CASE REPORT

Hypertrophic pyloric stenosis in a newborn: a diagnostic dilemma

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Infants with hypertrophic pyloric stenosis typically present at 2 to 4 weeks of age with non-bilious projectile vomiting. Hypertrophic pyloric stenosis is exceedingly rare in newborn infants and is scarcely reported in literature. Also, the diagnostic criteria for ultrasonographic measurements in newborn infants have yet to be determined. This report is of a newborn infant with hypertrophic pyloric stenosis. The patient presented with high-volume non-bile–stained output from a nasogastric tube and a dilated gastric bubble on abdominal radiograph. Contrast study ruled out intestinal malrotation. Two ultrasound tests showed that the pyloric muscle thickness and pyloric canal length were within normal limits. Subsequent laparotomy showed a thickened pylorus and pyloromyotomy was performed. The patient showed marked improvement in feeding postoperatively. A high index of suspicion is required for newborn infants presenting with gastric outlet obstruction. Ultrasound and contrast studies provide additional information, but definitive diagnosis may only be available intra-operatively.

Introduction

Hypertrophic pyloric stenosis is a well-recognised clinical condition that has an incidence of 1.5 to 4.0 per 1000 live births in Caucasian infants. Hypertrophic pyloric stenosis is less prevalent in African American and Asian children.[1] Infants with pyloric stenosis typically present with projectile, non-bilious vomiting, which may progress to dehydration, progressive weight loss, and characteristic hypochloraemic and hypokalaemic metabolic alkalosis. Pyloric stenosis is treated by pyloromyotomy with excellent prognosis.[1] Although controversy remains as to the aetiology, the typical age of presentation is in the second to fourth weeks of life. Hypertrophic pyloric stenosis is exceedingly rare in newborn infants and is scarcely reported in the literature. This report is of a newborn infant with pyloric stenosis, which was a challenge to diagnose preoperatively.

Case report

A 4015-g boy was delivered by emergency caesarean section for fetal bradycardia at 41 weeks of gestation. He was the first child of a family with a known unbalanced translocation of chromosomes 9 and 15 (46XY der(9)t(9;15)(p22q26.1)) diagnosed by antenatal chorionic villous sampling. Antenatal care showed a suspected congenital heart disease, but was otherwise uneventful. The infant's Apgar score was 10 at 1 and 5 minutes. He had multiple dysmorphic features, including small low-set ears, penoscrotal hypospadias, and right impalpable testis with right hypoplastic scrotum. He developed desaturation and respiratory distress 4 hours after birth. He was intubated and treated for clinical sepsis with intravenous antibiotics. Echocardiogram showed outlet ventricular septal defect and patent ductus arteriosus, and he was treated with prostaglandin E1 infusion for 2 days.

He was subsequently extubated and feeding was introduced on day 4 of life. However, feeding was poorly tolerated with a persistently high output of non-bilious gastric aspirate. Abdominal radiograph showed a distended stomach bubble and paucity of distal gas (Fig 1). Urgent contrast follow-through was done on day 4 of life, and showed slow gastric contrast transit time suggestive of delayed gastric emptying. Owing to the slow gastric emptying, the pyloric canal was not well delineated by the contrast. Subsequent images showed normal duodenojejunal flexure on the left side of the spine and free flow of contrast to the small bowel. The orogastric tube continued to yield a large amount of non-bilious aspirate and the gastric bubble remained distended on abdominal radiograph after the contrast study. Ultrasound of the abdomen done on day 8 of life showed that the thickness of the pyloric muscle (1.5 mm) and pyloric canal length (13 mm) were within normal limits. Ultrasound of the abdomen was repeated on day 11 of life (Fig 2) in view of the lack of clinical improvement, and showed that the pyloric wall thickness and canal length were 2.8 mm and 11.7 mm, respectively. There was no sonographic

Key words
Hypertrophy; Infant, newborn; Pyloric stenosis; Ultrasonography, Doppler

Hong Kong Med J 2011;17:245-7

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evidence of intrinsic mucosal obstruction or extrinsic compression at the distal stomach. Suboptimal peristalsis was also noted. Erythromycin was started on day 12 of life for prokinetic effect but there was no clinical improvement.

