



## Body Imaging

Esophageal pneumatosis: Case report and review of literature<sup>☆</sup>Mayur Virarkar<sup>\*</sup>, Revathy Iyer, Priya Bhosale, Tharakeswara Bathala, Wui Chong, Girish Shroff

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## ABSTRACT

Pneumatosis of the gastrointestinal tract is defined as presence of air in the wall of the gastrointestinal tract and can occur in any part of the gastrointestinal tract. It is most commonly seen in the intestine and very rarely in the esophagus. The exact pathogenesis is still unknown. It is managed primarily by conservative and non-surgical therapy, unless there are findings to suggest an acute abdomen or other co-morbidities. On review of literature, very few case reports of esophageal pneumatosis have been published. We present a rare case of pneumatosis of the esophagus with cytomegalovirus (CMV) infection.

## 1. Introduction

Pneumatosis of the gastrointestinal tract is characterized by the presence of air filled spaces in the submucosa and subserosa. It can occur in various locations in the gastrointestinal tract and can be asymptomatic or rarely present with acute abdomen. The most common clinical manifestations include chest pain, epigastric or abdominal pain and gastrointestinal bleed. Esophageal pneumatosis is a rare condition with diverse potential etiologies [1]. There are few case reports of esophageal pneumatosis in the literature. We report a unique case of esophageal pneumatosis with cytomegalovirus (CMV) infection and review of the literature.

## 2. Case report

41 year-old-female presented to the emergency room (ER) with chief complains of epigastric pain. She has a past medical history of myeloproliferative disease (MPD) and treated with chemotherapy. Currently, she is in complete remission. Her initial ER laboratory work up showed normal complete blood count, liver function test, amylase and lipase with mild increase in creatinine to 1.8 (normal – 1.5). The radiograph of the chest revealed no abnormality in bilateral lung fields with no air under the diaphragm (Fig. 1).

She underwent a Computerized Tomography (CT) of the abdomen and pelvis which reported circumferential air within the visualised distal esophageal wall and raised a suspicion of esophageal pneumatosis. No extraluminal air was noted to suggest perforation. There was

no evidence of acute intestinal obstruction. The continued epigastric pain raised a clinical suspicion of perforation and hence, a CT of the chest with esophageal leak protocol was ordered (Fig. 2). The CT of the chest esophageal leak protocol consists of a non-contrast CT chest and later an oral and intravenous (IV) contrast CT chest. It reported no evidence of extravasation of contrast to suggest active esophageal perforation. There was circumferential air in the wall of the proximal, mid and distal esophagus compatible with diffuse esophageal pneumatosis. There were small foci of air seen lining the left aspect of the proximal esophagus which could suggest walled-off microperforation. The oral contrast was seen in the stomach and small bowel loops. The Thoracic surgery service was consulted for esophageal pneumatosis. After examination, a conservative management was decided and an esophagogastroduodenoscopy (EGD) was planned. He was kept on parenteral nutrition, intravenous fluid and broad spectral antibiotics.

A barium esophagogram on next day identified diffuse shallow ulcers throughout the esophagus with a preponderance in the mid and distal esophagus. The findings were suggestive of esophagitis (Fig. 3). There is no evidence of esophageal perforation, stricture or mass. Normal transit of oral contrast was seen from the esophagus into the stomach.

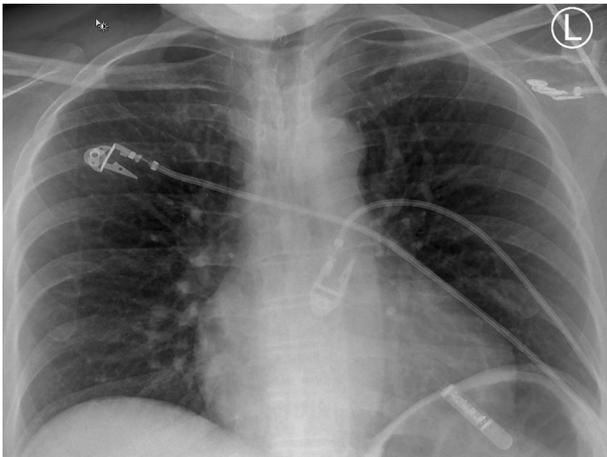
EGD performed on next day (Fig. 4) showed a 3 mm superficial esophageal ulcer without bleeding or stigmata of recent bleeding at the gastroesophageal junction. Los Angeles (LA) Grade D (one or more mucosal breaks involving at least 75% of esophageal circumference) esophagitis with no bleeding was found in distal esophagus. The entire examined stomach was normal. Patchy mildly erythematous mucosa

