fig. 4.). She had a benign clinical progression and was discharged on the fifth postoperative day with a 6-month follow-up and no signs or symptoms of recurrence.

3. Discussion

GSI was described and classified previously by Rodriguez-Hermosa JL y cols 2001 and Beuran et al., a typical clinical scenario is preceded by recurrent episodes of acute cholecystitis which produce high inflammation and adhesion formation between bowel and gallbladder that in conjunction with the increased intraluminal pressure and the secondary ischemia lead to the bilio-enteric

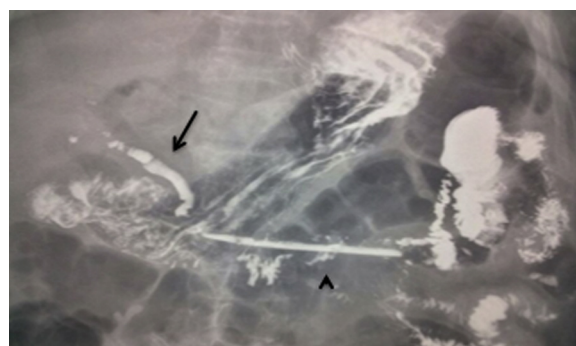


Fig. 4. Pneumobilia, Petrenis sign (black arrow), gastro-jejunal fistula (arrowhead).

fistula formation [2]. All the later eventually will allow the passage of stones to the GI tract [5].

The most common site for fistula formation is the duodenum (60–86%) [8]. Other reports include cholecystocolonic (5–25%), cholecysto-duodenal-colonic (2.5%), cholecystojejunal, cholecystogastric, choledochoduodenal and duodenum-left hepatic duct [2]. To date, this is the first reported case of a proved double fistula as noted in Fig. 4.

The diagnostic approach is non-specific, laboratories tests are not useful in leading to the diagnosis hence demonstrating some complication of GSI. Also, imaging tests reveal signs that are not easily seen by the inexperienced eye [8–10]. Furthermore, plain abdominal films have a diagnostic sensibility of 40–70% [11], with

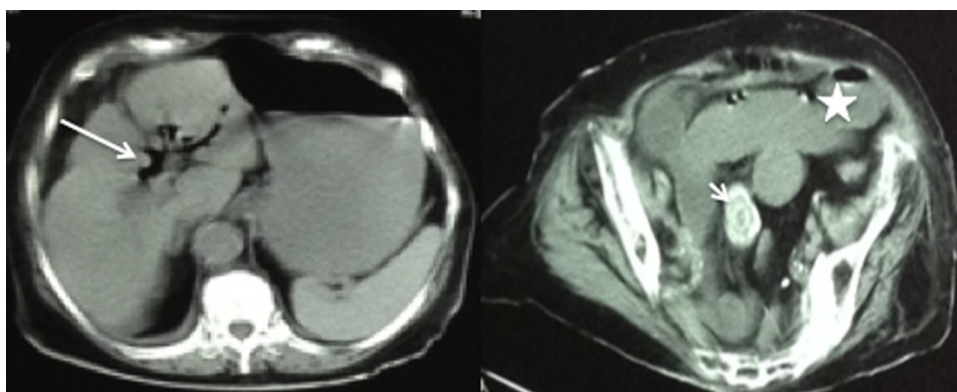


Fig. 2. Abdominal CT displaying Rigler's triad, pneumobilia (white arrow), bowel loop.

Rigler's triad presents in 40–50% of patients [8,1,12], although, it could be less [13,14].

Water-soluble contrast-enhanced abdominal CT is the gold standard diagnostic test for GSI with 93% sensibility [15]. It helps to establish the presence of pneumobilia [2], cholecysto-enteric fistula [16], the lodging site of the ectopic stone [2,5] and its dimensions [17,18].

Contrast-enhanced abdominal X-ray is no longer used as a routine diagnostic tool but remains useful when a CT scan is not available. It requires the administration of a water-soluble contrast, displaying at the lodging site a characteristic image of "snake head-like", known as the Forchet sign [7]. The passage of contrast to the biliary tree shows the presence of a patent bilio-enteric fistula, also known as, Petren sign [8] (Fig. 4). We suspected GSI in the initial abdominal X-ray (Fig. 1) and confirmed the diagnosis in the abdominal CT (Fig. 2). In this case the diagnostic approach to the fistula was made postoperatively.

Once the diagnosis was made, an emergency laparotomy was performed, extracting the stone by manual compression through the anus, without repairing the fistula. Several surgical approaches have been described, such as enterolithotomy, one-step approach, and two-step approach [4,9,16,19]. However, no evidence-based recommendation has been made to describe the best procedure in GSI. There is an apparent minor rate of complications with enterolithotomy compared with the rest of them, along with a slight recurrence rate 5% [3].

Even though there are not randomized trials nor systematic reviews and meta-analysis for the treatment of GSI, there are a few series of cases that compare the outcomes between enterolithotomy alone (EL) vs one-step approach (OSS) which show similar post-operative morbidity, with a major mortality, major operating time, and hospitalization length in the group of OSS [20,14].

Riaz et al., [17] shows that good stratification of the patients in two groups (hemodynamically-unstable and hemodynamically-stable) with an ASA score of 3 or more and less than 3, allow to be perceived that the former will receive an EL and the latter an OSS, since they had no mortality nor re-intervention. However, their sample is small to give a recommendation.

It has been described previously that the recurrence of biliary symptoms or GSI after EL is of less than 10%, and it presents predominantly in the first six post-operative months [4], that is the argument to perform OSS in most cases, nevertheless, re-intervention in both groups has not significant differences [20,21], with this results, no real benefit is seen in the OSS group.

Overall mortality of GSI is of 14.28% [7], even though, Rodríguez-Sanjuán et al., [21] Mallipeddi et al., [20] demonstrated no statistical significance difference between mortality in the group of EL vs OSS. This again shows, that even if there is not significant higher mortality in the OSS group, there is not a real benefit in performing the OSS.

The patient had an uneventful postoperative evolution and was discharged on the fifth postoperative day. During the six-month follow-up, no surgical complication or recurrence has been detected.

4. Conclusion

We present the first Karewsky syndrome with gastric-jejunal and gastric-choledochus double fistula reported in medical liter-

ature. There is no standard surgical protocol, but there is clear evidence that conservative surgical management can be useful for this clinical entity. We emphasize the importance of higher clinical suspicion in the elderly—particularly for those with previous episodes of biliary colic or other complications. Ultimately, although there are no evidence-based guidelines for this treatment, enterolithotomy is the recommended approach.

Disclosure

None of the authors reported conflict of interest.

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