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Unusual Presentation of Infantile Hypertrophic Pyloric Stenosis in a Ten-week old Infant

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Summary

Egri-Okwaji MTC, Njokanma OF, Uba CC, Osuoji RI, Fajolu IB. Unusual Presentation of Infantile Hypertrophic Pyloric Stenosis in a Ten-week old Infant. *Nigerian Journal of Paediatrics* **2005; 32: 56.** We report a case of infantile hypertrophic pyloric stenosis in a ten-week old, third-born girl. She presented with sudden onset of non-projectile vomiting of non-bilious material. The vomiting occurred consistently a few minutes after feeds. No peristaltic waves were seen in the abdomen. The typical olive-shaped mass was only felt under general anaesthesia despite repeated examinations. Definitive diagnosis was made with the aid of barium studies and confirmed intra-operatively. This case emphasizes the need for a high index of suspicion for infantile pyloric stenosis in a persistently vomiting infant irrespective of sex, birth order, presence or absence of projectile nature of vomiting, visible peristalsis or palpable abdominal mass.

Introduction

INFANTILE hypertrophic pyloric stenosis (IHPS) is the commonest cause of non-bilious vomiting in infancy. It has an incidence of 3-4/1000 live births in the United States and affects males of first birth order most commonly.[1] Male preponderance has also been reported among Nigerian children but the special predisposition of the first-born is not as prominent as it is in Caucasians.[2] The prevalence of IHPS in Nigeria is also lower than in Caucasians and it has been suggested that environmental, rather than genetic or racial factors are responsible.[2] Ameh and Chirdan[3] showed that IHPS is the seventh commonest cause of neonatal obstructive bowel disease in Nigeria. In another study,[4] it was found to account for 9.4 percent of gastric outlet obstruction. It is inherited in a multifactorial manner in seven percent of cases, among whom 50 percent have an affected mother.[5] Other factors implicated in the aetiology include abnormal pyloric muscle innervation, breastfeeding, maternal stress in the third trimester of pregnancy, increased serum prostaglandins, decreased level of pyloric nitric oxide synthase and infant hypergastrinaemia. It may be associated with certain syndromes such as trisomy 18, Turner, Smith-Lemli-Opitz and Cornelia de Lange.[1]

Since 1976, suggestions have been made about a possible link between erythromycin, a macrolide antibiotic and IHPS.[6] A recent study in the USA found a cluster of seven cases of IHPS among 200 neonates who received erythromycin as prophylaxis for pertussis.[7] Other workers[8] have confirmed this relationship and have reported an eight-fold risk in association with very early exposure. Exposure of the infant to erythromycin in breast milk within ninety days of delivery has also been associated with increased risk of the condition.[9] The early diagnosis of infantile hypertrophic pyloric stenosis requires a high index of suspicion. Any vomiting that is consistently postprandial should be properly evaluated. Early diagnosis is important to prevent electrolyte derangements resulting from the loss of

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hydrogen and chloride rich gastric fluid as well as to circumvent ensuing haemodynamic instability and minimize hospital stay.

In this communication, we present the case of a 10-week old baby with IHPS in order to highlight the diagnostic dilemma that sometimes plagues the clinical presentation of IHPS and the complications of surgical intervention.

Case Report

NE, a 10-week-old girl and the youngest of three siblings, presented at the outpatient department of Havana Specialist Hospital, Lagos, with one-day history of vomiting and excessive crying. She had vomited seven times on the first day and by the second day was vomiting consistently after every feed and occasionally, irrespective of feeds. Vomitus was non-projectile and non-bilious. The last time she passed stools was two days prior to presentation. Before then, she was known to be often constipated with bowel motions of once in three days and the stools were described as usually very hard. She was being exclusively breastfed. The baby had never been treated with erythromycin neither was the mother treated with the drug during or after pregnancy.

Physical examination revealed an afebrile, irritable child who was consoled only by suckling. The abdomen was full and soft. Repeated examination on test feeding showed neither visible peristaltic waves nor a palpable abdominal mass. Test feeding was followed three to five minutes later by non-projectile, non-bilious vomiting. Examination findings in other systems were not remarkable. Plain abdominal radiography showed gastric distension and paucity of bowel gas. Barium meal showed complete gastric outlet obstruction with failure of contrast to appear in the duodenum six hours post ingestion (Fig 1). Full blood count, serum electrolytes, blood urea and creatinine were normal except for mild hypochloraemia (chloride = 94mmol/L) and marginal hypokalaemia (potassium = 3.4mmol/L). The clinical impression was then infantile hypertrophic pyloric stenosis.