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**Fig. 1.** 41-year-old-female with esophageal pneumatosis. Radiograph of the chest shows normal bilateral lung fields. No air is seen under the diaphragm.

without active bleeding and with no stigmata of bleeding was found in the first and second portions of the duodenum.

The histology of the lower esophagus biopsy reported squamous mucosa with luminal necro-inflammatory debris, ulcerations, granulation tissue formation and viral cytopathic changes. No columnar epithelium, intestinal metaplasia, dysplasia or carcinoma was identified. Biopsy of the gastroesophageal junction ulcer showed squamo-columnar junction mucosa with active chronic inflammation and reactive epithelial changes. Detached luminal cells with viral cytopathic effects were noted. No evidence of intestinal metaplasia, dysplasia or carcinoma was identified. Immunohistochemical stains performed reported negative for Epstein-Barr virus. CD68 highlighted increase histiocytes. CD20 and CD3 showed distinct B cell and T cell compartments. CD30 stains were reactive for immunoblasts. Kappa and lambda light chain expressions were polytypic in the plasma cells. CD21 highlighted the follicular dendritic cell meshwork. The germinal center cells were positive for CD10 and negative for BCL2. Myeloperoxidase (MPO) stains showed admixed neutrophils. The overall findings support a reactive process, likely secondary to the underlying CMV infection. In addition, the CMV immunostains were also positive.

She was treated with valganciclovir 450 mg tablet twice a day and discharged. On follow-up after 2 months, she was asymptomatic. The follow-up CT of the chest showed resolution of the esophageal pneumatosis (Fig. 5).

### 3. Discussion

Pneumatosis of the gastrointestinal tract can be seen from the esophagus to the rectum. However, esophageal pneumatosis is a rare occurrence. Pneumatosis may be idiopathic or may occur secondary to underlying pathological process. Some causes include necrotizing enterocolitis and intestinal obstruction. Other non-gastrointestinal disorders include infectious enteritis, acquired or primary immunodeficiency conditions and hematological malignancies [1].

An extensive literature search was performed on PubMed and Ovid databases using the keyword- esophageal pneumatosis. The reference lists of all retrieved studies were scrutinized for additional articles to supplement the search result. All potentially eligible studies were checked to determine whether they fulfilled the following criteria: (a)

articles published in English language and (b) reported on human species. All the duplicates articles were excluded. The flowchart of the patient selection is shown in Chart 1. The initial literature search identified 19 articles. The application of addition search filter of Human species and English language resulted in exclusion of 5 articles and 14 potentially relevant articles were screened. Subsequently, 3 duplicates articles were excluded and yielded final 11 studies with 13 cases (Table 1). The mean age in the reported case reports was 70 years (range 39–87 years).

There are two important theories to explain the occurrence of pneumatosis: mechanical and infection theory. According to mechanical theory, air present in the gastrointestinal tract lumen dissects into the walls of the weakened gastrointestinal tract. Infection theory proposes that the air entering the walls of bowel is produced by the infection. In our case report, CMV was associated with esophageal pneumatosis. The probable cause would be dissection of air in the wall of the weakened and inflamed esophagus by the viral infection.