In view of the possibility of anatomical pathology causing persistent gastric outlet obstruction, the patient underwent operation on day 16 of life. The procedure started with upper endoscopy, which showed a distended stomach with an inconspicuous pylorus that did not open during the endoscopic examination. Laparotomy was then performed and showed a grossly distended stomach with a thickened pyloric wall. The rest of the bowel was normal. Pyloromyotomy was performed. On-table contrast study following the pyloromyotomy showed passage of contrast through the pylorus to the duodenum by gentle compression on the stomach. Although it is unusual for myopathy or neuropathy to cause delayed gastric emptying alone without affecting the distal intestine, random seromuscular biopsies were taken from the antrum, gastric body, small intestine, and transverse colon to be certain of the diagnosis. Pathology results showed the presence of normal myenteric ganglion cells in all biopsies without hypertrophic nerve fibres. There was no evidence suggestive of visceral myopathy or intestinal neuronal dysplasia on special staining and electronic microscopy.

The patient recovered from surgery without complications. Enteral feeding was resumed on postoperative day 3 and gradually stepped up to full feeding with good tolerance.

Discussion

Although exceedingly rare, hypertrophic pyloric stenosis in newborn infants has been reported.[2-7] The youngest patient was a 7-month fetus with pyloric stenosis demonstrated at autopsy. As Zenn and Redo[5] stated, 5% (range, 0.7-20.0%) of these patients had symptoms at birth, but despite the early symptomatology, surprisingly few cases undergoing operative intervention within the first week of life have been reported. This is probably related to the rarity of the condition at birth and the lack of diagnostic criteria. Therefore, a high index of suspicion is required to diagnose hypertrophic pyloric stenosis. Some patients were diagnosed intra-operatively when the neonates presented with other congenital malformation that required early surgical intervention, such as oesophageal atresia or intestinal malrotation.[6,7] This patient presented with intolerance to feeding with high-output non-bilious gastric aspirate at birth and a distended stomach on abdominal radiograph. Blood gases were not helpful as he had been in the neonatal intensive care unit since birth and his blood parameters had been corrected. Ultrasound of the abdomen was performed twice, and showed the thickness of the pyloric muscle to be 1.5 mm and 2.8 mm, and the length of the pyloric canal to be 13 mm and 11.7 mm.

新生兒的肥厚性幽門狹窄：一個診斷的難題

幼兒的肥厚性幽門狹窄症，一般在出生後第二至四週出現非膽汁性噴射性嘔吐。這種症狀在新生兒中非常罕見，相關的文獻也寥寥可數，而此症在新生兒超聲檢中的診斷標準亦尚未肯定。本文報告一宗新生兒的肥厚性幽門狹窄病例。患者病發時的鼻胃管被發現大量非膽汁性嘔吐物，腹部X光片中也出現擴張型胃泡。對比造影排除腸旋轉不良的可能性。兩次超聲檢均顯示幽門肌層厚度和幽門管長度正常，後經開腹術才發現幽門肌肉肥厚的情況，遂進行幽門肌切開術。術後病人進食明顯好轉。如果新生兒有胃出口梗阻，醫生必需有高度警覺性。超聲檢和對比造影可提供進一步資料，但不排除在手術過程中才可為病人確診。

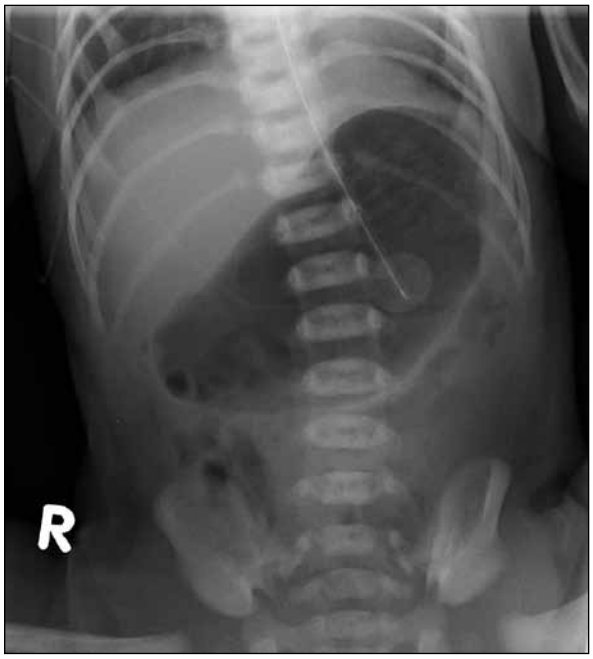

FIG 1. Abdominal radiograph showing a distended stomach bubble with the orogastric tube in situ

