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Gallstone ileus in a patient with amyotrophic lateral sclerosis: A case report

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Gallstone ileus is a rare disease that most commonly occurs in elderly females with a history of cholelithiasis. It has not been previously associated with Amyotrophic Lateral Sclerosis (ALS); a neurodegenerative disease that primarily affects the motor neurons at the spinal and bulbar levels. Autonomic malfunction, in particular, gastrointestinal dysfunction has been documented in ALS patients which may predispose this population to the development of gallstones and gut dysmotility. **CASE PRESENTATION:** In this paper, we report a case of gallstone ileus in a patient with diagnosed ALS. We performed an exploratory laparotomy, enterolithotomy, and an open cholecystectomy with take-down/closure of a cholecystoduodenal fistula. The patient had a relatively uncomplicated postoperative course and was discharged from the hospital on postoperative day nine.

CLINICAL DISCUSSION: Delays in gastric emptying and colonic transit times in ALS patients may pose a risk for the development of gallstones and the potential impaction of a gallstone ileus in patients who are left untreated. Multifactorial evaluation of this patient population is necessary when assessing a potential causal pattern of gallstone ileus in patients with significant comorbidities.

CONCLUSION: We present an unusual pathology without an established incidence, which has pertinent multidisciplinary implications. The suspicion of ALS as a potential cause for the development of a gallstone ileus is relevant and essential in the diagnostic workup for an elderly patient who develops a small bowel obstruction with multi-comorbidities.

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1. Introduction

Gallstone ileus (GI) is an uncommon sequela of cholelithiasis, defined as a mechanical impaction and/or obstruction of one or more gallstones within the gastrointestinal tract. The term “ileus” is a misnomer because this pathology is not a result of cessation of movement, rather mechanical obstruction [1]. In patients diagnosed with cholelithiasis, approximately 0.3–0.5% develop a GI [1]. The incidence of GI is increased in individuals over the age of 60 years with a higher prevalence in females (up to 6:1 female to male ratio) [2]. The fistulous connection between the gallbladder and adjacent abdominal organs is a product of a chronic inflammatory process resulting in adhesive formation [1]. Most commonly, a cholecystoduodenal fistula forms, however, a chole-

cystocolic, cholecystogastric, and choledochoduodenal connection may occur [1,2]. Patients have often developed GI as a consequence of chronic and/or poorly controlled Crohn's disease, post-ERCP, Mirizzi syndrome, and post laparoscopic cholecystectomy with failure to retrieve intraabdominal gallstones [3]. Until the present time, there has not been any well-established literature on patients with ALS who have developed GI, however, potential autonomic dysfunction stemming from ALS has been reported. This report has been written in conjunction with the surgical case report guidelines (SCARE) criteria [4].

2. Case presentation

We report a case of a 69-year-old woman with a 30-year history of Amyotrophic Lateral Sclerosis, who has not utilized medical management since her diagnosis. The patient has a confirmed history of cholelithiasis in 2011 and elected against surgical intervention at that time. She presented to the emergency department complaining of four days of constant, right upper-quadrant (RUQ)/epigastric pain associated with a diminished appetite, nausea/vomiting, and constipation. On clinical examination, the patient had a positive murphy's sign without indications of peri-

Abbreviations: GI, gallstone ileus; ALS, Amyotrophic Lateral Sclerosis; CT, computed tomography; WBC, white blood cell; ERCP, endoscopic retrograde cholangiopancreatography; NGT, nasogastric tube; POD, post-operative day.