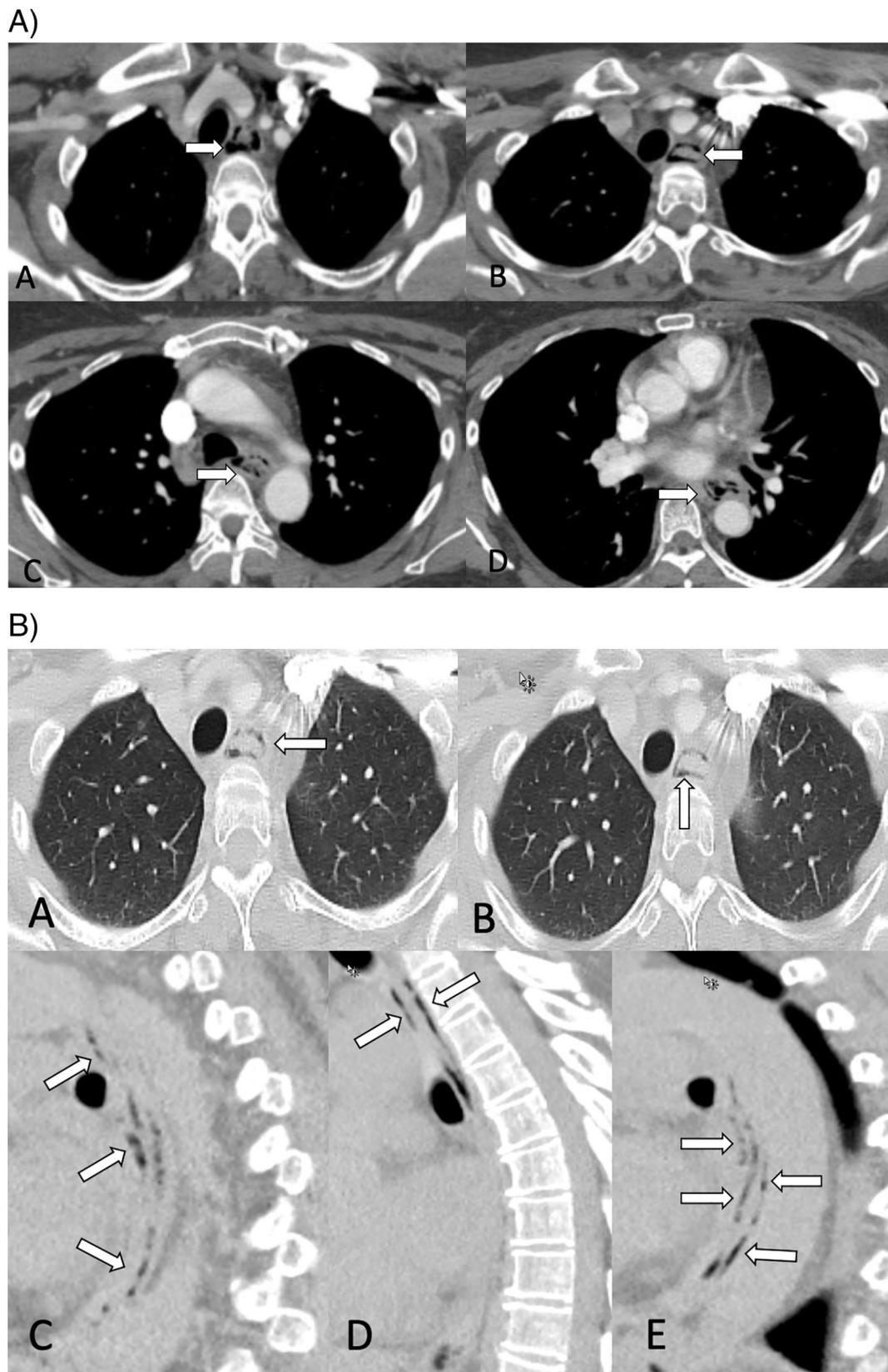
CMV is a type of herpesvirus with multiple organ involvement. It causes various diseases, such as retinitis, hepatitis, encephalitis, pneumonia, and gastrointestinal infection in immunocompromised patients [2]. Esophagitis is the common gastrointestinal manifestation of CMV. CMV esophagitis has been reported in immunosuppressed patients. Its manifestations include, difficulty in oral painful swallowing, nausea/vomiting, abdominal pain, weight loss, and diarrhea. The diagnostic procedure of choice is endoscopy to visualise any mucosal lesions and to obtain a tissue biopsy [2,3].

