Case Report

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Atypical Presentation of Pyloric Stenosis in Babies in Critical Care

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1. Abstract

Pyloric stenosis usually presents with well defined symptoms and signs. However, these may be absent in neonates in intensive care for other conditions. This paper reports 2 cases of congenital diaphragmatic hernia that developed pyloric stenosis during the critical care period and illustrates the atypical presentation of pyloric stenosis and the diagnostic difficulties encountered in this setting.

2. Keywords:
Congenital diaphragmatic hernia, critical care, pyloric stenosis

3. Introduction

Pyloric stenosis (PS) is a common surgical anomaly. Congenital diaphragmatic hernia (CDH) occurs in 1:2500 births [1]. However both anomalies rarely occur simultaneously. This paper reports on 2 cases of congenital diaphragmatic hernia that developed pyloric stenosis during the critical care period. The atypical presentations and the diagnostic difficulties are outlined.

4. Case Report

4.1. Case 1:
A 1.540 Kg male baby was born by emergency lower segment caesarean section at 33 weeks of gestation following abruptio placenta. An antenatal diagnosis of left sided congenital diaphragmatic hernia was made on a 20 week ultrasound scan. Post delivery the baby was immediately intubated and ventilated. He had an initial stormy period but by day 15 of life was stable enough to undergo surgery. A large left diaphragmatic defect was repaired using Gore – Tex patch. Post operatively the child developed sepsis. He was started on trophic feeds on day 21 of life. He however continued to have brown - coloured aspirates from his nasogastric tube and was commenced on intravenous ranitidine. On day 44 of life an abdominal ultrasound scan was done which showed an elongated non – relaxing pylorus with a thickened pyloric muscle at 4mm. These findings were suggestive of pyloric stenosis but pylorospasm could not be ruled out. A week later, a repeat ultrasound scan showed the elongated pylorus was absent suggesting that the previous appearances were due to pylorospasm. Unfortunately the child developed another bout of sepsis and enteral feeding was stopped.

As the sepsis resolved, small amounts of enteral feeds were commenced but again there were large non - bilious nasogastric aspirates. An ultrasound scan was done on day 70 of life which showed a pyloric muscle thickness of 3mm. A contrast was done on the same date which showed a pylorus which was very slow to open remaining suspicious but not confirmatory for pyloric stenosis. The contrast however showed severe gastro – esophageal reflux. Full antireflux medication was started. The baby started tolerating feeds and was on full feeds by day 90. However he started having large milky vomits which were non - projectile followed by large brown – coloured nasogastric aspirates despite full antireflux medication. His serum electrolytes were normal throughout. A repeat ultrasound scan on day 92 of life showed a definitive pyloric stenosis with a pyloric muscle thickness of 4mm and a canal length of 15mm. The child had a laparotomy on day 93 of life. Laparotomy showed intestinal adhesions and adhesiolysis had to be performed before the pylorus could be adequately exposed. A thickened pylorus was noted and a pyloromyotomy was performed. Unfortunately the baby succumbed to a further bout of chest infection and sepsis and died.

4.2. Case 2:
A 38 week gestation male baby was born by vaginal delivery. Birth weight was 3.08Kg. He had an antenatal diagnosis of left sided CDH at the 20 week anomaly scan. He underwent surgery at 4 days of age and had a patch repair of his diaphragm. Trophic feeds were started at 6 days post op. He however developed non - bilious vomiting at day 12 of life. Blood gases on day 18 of life showed a pH of 7.4 and serum bicarbonate of 34mmol/L. The rest of the electrolytes were normal. An ultrasound scan done at day 19 of life revealed pyloric stenosis. A pyloromyotomy was done the same day. The child was later treated for gastroesophageal reflux. The child survived and was eventually discharged.

5. Discussion

The association of pyloric stenosis and congenital diaphragmatic hernia is rare with only few cases reported in the literature [2,3]. The above 2

cases demonstrate this association in the critical care setting. The children developed pyloric stenosis whilst still on the ventilator post CDH repair. The difficulty in diagnosis here lies in the fact that the classic clinical presentation of PS in these children was absent. There was no projectile vomiting. Abdominal examination did not reveal a mass probably due to the adhesions from the previous surgery. The first child was particularly difficult as the blood gases did not suggest pyloric stenosis. The initial diagnosis in the first child was reflux which may occur after CDH repair [4]. It was only after repeated ultrasound scans that the diagnosis of PS was finally made. The second child did have a blood gas suggestive of PS despite the electrolytes being normal.

This prompted an early ultrasound scan with the diagnosis confirmed earlier than the first child. These cases illustrate the different atypical forms of presentation of PS in babies in critical care. The classic presenting features are absent and blood gases could be normal. The electrolytes would be expected to be normal as these babies are usually on intravenous fluids from birth. Ultrasound scans could be difficult to interpret after neonatal abdominal surgery and repeated scans may be necessary before the diagnosis is confirmed. A recent thirty year review of pyloric stenosis following neonatal thoracoabdominal surgery revealed ten cases with atypical presentation but only one associated with diaphragmatic hernia [5]. The authors however could not offer any specific insight to help with the diagnosis. We feel that a high index of suspicion is needed in achieving a diagnosis in these cases. This paper thus adds to the growing awareness of this condition in patients in critical care.

6. Conclusion

Pyloric stenosis may present atypically in babies in critical care because of other malformations, delayed normal oral feeding and other factors. Ultrasound scans are difficult to interpret immediately after abdominal surgery. Repeated scans and a high index of suspicion are needed to achieve a diagnosis and early treatment.

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