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The diagnostic value of the triple bubble sign in proximal jejunal atresia: a case report

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Summary

Proximal jejunal atresia (PJA) is a common cause of intestinal obstruction in the newborn. Despite the need for an early surgical intervention to minimize morbidity and mortality, a timely identification is frequently precluded by the absence of specific clinical and investigative clues. Against the background of the limitations in making a timely diagnosis of PJA in a tropical setting, where opportunities for high-tech imaging tools are few, we report the diagnostic value of the "triple bubble" sign on the plain radiograph of a Nigerian infant. This radiologic finding led to an early diagnosis and ultimately a prompt surgical extirpation. The paper suggests that the presence of this sign should be a pointer to an early diagnosis of PJA.

Keywords: *Jejunal atresia; "Triple bubble" sign; radiological diagnosis.*

Résumé

L atrèsie proximale jèjunale (PJA) est une cause commune de l'obstruction intestinale chez les nouveau nès.La nècessitè d'une chirugie d'intervention urgente pour minimiser les symptomes et la mortalitè , et une indication a temps est frequetement inachevèe par l'absence des cliniques spècifiques et viable d 'investigation. En vue de ces limitations de faire un diagnostie a temps du PJA dans les zones tropicales où les opportunitès d'emploi des outils de qualitè supèrieur sont limitès. Nous avons reportè la valeur diognostique de signe de triple bubles sur le fichier radiographique d'un enfant nigèrian. Cette radiographie a conduit à un diagnostie tot et ultimement à une chirugie effective. Ce papier suggère que la presence de ce signe pourrait etre une indicateur du premier symptome de l'astrèsie proximale jèjunale.

Introduction

Jejunal atresia is an entity within a spectrum of congenital intestinal anomalies, which include ileal and colonic atresia among others With a prevalence rate of about 1 per 3000 live births [1], it probably constitutes one of the commonest congenital anomalies of the alimentary canal. In affected infants, the typical clinical features of intestinal obstruction are frequently absent [2], making early surgical suspicion difficult. However, a careful radiologic evaluation of such infants constitutes an indispensable diagnostic tool.

A number of radiologic features which had been described in congenital intestinal atresia [3-5] include massively dilated bulbous terminal segment, fetal ascites and echogenic bowel, usually described as more echogenic than liver or bone. Radiologic evaluation of plain abdominal radiographs, ultrasonography and contrast studies constitute the common investigative practice in a resource-poor community like ours. Furthermore, many of these previously described radiological features are not entirely specific. While most clinicians are familiar with the diagnostic utility of the "double bubble" sign in duodenal atresia [6] the potential diagnostic value of the radiographic "triple bubble" sign in isolated proximal jejunal atresia (PJA) had eluded appropriate emphasis to date. A recent review of the literature showed only two relevant citations [3,7] on the potential diagnostic value of the sign. Inded, neither of these two reports emanated from sub-Saharan Africa, where facilities for modern imaging modalities remain limited or absent.

The present report, borne out of our experience in managing a Nigerian infant with isolated PJA, highlights the diagnostic importance of the "triple bubble" sign on a plain abdominal radiograph.

Case report

Baby A.A, was a full -term male singleton born to unrelated Nigerians. He was a product of normal vaginal delivery after an uncomplicated pregnancy. The patient presented at the University of Ilorin Teaching Hospital, Ilorin, Nigeria on the second day of life, with progressive abdominal distention from birth, recurrent vomiting, and failure to pass meconium. Prenatal clues of possible polyhydramnios were denied. Sonographic findings at the gestational age of 28 weeks were normal. The vomiting was said to be projectile, and the vomitus was essentially bile stained with recently offered feed/gastric contents. He was said to be making urine, and the stream had been noticeably good.

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At presentation, the baby was apparently uncomfortable and was crying inconsolably. On physical examination, he was neither pale nor jaundiced, and the hydration status was adjudged sub-optimal. The admission temperature was 37.0 °C. The abdomen was grossly distended, soft and tympanitic; and there were no visible peristalsis. No palpable abdominal mass or demonstrable ascites were noted. Widespread crackles were heard over both hemithoraces, especially at the lung base. A working diagnosis of intestinal obstruction was made, and this was localized to the upper segment, while the chest findings were attributed to an associated pneumonitis, presumably from aspiration of gastric content.