The clinical presentation of esophageal pneumatosis includes retrosternal chest pain, abdominal pain, septic shock or asymptomatic. The symptoms vary depending upon location and extent of the pneumatosis. Initial workup of pneumatosis should be focused on ruling out diagnosis of acute abdomen or sepsis and identification of the underlying pathology with radiographs of the chest and/or abdomen. It should be followed by CT of the abdomen and pelvis for identifying intestinal obstruction or pneumoperitoneum [1,4]. The esophagogram can illustrate edematous mucosa, superficial ulceration, stricture or mass. After stabilisation of the patient, EGD can be helpful for direction visualisation and taking biopsy. Endoscopy can demonstrate esophagitis, ulcer, pneumatosis or active bleeding. CMV esophagitis can present with superficial ulceration, erosions, haemorrhage and nodules [5]. The diagnosis is confirmed on histopathologic identification of viral-infected cells in biopsy and immunohistochemistry.

Management depends on the underlying etiology. Various treatment modalities have been utilized including oxygen therapy and conservative. Endoscopic therapy like cyst puncture and sclerotherapy has also been tried in the past. However, the effective management is conservative and non-surgical therapy, unless there are findings to suggest acute abdomen or other co-morbidities requiring emergent surgery. CMV esophagitis has been treated with oral Ganciclovir [3]. In our case, the patient did not response to initial proton-pump inhibitor treatment, and symptoms eventually resolved only after ganciclovir treatment.

### 4. Conclusion

We present the first case report of esophageal pneumatosis with CMV infection. In our patient, pneumatosis esophagitis was effectively treated with conservative management and in concordance with the available literature.



**Fig. 2.** 41-year-old-female with esophageal pneumatosis. CT of the chest (2.1 A, B, C, D) axial soft tissue window, (2.2 A, B) axial lung window, (2.2 C, D, E) sagittal lung window, (2.3 A, B, C, D, E) coronal soft tissue window and (2.4 A, B, C, D, E, F) sagittal soft tissue window images show circumferential air (arrows) in the wall of the proximal, mid and distal esophagus compatible with esophageal pneumatosis.