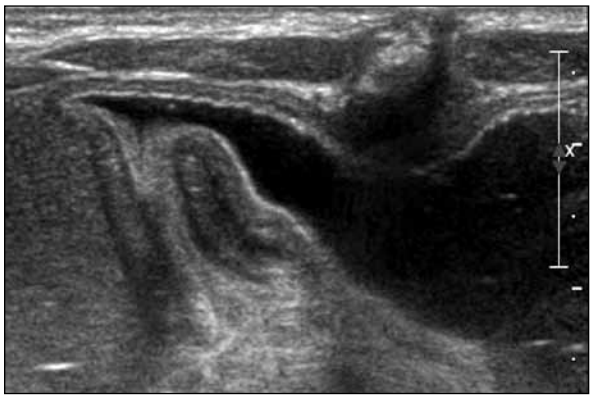

FIG 2. Transverse image of the pylorus using high-resolution linear ultrasound transducer shows prominent pyloric muscle of 2.8 mm thickness. Note the stomach is distended with water and there is no visible gastric emptying during the real-time scanning

The universal diagnostic criteria for hypertrophic pyloric stenosis include muscle thickness greater than 4.0 mm and pyloric canal length greater than 15 mm.[8] However, it is noteworthy that such measurements are applicable to infants born at full-term and who usually present at the age of 4 to 6 weeks. Leaphart et al[9] have suggested a linear relationship of the pyloric muscle thickness and canal length with the patients' age, and have advocated the use of 3.5 mm as a 'cut-off' in patients younger than 21 days old, but to date, there are no reported data on newborns. On the other hand, Keckler et al[10] have raised concerns about the possibility of false-negative results of the initial ultrasound examination as hypertrophic pyloric stenosis is a progressive and dynamic process, and have suggested a low threshold to repeat ultrasound should symptoms persist. In this patient, ultrasound of the abdomen was repeated since there was no clinical improvement. These two ultrasound examinations did show an increase in thickness of pyloric muscle (from 1.5 mm to 2.8 mm), but the parameters were not diagnostic of hypertrophic pyloric stenosis. To date, there are still no universally accepted guidelines on sonographic parameters for the diagnosis of hypertrophic pyloric stenosis in a newborn.

Kosiak et al[11] reported prostaglandin-induced gastric foveolar hyperplasia mimicking hypertrophic pyloric stenosis in neonates. In both patients, the gastric outlet obstruction resolved spontaneously after cardiac surgeries were performed.[11] Callahan et al[12] also discussed prostaglandin-induced foveolar hyperplasia subsequently leading to the development of 'secondary' hypertrophic pyloric stenosis. However, this patient was given prostaglandin infusion for only 2 days, which is much shorter than the periods reported previously to induce gastric foveolar hyperplasia.[12] Moreover, the ultrasound findings were not typical in that the antral mucosa was not thickened.[12] Babyn et al[13] reported that infants who were treated with a prolonged course of prostaglandin had distinct sonographic features of gastric mucosal abnormality. The main feature was papillary or polypoid appearance of the mucosa and submucosa. The enlarged fold surface impinged upon the lumen of the antropyloric region contributing to gastric outlet obstruction.

The intra-operative findings and postoperative course of this patient have confirmed hypertrophic pyloric stenosis to be the cause of the gastric outlet obstruction at presentation. Although the aetiological process of hypertrophic pyloric stenosis usually occurs several weeks after birth, this patient demonstrates that very rarely the pyloric tumour can evolve from the time of birth and the patient presents with a clinical picture suggestive of congenital pathology. Owing to the lack of radiological diagnostic criteria, the diagnosis was made only at the time of surgery. Establishing diagnostic criteria requires a large case volume, which is unlikely to exist for hypertrophic pyloric stenosis presenting at birth as the incidence is exceedingly low. Newborn babies presenting with persistent features of poor gastric emptying associated with distended gastric shadows on repeat abdominal radiograph should raise the suspicion of this rare condition. Although contrast study and ultrasound provide additional information, the final diagnosis may only be made at the time of surgical exploration, which should not be unduly delayed when there is suspicion of gastric outlet obstruction in a newborn infant without obvious causes.

Acknowledgement

We would like to thank Dr Vivian YF Leung for providing the ultrasonographic images.

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