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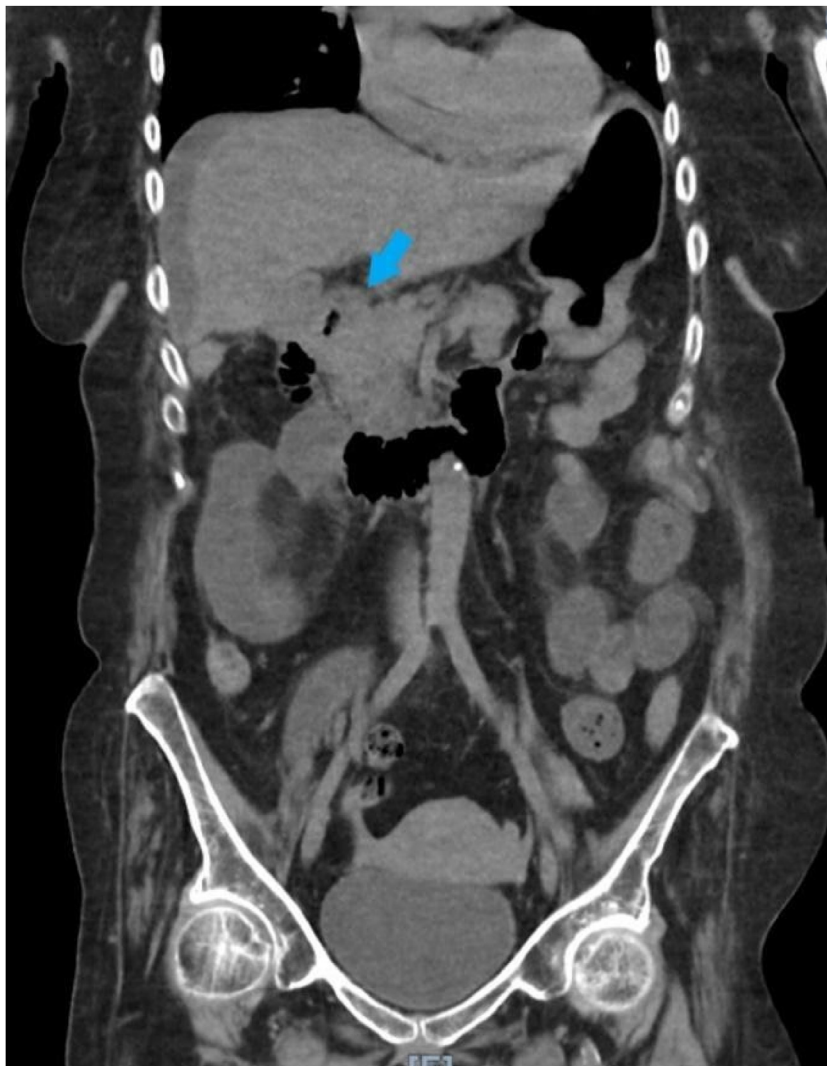


Fig. 1. Coronal view of abdominal CT scan showing fistulation between the gallbladder and duodenum (blue arrow).

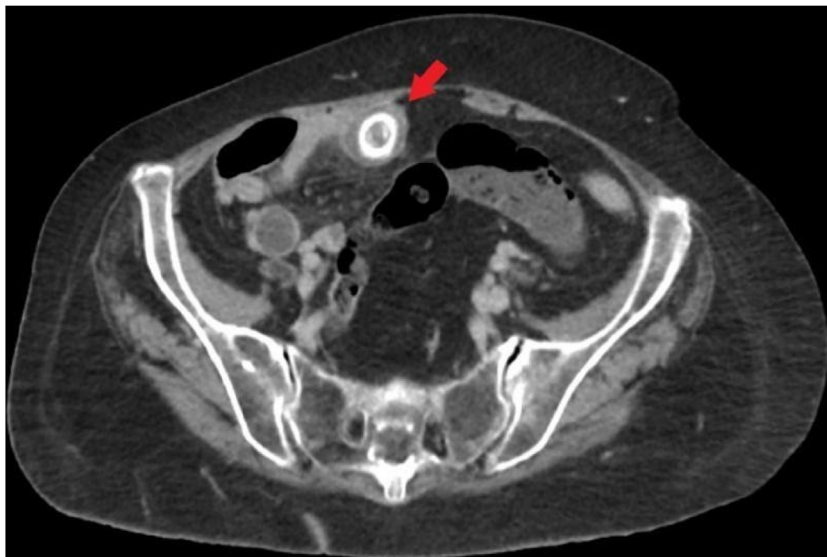


Fig. 2. Axial view of abdominal CT scan displaying a gallstone in the terminal ileum (red arrow).