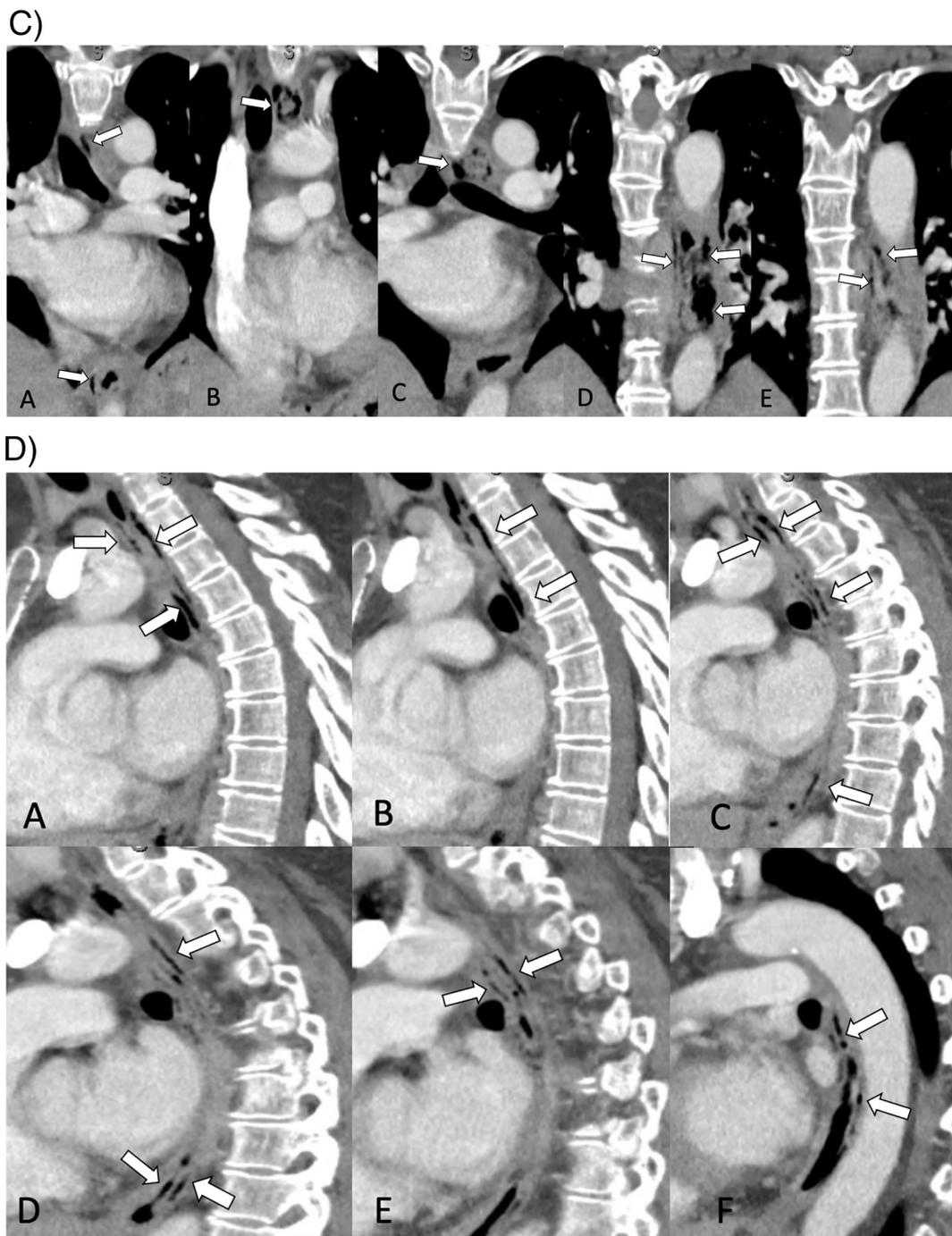


Fig. 2. (continued)

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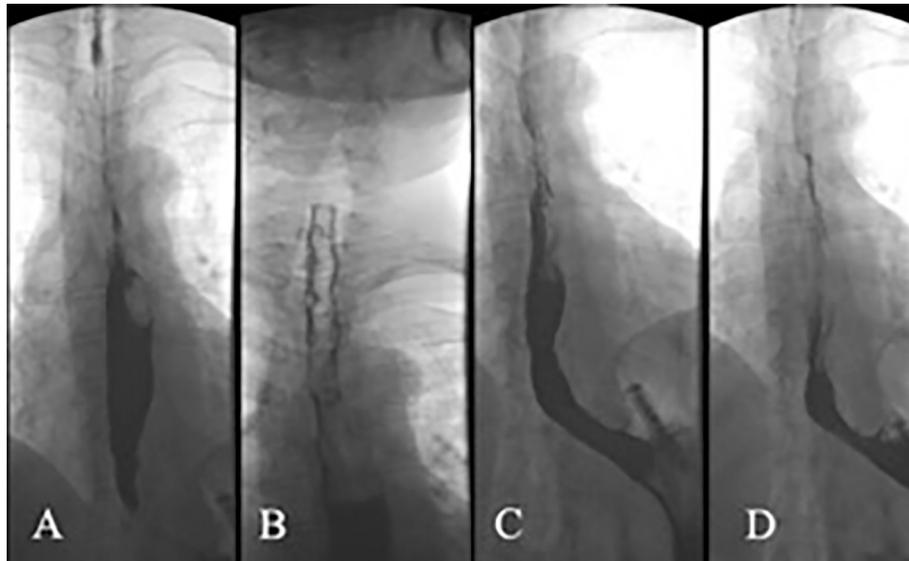
**Ethical approval**

