An unusual presentation of infantile hypertrophic pyloric stenosis with severe late-onset neonatal sepsis

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Abstract

A 27 days old neonate presented with poor feeding, severe respiratory distress and septic shock without any palpable abdominal mass. On evaluation found to have features of overt late onset neonatal sepsis and right sided pneumonia. The initial working diagnosis was poorly resolving pneumonia however upon further evaluation found to have blood gas and ultrasonographic evidence of infantile hypertrophic pyloric stenosis (IHPS). Good perioperative care is required to prevent complications.

Keywords: Infantile hypertrophic pyloric stenosis, late onset neonatal sepsis, metabolic alkalosis, ramsted pyloromyotomy

INTRODUCTION

Infantile hypertrophic pyloric stenosis (IHPS) is a common surgical cause of recurrent vomiting in infants. It is characterized by an enlarged pyloric muscular mass and gastric outlet obstruction. In literature, IHPS was first described by Harald Hirschsprung in 1888. The usual incidence varies from region to region and is around 1–4/1000 live births. IHPS is more common in males 4–6:1 and in preterm. The common presentation varies from recurrent nonbilious vomiting and failure to thrive to dehydration and occasionally self-resolving hypertrophy. The usual age for diagnosis varies from 2nd week to 8 weeks. Initially, most infants are dehydrated with weight loss and have an “olive mass” in the right upper quadrant of the abdomen. The diagnosis may be delayed when the presentation is not classic. We report one such case of IHPS in a 4-week-old infant who had features of late-onset neonatal sepsis (LONS) with septic shock and pneumonia.

CASE REPORT

A 27-day-old male neonate referred from a tertiary care hospital with a history of two episodes of vomiting with poor feeding and severe respiratory distress. On arrival, the baby was lethargic and had heart rate 210/min, respiratory rate 80/min, blood pressure 56/38 mmHg, and SPO2 of 89% improving to 93% on non-rebreathing mask. The baby had cold and cyanosed peripheries. Air entry on the right side was reduced with marked intercostal and subcostal retractions. Initially, the neonate was managed for late-onset neonatal sepsis (LONS) with shock with intravenous (IV) fluid boluses, adrenaline, milrinone and dobutamine infusion, and broad-spectrum antibiotics. Investigations are summarized in Table 1. Figure 1 shows chest X-ray depicting...
right-sided pneumonia. Her shock was persistent and hence was intubated on the same day and required higher settings tailored to satisfactory improvement. Echocardiography and abdominal ultrasonography (USG) were normal. His bronchoalveolar lavage done due to poor clinical response revealed numerous pus cells; however, culture was negative. His initial immunological workup including flow cytometry was normal. The infant also required one packed red cell transfusion (lowest Hb 9.1 g/dl) and IV immunoglobulin infusion due to persistent sepsis. The baby was extubated on the 9th day of admission and put on continuous positive airway pressure of 6 cm. The baby continued to be distressed suggestive of protraction toward persistent pneumonia. The infant was discharged against medical advice and got readmitted at our center. In view of significant respiratory distress on arrival, he was put on HHFNC at 6 L/min, FiO2 50%. Investigations revealed polymorphonuclear-leukocytosis with dilated gas-filled single stomach shadow on X-ray [Figure 2]. On suspicion of earlier mixed metabolic acidosis with alkalosis and pure metabolic alkalosis noted at our center, an abdominal USG was carried out with clinical suspicion of IHPS. USG confirmed IHPS with pyloric length of 22 mm and wall thickness of 5.9 mm [Figures 3 and 4]. He underwent Ramstedt’s pyloromyotomy [Figure 5] after metabolic and hemodynamic stabilization. In view of a small intraoperative mucosal tear that was sealed at surgery, enteral feeds were delayed till 72 h postsurgery. The infant made complete recovery and was discharged after 10 days of hospital stay on exclusive breast feeds, consistent weight gain, and without any further complication. The baby was thriving well on follow-up during 60 days postoperative OPD visit.

**DISCUSSION**

This index case highlights the diagnostic conundrum encountered occasionally in the diagnosis of IHPS. In our case, the infant had presented with features of LONS with pneumonia and septic shock with antecedent history of vomiting and poor feeding. On evaluation, he had signs of poor perfusion, high lactate and laboratory features of leukopenia, and increased C-reactive protein. He continued to have features of poorly resolving pneumonia with high delta ratio on ABG with metabolic alkalosis prompting the suspicion of IHPS. This case highlights the need to look through the patient’s clinical profile and blood gases diligently to arrive at an early and correct diagnosis and management.

