Coexistence of Hypertrophic Pyloric Stenosis and Long-gap Isolated Esophageal Atresia: A Case Report and Review of the Literature

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Abstract

The coexistence of hypertrophic pyloric stenosis (HPS) and esophageal atresia (EA) is a rare condition. In the literature, also, there are only a few publications written about this rare coexistent condition. In this study, an incidental HPS case, which was discovered during gastric pull-up surgery for long-gap isolated EA, is investigated. A 50-day old girl who had isolated EA applied to our clinic for gastric pull-up surgery, which was planned to replace her currently existing gastrostomy which was performed in the newborn period. During the operation, it was seen that the stomach was larger than expected because of HPS. Pyloromyotomy was carried out before gastric pull-up. Following this, the stomach was pulled upwards from the mediastinum and anastomosed with the proximal esophagus. No complication developed during or after the surgery. In cases where patients with EA come with nutrition problems, and also stricture of the anastomosis line or gastroesophageal reflux are excluded, it should be kept in mind that the reason may be HPS.

Keywords: Esophageal atresia, Infantile hypertrophic pyloric stenosis, pediatric surgery

INTRODUCTION

Esophageal atresia (EA) is a congenital pathology. It can present as an isolated defect but is often seen with additional anomalies.¹ Additional anomalies should be investigated in the pre-operative period and detailed preparation should be made for the appropriate treatment of the patient. The EA and hypertrophic pyloric stenosis (HPS) combination seems extremely rare. In the literature, there are few reports where the coexistence of these two pathologies has been described. However, in these reports, it was seen that HPS was diagnosed pre-operatively.²⁻⁶ In this paper, we aimed to present an incidentally diagnosed HPS in a newborn during a gastric pull-up procedure due to long-gap isolated EA.

CASE PRESENTATION

A 50-day-old female patient with isolated EA was referred to our clinic for a gastric pull-up operation. When the patient was born, a nasogastric (NG) tube could not be advanced into the stomach. On the patient’s chest X-ray, no gas mark was seen. The patient was diagnosed with isolated EA. After diagnosis, the patient underwent laparotomy to perform the gastrostomy procedure. No additional anomalies were detected. The patient was discharged after feeding from a gastrostomy tube and referred to a hospital for the gastric pull-up procedure.

When the patient was admitted to our clinic, the patient was being fed via the gastrostomy tube. There was no additional problem seen in the pre-operative examination. Information was given to the family about the coexistence of HPS and EA. The operation was planned to replace the gastrostomy tube with a gastric pull-up. During the operation, it was seen that the stomach was larger than expected because of HPS. Pyloromyotomy was carried out before gastric pull-up. Following this, the stomach was pulled upwards from the mediastinum and anastomosed with the proximal esophagus. No complication developed during or after the surgery.

Keywords: Esophageal atresia, Infantile hypertrophic pyloric stenosis, pediatric surgery

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the procedure and its possible risks. Informed consent was obtained from the family.

At laparotomy, the stomach seemed large and more distended than expected (Figure 1). Intra-operative ultrasound was performed, pyloric muscle thickness was measured at 5.5 mm and pyloric channel length was measured at 17 mm. The pyloric muscle layer was cut parallel to the muscle lines and myotomy was performed. The stomach and distal esophagus were separated and released from the surrounding tissue. The distal esophagus was excised. The gastrostomy site was repaired. Following this, the proximal esophagus was found via neck incision. A retrosternal tunnel was created. The stomach was brought from the longitudinal tunnel. The proximal esophagus and stomach were anastomosed.

On the 4th post-operative day, feeding by NG tube was initiated. On the post-operative 8th day, contrast study was performed. No leakage or luminal stenosis were identified. Additionally, there was no abnormal finding in the pyloric passage (Figure 2). The NG tube was removed and the patient was discharged after oral intake was seen to be adequate.

DISCUSSION

Concomitant congenital anomalies are common in patients with EA. Detection of these anomalies is important in terms of pre-operative evaluation and post-operative follow-up time. Although there are not many cases presented in the literature, retrospective studies have shown that the association of EA and HPS is too high to be negligible.

EA is a life-threatening malformation in newborns and it is associated with morbidity and mortality. Pathologies which are associated with EA may affect prognosis and survival in these patients.7

HPS presents projectile vomiting after feeding in the first 2-12 weeks of age.8 Progressive hypertrophy of the pyloric muscle causes obstruction of gastric emptying. Non-bilious vomiting, visible peristaltic waves in the left upper part of abdomen, palpations of the hypertrophic muscle (olive) and hypokalemic hypochloremic metabolic alkalosis are cardinal symptoms of HPS.8 The accepted criteria for a positive United States study are a pyloric muscle thickness of more than 3 mm, and a pyloric channel length of 15 mm or more.8

In a retrospective study, 267 cases with EA who had been admitted to a clinic during the previous 20 years were evaluated and 24 of them had HPS.8 The authors emphasize that this ratio is actually too high to be ignored and that pyloric stenosis should be considered in patients presenting with vomiting or non-effective feeding after EA repair.8 Carazo Palacios et al.10 presented two cases accompanied by these two pathologies. It was determined that these two diseases were seen together in 2 out of 66 of the patients treated in their clinic. Qvist et al.11 found that 2 out of 74 of these two pathologies accompanied each other in their study. After repair of EA, in less than 8 weeks, patients with symptoms of gastroesophageal reflux and stricture must be investigated for absolute HPS.11 In another case series, the authors presented cases of EA accompanied by HPS over a 5-year period. Four out of 42 patients who were admitted to the clinic during a 5-year period had HPS. Approximately 10% of these two pathologies are seen together.6

The diagnosis of HPS can be made between 2 and 8 weeks, since its symptoms may be delayed.6 Repair of EA is performed in the newborn period unless it is long gap. In the early period after the repair of EA, diagnosis of HPS may be delayed due to the long duration of total parenteral nutrition and the completion of total enteral feeding.5

In our case, the patient’s growth and development were not affected due to the undiagnosed pyloric stenosis. If the patient had not achieved the desired level of development, we might have had to have waited longer for the planned operation.

Figure 1. When laparotomy was completed, the stomach was found to be larger and more distended than expected.

Figure 2. Hypertrophic pyloric muscle.
The stenotic stomach seemed large and wide. In this way, it was easy for the stomach to reach the neck of the patient and a tension-free anastomosis was provided. Although the large stomach was placed in the retrosternal region, there was no respiratory distress due to pressure. The patient also did not have such problems during their early post-operative period.

**Main Points**

- When patients present with vomiting, preliminary diagnoses are primarily due to gastroesophageal reflux and stenosis in the anastomosis line and the first symptom of HPS is vomiting after feeding.

- It should be kept in mind that these two diseases may be seen together in patients with recurrent vomiting during follow-up since the diagnosis of HPS cannot be made in the neonatal period and because symptoms are the same as reflux and stenosis, and these patients should be examined when clinical suspicion occurs.

**Ethics**

**Informed Consent:** Informed consent was obtained from the family.

**Peer-review:** Externally and internally peer-reviewed.

**Authorship Contributions**


**Disclosures**

**Conflict of Interest:** No conflict of interest was declared by the authors.

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**References**


