Pyloric duplication cyst in a newborn with goldenhar syndrome

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Background: Pylorus is a rare location of duplication cysts. Its association with Goldenhar syndrome has not been documented. Presentation is very similar to hypertrophic pyloric stenosis. Imaging studies are required, but diagnosis is done intra-operatively. Complete cyst excision is desired.

Case Presentation: A newborn, Filipino, male presented with non-bilious vomiting and epigastric mass. Congenital scan revealed cardiac anomalies, duodenal atresia, and shortened long bones. Patient was delivered via normal spontaneous delivery. Patient had craniofacial anomalies consistent with Goldenhar syndrome, and an epigastric mass. Patient had multiple non-bilious vomiting post-feeding hence referred to Pediatric Surgery. Ultrasound noted well-circumscribed cystic focus in the epigastrium. Further investigation with abdominal CT scan with contrast revealed well-defined, homogenous cystic focus, non-enhancing mass in the midline of abdomen measuring 4.1 x 3.5 x 3.4 cm inferior to the pancreatic head, in close relation to the pylorus and duodenum. Gastric outlet obstruction secondary to pyloric duplication cyst was considered. Patient underwent exploratory laparotomy with partial excision of mass. Intraoperatively, cystic mass had a shared antimesenteric wall with the pylorus, extending into the first part of the duodenum. Histopathology revealed gastric tissue with pyloric mucosa and intestinal metaplasia without heterotropic tissues. Postoperatively, patient tolerates feeding without significant recurrences of vomiting. Patient's medical problems from his Goldenhar syndrome were managed accordingly. On 36th day of life, patient expired due to septic shock secondary to neonatal pneumonia.

Conclusion: Literatures have described gastrointestinal complications of Goldenhar syndrome but did not include duplication cysts. This is the first paper to present their relationship, and further research work may be done to investigate it further. Nutrition is critical to neonates, immediate referral subspecialty services is needed to provide intervention and relief of obstruction caused by duplication cysts while managing other complications from Goldenhar syndrome.