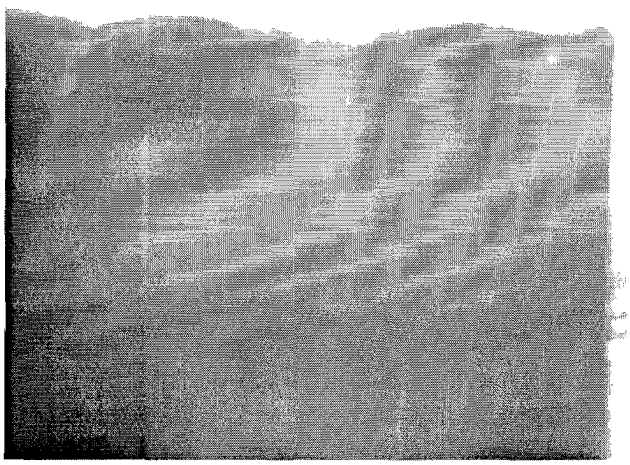

Fig. 1 Radiogram taken 6 hours after ingestion of contrast showing no contrast beyond the grossly dilated stomach

Just before laparotomy, repeat abdominal palpation under anaethesia revealed the typical olive-shaped pyloric mass. The diagnosis of hypertrophic pyloric stenosis was confirmed intra-operatively and pyloromyotomy (Fredet-Ramstedt operation) was done. There was an accidental incision into the pyloric lumen which was repaired immediately in two layers of catgut sutures. Postoperative management included intravenous fluids, intravenous ceftriaxone, metronidazole and intramuscular pentazocine. The baby's haemoglobin level dropped from 9.1g/dl to 7.3g/dl for which she was transfused with 60ml of whole blood.

On account of the accidental incision into the gut lumen, oral feeds were not re-commenced until after about 72 hours. The patient however, resumed vomiting soon afterwards, necessitating a withdrawal of feeds 24 hours later. The vomitus this time was bilious. Bowel motion resumed on the third post-operative day: the first stool was dark and copious but subsequent motions were loose and greenish with a frequency of three or four times daily. On the fifth post-operative day, her abdomen became distended with minimal bowel sounds. A repeat serum electrolyte evaluation revealed further decline in potassium level to 2.4mmol/L and chloride to 92mmol/L. A clinical diagnosis of paralytic ileus was made and supplemental potassium was added to the intravenous fluid at 3mmol/kg of body weight per day. The baby showed remarkable response with resolution of features within twenty four hours. Repeat investigations forty eight hours later showed improvements in potassium and chloride levels to 3.5mmol/L and 96 mmol/L, respectively. Feeds were then re-introduced without adverse sequelae. She was discharged on the 12th postoperative day with clinical and laboratory evidence of full recovery.

Discussion

This case illustrates the need for a high index of suspicion for IHPS despite the absence of "classical features". Typically, the clinical features of IHPS may begin to manifest as early as two days or as late as five months of life. Vomiting is often the earliest symptom and the vomitus is invariably non-bilious.[1] In these respects, the presentation of our patient was 'typical'. On the other hand, the baby was a female rather than a male and the youngest rather than the oldest sibling in a family with neither an affected parent nor sibling. Also untypical was the patent absence of the expected projectile nature of vomiting. Furthermore, despite repeated examinations and diligent search, no visible peristaltic waves moving from left to right across the upper abdomen were seen. The classical pyloric lump

could only be felt when the baby was under anaesthesia. It is possible that the lump had become appreciable as a result of better relaxation of the anterior abdominal wall musculature. The main feature therefore, that sustained the clinical diagnosis of IHPS was the fact that the baby vomited consistently after every meal.

The absence of a 'typical' presentation necessitated the recourse to radiographic investigation. Although gastrografin whose toxic effects are milder than those of barium should there be aspiration into the respiratory tract, is usually preferred to barium as a contrast medium for upper gastrointestinal studies in children, the former was not available at the time of the investigation. The expected pattern was that of a 'string sign' indicating the presence of contrast medium in the narrow pylorus.[1] However, the thick barium paste probably blocked an already narrow canal resulting in a severe degree of gastric outlet obstruction with no barium at all in the pylorus as late as six hours after ingestion. With reference to the mucosal perforation which occurred in the patient, the use of barium may have been an advantage over gastrografin since the latter causes much more severe peritonitis. Faced with the radiographic evidence of complete gastric outlet obstruction, the regurgitant, rather than projectile nature of vomiting remains enigmatic. Ultrasonography might also have been helpful in making a diagnosis by showing an elongated (>14mm) or thickened (>4mm) pylorus, but this investigation was not done.