IHPS is characterized by postnatal hypertrophy of longitudinal and circular muscular layer of pylorus. This results in pyloric swelling merging into antrum and ending abruptly at duodenal end.[4] Earlier this was also called as congenital pyloric stenosis; however, this terminology is not used now. The pyloric canal gets elongated and thickened, hence causing gastric outlet obstruction. The IHPS is more common in males and white population. There was no history of any antiepileptic intake during pregnancy or any neonatal exposure to macrolide antibiotics in this case. Their usages have been associated with development of IHPS. The usual age of presentation is 4–6 weeks, similar to our case.[8] The typical clinical presentation with recurrent nonbilious vomiting and failure to thrive was not present in our case. The clinical and laboratory features in our patient were overwhelmed by pneumonia, LONS, and septic shock. A usually seen feature in IHPS, an epigastric mass or peristaltic wave, was not seen in our case. In a retrospective study by Chalya et al., consisting of 102 patients, only 24% had palpable mass and 44% had peristaltic wave. As in our infant, dehydration was evident in 59% of cases in this series.[6] In our patient, there was no associated anomaly, which is usually seen in up to 33% of cases.[11] Such anomalies are usually seen in central nervous system, gastrointestinal system, urinary

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**Table 1: Baseline investigations**

| Investigation          | Report         |
|------------------------|----------------|
| Hb (gm/dL)             | 13.2           |
| TLC (/µL)              | 2200           |
| Platelet count (/µL)   | 330,000        |
| Creatinine (mg/dL)     | 0.8            |
| C-reactive protein (mg/L) | 57.6        |
| Procalcitonin (ng/ml)  | 0.6 (already on antibiotics) |
| ABG                    | pH-7.28, pCO2 -37.1, HCO3- 20.1, AG-25, delta ratio-3.3 |
| Arterial lactate       | 6.6            |
| Blood culture/BAL culture | No growth     |

Hb: Hemoglobin, TLC: Total leukocyte count, ABG: Arterial blood gas, BAL: Broncho-alveolar lavage

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**Figure 1:** Chest X-ray depicting right-sided pneumonia
tract, and occasionally cardiovascular system. One of the common associations is hyperbilirubinemia, also known as icteropyloric syndrome. They usually have unconjugated hyperbilirubinemia and resolves after surgical correction. It is usually due to decreased level of glucuronyl transferase. Rarely, other coexistent clinical conditions have been described, such as hiatal hernia, peptic ulcer, eosinophilic gastroenteritis, congenital nephrotic syndrome, and congenital hypothyroidism.8

The etiology has been considered multifactorial with interplay between genetic predisposition and environmental causes including use of macrolides and nalidixic acid. On laboratory investigation, they depict varying severity of hypochloremic and hypokalemic metabolic alkalosis with paradoxical aciduria depending on the duration of symptoms.9 Treatment is stabilization of these infants for fluid and electrolyte deficit and definitive surgery. The surgery for IHPS was first described in literature by Ramstedt's in 1912. However, surgery may be delayed in cases of dehydration and electrolyte deficit. Abdominal USG is confirmatory and on transverse sonogram a cutoff of 4 mm in pyloric muscle thickness >15 mm pyloric channel length on horizontal image and a pyloric diameter of 10–14 mm has sensitivity of 95%–98%.9 Occasionally, a contrast study may be required, which depicts an elongated pyloric channel-string sign, a bulge of muscle in antrum-shoulder sign and parallel streak of barium in narrow channel simulating double-track sign.9 Our patient did not require contrast study as he had already confirmatory findings on USG.

The preoperative management consists of correction of fluid and electrolyte imbalances.71 Correction of hypokalemia is required to prevent arrhythmia, and correction of alkalosis is required for the prevention of postoperative apnea.89 Occasionally, for persistent vomiting, nasogastric suction may be required. Most of the patients require surgical management by Ramstedt's pyloromyotomy, standard procedure of choice.111 In the
1990s, laparoscopic pyloromyotomy came in vogue and it has comparative mortality outcome with better cosmetic results.[12] For patients who are poor surgical candidates, oral/IV atropine has shown some favorable outcome with prolonged treatment in up to 80% of cases. At our center, open pyloromyotomy was done. Mucosal perforation has been described rarely and this intraoperative complication results from inadvertent extension of the myotomy beyond the pyloric-duodenal junction and is indicated by bilious fluid appearance. In such cases, mucosal repair is required with omental cover. In the series by Chalya et al., 5.9% of infants had this complication.[6] The overall mortality in IHPS is <0.4%, and with improved preoperative care, it is likely to decrease further.[13]

CONCLUSION

Although IHPS is a common cause of nonbilious vomiting and gastric outlet obstruction in neonates and infants, the usual presentation may not be evident in every case. This case also highlights the meticulousness required in reading the detailed arterial blood gas analysis in the given clinical profile of each case. An early diagnosis and preoperative stabilization of acid base and electrolyte anomaly will improve the outcome with negligible mortality and morbidity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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