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/

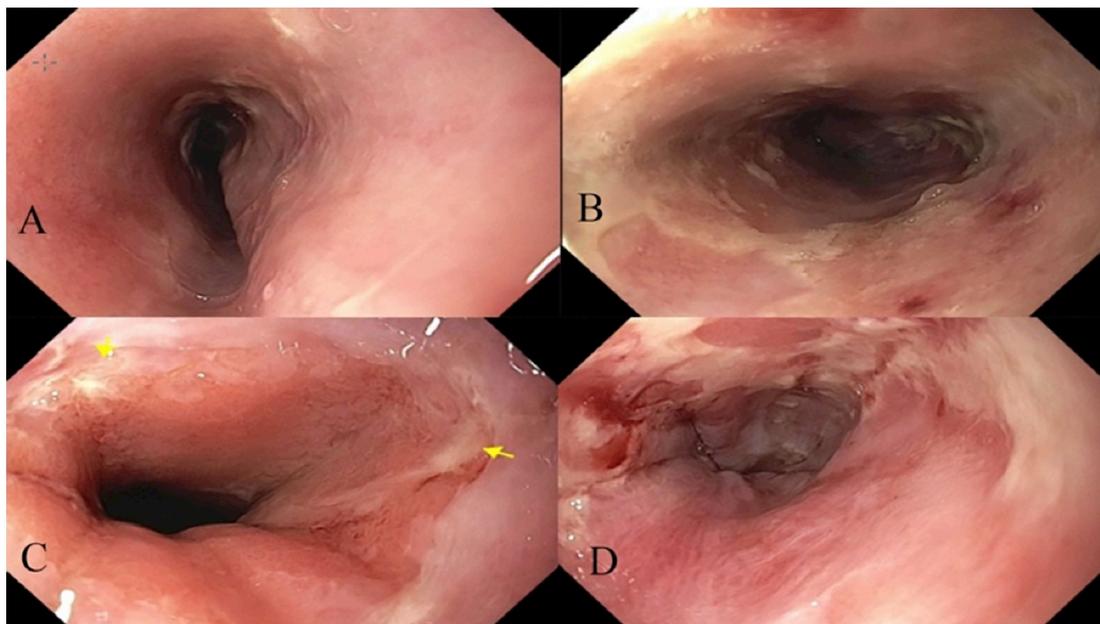
or national research committee and with the 1964 Helsinki declaration, and its later amendments or comparable ethical standards.

**Informed consent**

Informed consent was obtained from the patient before enrollment in the study.



**Fig. 3.** 41-year-old-female with esophageal pneumatosis. (A, B, C, D) Esophagogram images show diffuse shallow ulcers in the esophagus compatible with esophagitis. There is no evidence of esophageal perforation, stricture or mass.



**Fig. 4.** 41-year-old-female with esophageal pneumatosis. Esophagogastroduodenoscopy (A) upper third, (B) middle third, (C) gastroesophageal junction and (D) lower third show a 3 mm superficial esophageal ulcer (long yellow arrow) and esophagitis (short yellow arrows) with no bleeding or stigmata of recent bleeding at the gastroesophageal junction. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

