Case Report

Gastroduodenal Emphysema with Portal Venous Air due to Congenital Duodenal Web in a Child: A Case Report and Review of Literature

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1.Introduction

Gastroduodenal emphysema is a very rare clinical presentation in infancy [1, 2]. Evidence suggests that gastroduodenal emphysema is associated with gastric outlet obstruction secondary to pyloric stenosis and with duodenal obstruction [1, 2]. Out of the reported cases of gastric emphysema in infants, only a few cases were associated with duodenal obstruction [3]. Depending on the degree of duodenal obstruction, the age of presentation of the related clinical symptoms varies [3]. However, usually, these patients present in the first few months after birth [3]. Late presentations are rare, which need a high degree of suspicion for the correct diagnosis [4].

Congenital duodenal web is a known cause of neonatal duodenal obstruction, and depending on the type of the web, the clinical presentation and the age of the presentation may vary [5]. Only a limited number of case reports have been published on congenital duodenal web leading to duodenal obstruction and presenting with gastroduodenal...
emphysema in infancy [1, 2, 6]. Windsock deformity is another uncommon association with duodenal web reported in the literature [5, 7].

Portal venous air is very rarely associated with gastro-duodenal emphysema [8, 9]. Even though portal venous air is usually considered as an ominous imaging finding, it could also have benign aetiologies [8, 9].

We present a very rare case of an eighteen-month-old child who developed gastroduodenal emphysema and portal venous air secondary to proximal duodenal obstruction due to a congenital duodenal web.

2. Case Presentation

An eighteen-month-old male child, who was under investigation for failure to thrive, presented with a history of frequent vomiting for two days. He had been having episodes of similar symptoms for the past one year, which were self-limiting and usually settling within one day. During these episodes, initially, the vomitus contained clear fluid; however, later it turned into coffee-ground colour. In addition, he developed progressive abdominal distension. On examination, he was afebrile and not drowsy; however, he was mildly dehydrated. The child was in discomfort due to abdominal distension.

Initial basic haematological investigations such as full blood count, C-reactive protein, serum electrolytes, and urine full report did not reveal any significant derangements.

2.1. Imaging Findings. Initially, the child was referred for an abdominal ultrasound scan. It revealed multiple echogenic foci in the portal vein suggestive of intravenous air. In addition, a small amount of free fluid was seen in the abdomen. The stomach was distended with echogenic foci within the gastric wall. Furthermore, distal small bowel loops were collapsed.

A plain radiograph of the abdomen showed grossly distended stomach and proximal duodenum. Linear and circular air lucencies were present in the gastric wall and in the upper duodenum (Figure 1). The portal venous air was not seen in the plain radiograph.

The collective evidence from clinical, basic haematological investigations, plain radiography, and ultrasonography was suggestive of proximal duodenal obstruction. However, the reason for the gastroduodenal emphysema or the obstruction could not be explained by the available imaging findings, and it was decided to proceed with upper gastrointestinal contrast study in view of excluding gastric or midgut volvulus. Although the presence of portal venous air is considered as an ominous sign historically, as the foregut obstruction can cause portal venous air, an upper gastrointestinal contrast study was performed.

The contrast study was carried out using low osmolar nonionic contrast media. The distended stomach and the proximal duodenum with gastroduodenal pneumatoses were seen during the fluoroscopic study. Although the stomach and the proximal duodenum were filled with contrast, no passage of contrast was seen beyond the obstruction (Figure 2). At the site of the obstruction, there was contrast accumulation with ballooning suggestive of a windsock deformity. However, a radiolucent line suggestive of a membrane was not noted.

The diagnosis of proximal duodenal obstruction was substantiated by the clinical and imaging findings. Following these investigations, the child was transferred to a specialized children hospital for the surgical intervention.

2.2. Surgical Findings. During the surgery, the stomach and the proximal duodenum were found to be dilated and the crepitus of the stomach wall was felt. An incision was made...
on the proximal duodenum, and obstruction was felt at the
second part of the duodenum. A thin membrane attached to
the duodenal wall was revealed by a vertical incision made
along the proximal second part of the duodenum (Figure 3).

The web was stretched distally giving rise to a windsock
deformity (Figure 4). The obstruction was proximal to the
bile duct opening. The gastric contents were not bile mixed.
The membrane was excised. The child had an uneventful
recovery with no adverse or unanticipated events recorded
postsurgically.

3. Discussion

The condition of gastroduodenal emphysema associated
with duodenal obstruction secondary to congenital duodenal
web seen in our patient is an exceedingly rare condition in
eyear childhood [10, 11]. Comparison of the present case
with reported cases of gastroduodenal emphysema due to
congenital duodenal web is presented in Table 1.

Amongst the several postulated theories for the devel-
opment of gastric emphysema in infancy secondary to
obstruction, mucosal damage due to mechanical pressure
developed in the distal obstruction is at the forefront. A tear
in the mucosa is thought to be the cause for air tracking in
the submucosa leading to emphysema [10, 15].