The re-introduction of feeds 72 hours post-operatively must have been premature considering the accidental incision and therapeutic suturing of the gut. Ensuing secondary vomiting, this time bilious in nature, led to hypochloraemia and hypokalaemia, with the latter leading to the worsening of paralytic ileus. Indeed, accidental mucosal breach at pyloromyotomy is not unusual. An earlier report of this complication was made by Eke[10] in 2001. The patient in that report subsequently experienced post-operative abdominal distension but for a different reason: he developed pyloric fistula at a site different from the operative mucosal breach.

Infantile hypertrophic pyloric stenosis should be differentiated from gastro-oesophageal reflux disease, raised intracranial pressure, duodenal atresia and volvulus neonatorum. Duodenal atresia and volvulus neonatorum present with bilious vomiting and duodenal atresia will also typically present with a "double-bubble sign" on abdominal radiography. Gastro-oesophageal reflux and raised intracranial pressure may be differentiated by the absence of a pyloric lump and radiographic findings.[1]

The treatment of choice is Fredet-Ramstedt pyloromyotomy in which the pyloric muscle is split to allow the intact mucosa bulge through. When done early, the prognosis is good with an associated mortality rate of 0.5 percent or less.[1] Some workers[11] have used non-surgical techniques in the management of this condition. They include a regimen of small, frequent feeds of thickened cereal, maintenance in an upright posture for about one hour after feeds, sedation, administration of anti-cholinergic drugs and parenteral fluids. This option is fraught with problems of slow recovery (two months to eight months), prolonged hospitalization and its attendant adverse effects of the emotional development of the child, higher mortality and high cost.[12] Furthermore, the chances of failure are high whereupon the baby would still have to undergo surgery.

References

1. Wyllie R. Pyloric stenosis and other congenital anomalies of the stomach. In: Behrman RE, Kliegman RM, Jenson HB, eds. Nelson Textbook of Pediatrics. Philadelphia: WB Saunders Company, 2000: 1130–1.
2. Johnson A, Adekunle OO. Congenital hypertrophic pyloric stenosis in Nigeria. *Trop Geogr Med* 1976; **28**: 191-3.
3. Ameh EA, Chirdan LB. Neonatal intestinal obstruction in Zaria, Nigeria. *East Afr Med J* 2000; **77**: 510–3.
4. Dogo D, Yawe T, Gali BM. Gastric outlet obstruction in Maiduguri. *Afr J Med Sci* 1999; **28**: 199–201.
5. Wastell C. The stomach and duodenum. In: Mann CV, Rusell RC, Williams NS, eds Bailey and Love's Short Practice of Surgery. London: Chamman and Hall Medical, 1995: 673–5.
6. SanFilipo JA. Infantile hypertrophic pyloric stenosis related to ingestion of erythromycin estolate: a report of five cases. *J Pediatr Surg* 1976; **11**: 177–80.
7. Honein MA, Paulozzi LJ. Himelright IM, Lee B, Cragan JD, Patterson L, Correa A, Hall S, Erickson JD. Infantile hypertrophic pyloric stenosis after pertussis prophylaxis with erythromycin: a case review and cohort study. *Lancet* 1999; **354**: 2101-5.
8. Cooper WO, Griffin MR, Arbogast P, Hickson GB, Gautam S, Ray WA. Very early exposure to erythromycin and infantile hypertrophic pyloric stenosis. *Arch Pediatr Adolesc Med* 2002; **156**: 647-50.
9. Sorensen HT, Skriver MV, Pedersen L, Larsen H, Ebbesen F, Schonheyder HC. Risk of infantile hypertrophic pyloric stenosis after maternal post natal use of macrolides. *Scand J Infect Dis* 2003; **35**: 104-6.
10. Eke N. Postoperative tension pneumoperitoneum in an infant. *Pediatr Surg Int* 2001; **17**: 204-5.
11. Kawahara H. Imura K, Nishikawa M, Yagi M, Kubota A. Intravenous atropine treatment in infantile hypertrophic pyloric stenosis. *Arch Dis Child* 2002; **87**: 71-4.
12. Hamilton RJ. Congenital and perinatal anomalies of the gastrointestinal tract and intestinal obstruction. In Behrman RE, Heigman RM, Jensen HB, eds. Nelson Textbook of Pediatrics. Philadelphia: WB Saunders Company, 1992: 948–50.