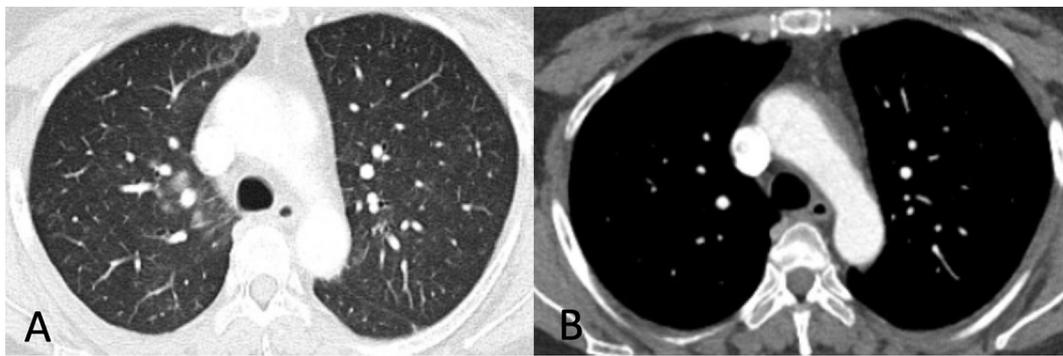


Fig. 5. 41-year-old-female with esophageal pneumatosis. CT of the chest (A) Axial lung window and (B) soft tissue window demonstrate resolution of esophageal pneumatosis.

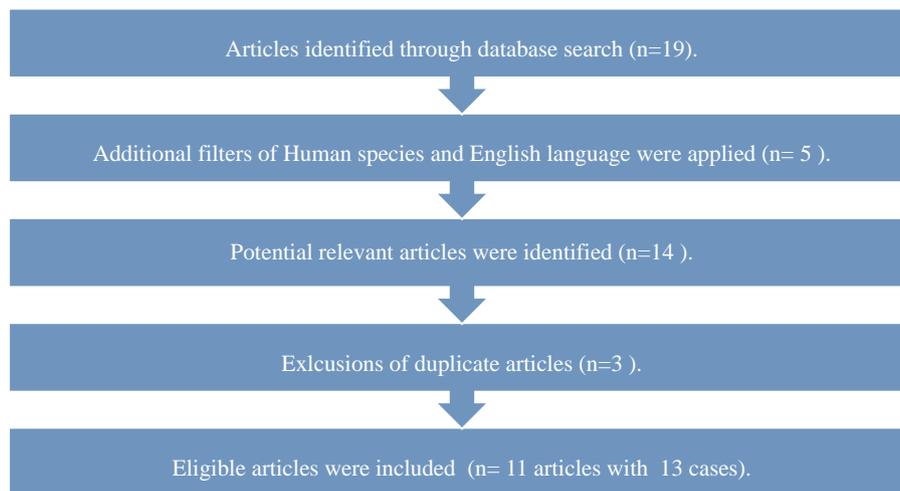


Chart 1. Flowchart of selected patients.

Table 1  
Clinical and pathological characteristics and management of Esophageal pneumatosis.

Sr. no.	Author (year)	Age (years)	Sex	Primary cause	Management	Follow up
1	Vanasin et al. [6] 1971	62	F	Idiopathic esophageal stricture	None	Died due to aspiration pneumonia
2	Katz et al. [7] 1972	85	F	Esophagoscopy procedure complication	Exploratory laparotomy to rule out perforation.	Died
3	Katz et al. [7] 1972	46	F	Esophagoscopy procedure complication	Thoracotomy, esophagectomy and esophagogastrotomy for carcinoma.	Survived
4	Katz et al. [7] 1972	39	F	Esophagoscopy procedure complication	Antibiotic	Survived
5	McKelvie et al. [8] 1994	76	M	Infection	None	Died
6	Tixedor et al. [9] 1998	74	F	Abdominal surgery complication	Conservative treatment and nasogastric Aspiration	Survived
7	Mclaughlin et al. [10] 2007	77	M	Stomach dilation	Conservative treatment and Oxygen	Survived
8	Yahng et al. [11] 2011	62	F	Chemotherapy	Conservative	Expired due to ARDS
9	Chelimilla et al. [4] 2013	78	F	Laryngocele induced.	Conservative	Survived
10	Lin et al. [12] 2015	85	M	Ischemic bowel disease	Conservative	Died due to sepsis.
11	Huang et al. [13] 2015	70	M	Post esophagoscopy	Medical Management	Survived
12	Muhammad et al. [14] 2017	70	M	Septic shock	Medical Management	Died
13	Tewari et al. [15] 2017	87	M	Small bowel ileus	Medical Management	Survived

## Declaration of Competing Interest

The authors declare that they have no conflict of interest.

## Acknowledgments

None.

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