It is believed that following the mucosal damage, air
tracking through tissue planes enter portal veins leading to
portal venous air. Although portal venous air is historically
considered to be an ominous sign, this had been described
in more benign conditions like gastric outlet obstruction
[8, 9]. Thus, a more conservative approach to patient
management could be practiced rather than a radical
surgical intervention, if the clinical and biochemical fea-
tures are suggestive of a benign aetiology. Since the clinical
and biochemical evidence did not suggest a grave aetiology
for the gastroduodenal emphysema or portal venous air,
but more of a gastric outlet obstruction in our patient, the
contrast study was performed to determine the site and the
possible cause of obstruction. This also avoided CT
scanning, which would have exposed the child to a higher
radiation dose.

In the literature, gastric emphysema is classified into two
types, viz., emphysematous gastritis and gastric emphysema.
It is important to differentiate these two entities as gastric
emphysema has a benign self-limiting course, whereas
emphysematous gastritis has more grievous course, which
needs prompt intervention [8, 16, 17]. Radiologically, it is
often difficult to differentiate the two entities and the dif-
ferentiation is mainly based on the clinical presentation [1].

Literature suggests that the characteristic radiological
finding in emphysematous gastritis is the presence of streaky
and linear air in the gastric intramural area, whereas more
round gas bubbles are seen in gastric emphysema [1, 17].

The emphysematous gastritis presents with acute abdomen and
often with elevated inflammatory markers. In gastric em-
physema, patients are often haemodynamically stable and do
not demonstrate features of acute abdomen [8, 17]. Our
patient was haemodynamically stable and had no features to
suggest acute abdomen. Thus, careful evaluation by the
clinical team helped in deciding the appropriate radiological
investigations and final patient management.

Duodenal webs are small membranes, which consist of
mucosa and submucosa and are usually seen in the second
part of the duodenum. However, there are a few cases re-
ported in the literature, in which the webs were found in the
third and fourth part of the duodenum [7]. In this patient,
the web was found in the second part of the duodenum.
These are usually attached to the duodenal wall partially,
though circumferential attachment is seen in few instances.
In our patient, the web was attached circumferentially in the
duodenum.

Usually, there is a small central pinhole aperture in the
congenital duodenal membrane. Long-term pressure of
peristalsis against the membrane leads to distal stretching of
the web, forming an intraluminal pseudodiverticulum,
which is known as a windsock diverticulum or windsock
deformity [7].

Duodenal webs can cause complete or partial obstruc-
tion depending on the size of the central aperture. This
affects the age of presentation and also the mode of the
presentation in these patients. Obstruction of the aperture in
the membrane with food particles or gradual onset atony
with ineffective peristalsis against dilated proximal duo-
denum can result in delayed presentation [5]. These webs are
seen as a thin radiolucent line in upper gastrointestinal
contrast studies [10, 18]. In addition, the contrast pooling at
the site of obstruction may give rise to typical windsock
appearance on contrast studies [18]. In our patient, the windsock deformation of the web was noted in the contrast study as well as during the surgery.

The evidence suggests that there is an association between Down’s syndrome and anatomical abnormalities in the duodenum [4, 6]. Most of the reported cases on paediatric gastric

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<td>Progressive, nonbilious vomiting and abdominal distension for 2 days</td>
<td>Mildly dehydrated; abdominal examination: abdominal distension; biochemical investigations: normal</td>
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emphysema with duodenal obstruction have described the condition in children with Down’s syndrome [1, 2, 10, 13]. However, our patient had no chromosomal abnormalities.

Gastric decompression and the treatment for the aetiology of the obstruction are known to relieve the gastric emphysema [1], and this was seen in our patient. Although this patient had normal serum electrolytes, these children usually present with hyponatraemia. Thus, before embarking on any surgical interventions, patient stabilization should be a priority.

Majority of the patients with duodenal webs present early during the first year of life, but atypical presentations as in our patient, should be anticipated. Even though rare, high degree of suspicion should be maintained when a child presents with features of proximal bowel obstruction.

4. Conclusions

This case clearly illustrates the importance of correlating clinical, biochemical, and imaging findings in formulating a final diagnosis and in patient management. Although presence of both the gastric emphysema and portal venous air are concerning, benign aetiologies can also cause these radiological findings and interpretation should be made considering clinical picture. Variable presentations of congenital duodenal webs should be borne in mind, as delayed diagnosis can lead to detrimental effects on the child.

Consent

Informed consent was obtained from the father of the patient to present or publish the findings without revealing the identity of the child.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

Authors’ Contributions

MCW collected all relevant clinical, radiological, and surgical data and performed radiological investigations. SR performed radiological investigations and interpreted results. SK performed the surgery. NDW conducted the literature review. MCW and NDW drafted the initial manuscript. SR and SK were involved in revising it critically for important intellectual content. All authors read and approved the final manuscript.

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