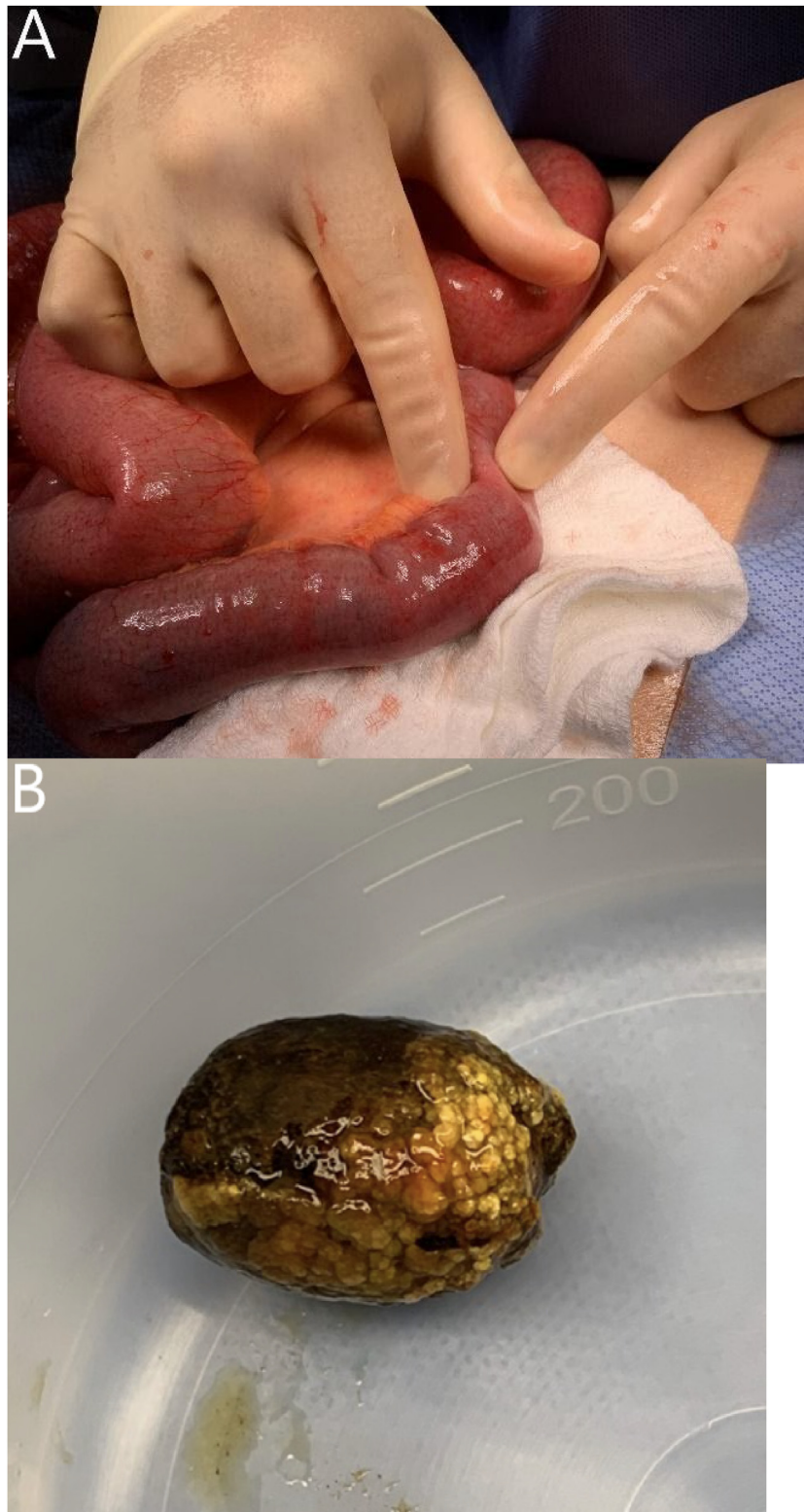


Fig. 3. Distal impaction of the gallstone identified at the terminal ileum (A). Successful enterolithotomy of a 2 cm stone (B).

tonitis. A RUQ ultrasound was performed, demonstrating gallstones and sludging within an amorphous gallbladder. A possible gallbladder mass with chronic inflammatory changes and perforation could not be ruled out; prompting an abdominal CT scan. A poorly defined gallbladder with fistulation to the second portion of the duodenum was noted (Fig. 1). A large gallstone measuring approximately 2 cm was found in the terminal ileum (Fig. 2). The patient was noted

to have a leukocytosis of 14,500 per cubic millimeter with 92% neutrophils. Isotonic fluid resuscitation was initiated, as well as, intravenous Piperacillin-Tazobactam 3.375 g every eight hours.

The patient was taken to the operating room for endoscopic retrograde cholangiopancreatography (ERCP) with biliary sphincterotomy, which returned no findings. Post-ERCP, an exploratory laparotomy was performed by the attending hepatobiliary surgeon

with the chief surgical resident and senior surgical resident assisting throughout the procedure. Stone impaction at the terminal ileum was identified (Fig. 3a), followed by a successful enterolithotomy and stone retrieval (Fig. 3b). An open cholecystectomy, and takedown/closure of cholecystoduodenal fistula was completed. Prior to leaving the operating room, a nasogastric tube (NGT) and blake drain were placed. The following management was focused on pain control, daily incisional care, appropriate return to functional baseline, and continuation of antibiotic-based treatment with 3.375 mg of intravenous Piperacillin-Tazobactam every eight hours for a total of seven days. The patient's postoperative course was relatively uncomplicated. An initial white blood cell count (WBC) and hemoglobin (Hgb) concentration of 25.2 and 13.5 was reported on post op day (POD) one. By POD four, her WBC count had normalized to 5.9, with an increase in Hgb to 11.9 from the previous day. The patient's bowel function had returned by POD three. The NGT was removed on POD seven and the blake drain was removed prior to discharge on POD nine. The patient returned to the clinic for two and four-week post op checks; during this time her incision was healing appropriately with daily dressing changes performed by a home-nurse. She was maintaining a regular diet with normal bowel function.

3. Discussion

The incidence of GI is approximately 30–35 cases/1,000,000 admissions [1], contrasted to approximately 2.7 cases/100,000 for ALS in North America and Europe [5]. ALS is characterized as a fatal neurodegenerative disease of the upper and lower motor neurons at the spinal and bulbar levels [5], with symptoms classically developing as limb weakness, progressing to speech and respiratory deterioration [5,6]. However, there is mounting evidence suggesting ALS extends beyond the anterior horn cells and corticospinal tracts. Autonomic symptoms have been reported in over 25% of patients with diagnosed ALS; most commonly urinary or gastrointestinal dysfunction, and orthostatic intolerance [7–10].

The pathophysiology of ALS is complex and the degree of autonomic dysfunction remains largely unknown [7]. It has been reported, however, that ALS patients with chronic tracheostomy ventilation were found to be at significantly higher risk of developing cholelithiasis/cholecystitis [11]. The amount of intestinal hypoperistalsis may factor into poor gallbladder contractility, resulting in biliary stasis and sludge development [8,12]. Also, hyperfunction of the adrenergic sympathetic nervous system may contribute to splanchnic hypoperfusion [11], which in-turn, may lead to worsening gallbladder contractility, stasis, sludge and gallstone formation. While patients with significant respiratory compromise may be at higher risk for gallstone formation, the risk of cholelithiasis was not evaluated in patients whose neurodegenerative function was not severely compromised, like in our patient. The neurodegenerative changes affecting gastrointestinal motility in ALS patients was further verified when comparing gastric emptying with healthy controls after ingesting a radioactive isotope tracer [9]. A total of 83% of patients in this study with diagnosed ALS had a delay in gastric emptying by more than 160 min [9]. Colonic transit times were also found to be significantly delayed in the right and left colon of ALS patients when compared to age-matched controls [10]. Interestingly, the rectosigmoid transit time was within normal limits when compared to the healthy control group [10]. A significant limitation that should be considered with both of these studies is the relatively small patient populations of 14 and 18 patients respectively.

It is noteworthy to comment that our patient had a diagnosis of ALS more than thirty years prior and has been living without the support of medical management. The degree of progression

of her disease should be considered when questioning the level of dysfunction it may contribute to the autonomic nervous system. Other important factors including; nutritional status/fiber content in the diet, age-related deconditioning/inactivity, surgical and past medical history should be thoroughly evaluated when trying to determine a causal relationship of ALS with a gastrointestinal pathology such as GI [1,3,13]. Further assessment of small intestinal peristaltic function, especially in the terminal ileum of ALS patients is necessary to determine if this poses a risk to this patient population.

4. Conclusion

A gallstone ileus is a rare disease that commonly affects elderly female patients with or without signs and symptoms of intestinal obstruction. Neurodegenerative comorbidities, such as ALS may pose a potential risk for the development of gallstones in patients with advanced disease and compromise the physiological gastrointestinal transit. Since this pathology typically affects an aged population, it is imperative to keep in mind that there may be other physiological factors that can pose a risk for development of gallstone ileus.

Declaration of Competing Interest

No.

Funding

No.

Ethical approval

The ethical approval has been exempted by our institution. Not required for this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Vincent Marcucci and Sindi Diko wrote the article. Vincent Marcucci, Sindi Diko, and Derick Christian participated in the patient's care. Derick Christian supervised the article writing.

Registration of research studies

Not Applicable.

Guarantor

Derick Christian.

Provenance and peer review

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