A sepsis work-up revealed normal white blood cell count and absence of significant growth of bacteria on blood culture. An abdominal ultrasound showed grossly dilated small bowel loops filled with fluid and gas. No increased intra-peritoneal fluid was demonstrated. The liver, spleen, and both kidneys were essentially normal. His biochemical profile showed a mild to moderate hyponatraemia, hypochloraemia, and alkalosis. Serum creatinine was within the normal reference values. The chest radiograph revealed widespread inflammatory opacities in both lungs. The heart, bony thorax and soft tissue were also essentially normal. A plain abdominal radiograph (Fig 1 & 2) showed a grossly distended abdomen,

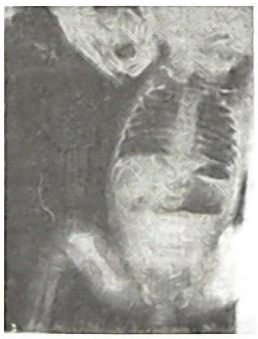

Fig. 1: The "triple bubble" sign on the erect plain antero posterior abdominal radiograph.

the lower half of which was devoid of gas. Three distinct air fluid levels, consistent with the "triple bubble" sign were evident in the upper half. Associated meconium ileus, with or without perforation and meconium peritonitis were excluded by the absence of a right lower quadrant "ground glass" appearance, flank calcification sub-diaphragmatic peritoneal gas[1,6]. Based on the presence of this sign,

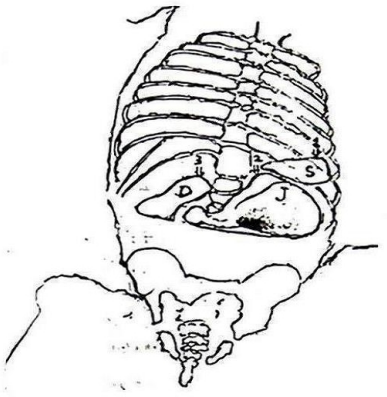

Fig. 2: Schematic diagram of the plain abdominal radiograph as shown in Figure 1. The "triple bubble" sign are as indicated by the numbers.

Key

1.....Bubble 1,	2.....Bubble 2
3.....Bubble 3,	S.....Stomach
D.....Duodenum,	J.....Jejunum

and the subsequent findings, of dilated and bulbous terminal segment of proximal jejunum, from the limited barium studies (Figure 3), a pre-operative diagnosis of an

Fig. 3: The barium meal radiograph of the same patient showing dilated stomach with abrupt termination of the barium column at the proximal jejunum.

isolated proximal jejunal atresia was made. This was corroborated by the surgical findings as those of a Type II atresia of the proximal jejunum, in which the blind ends of the jejunum were separated by a fibrous cord [6]. The atretic segment was resected and an end-to-end anastomosis carried out. The intra- and post-operative managements were uneventful, and the baby was discharged on the 10th

post-operative day. Follow-up evaluations over a period of six months at the Paediatric Surgical Clinic showed that the infant was thriving well.

Discussion

Jejuno-ileal atresia is the commonest cause of congenital intestinal atresia and also the most frequent cause of neonatal intestinal obstruction [1,8]. In affected patients, there is no known gender skewing in the incidence [9], but when compared with dizygotic twins and singletons, monozygotic twins are reportedly at a higher risk [6]. The conspicuous absence of prenatal clues of polyhydramnios in the current case is consistent with the reported incidence of only 25% of cases [6]. The association of PJA with cystic fibrosis, renal dysplasia, atresia of the appendix and biliary tract have been reported ealier [10-13]. None of these was however present in this case.

Various workers have dwelt on the pathogenesis of jejuno-ileal atresia [2,6]. It has been ascribed to a late intrauterine mesenteric vascular occlusion, presumably from intrauterine accidents like volvulus, malrotation, internal hernia, intussuceptions or strangulation in a tight abdominal defect [2,6]. Pre-natal parvovirus B19 infection and genetic predisposition have been suggested in more recent reports [13-15].

Jejuno-ileal atresia has been classified into four major anatomic types [16]. The nature of the lesion(s) varies from a luminal diaphragm associated with the type I variety, to the rare type IV that is characterized by the presence of multiple atretic segments of the bowel. The type II variety which was discovered in this patient at surgery was associated with a solid cord connecting the proximal and the distal bowel loops. Though the type IIIa is associated with blind intestinal loops, the typical anatomic defect in the type IIIb is the so called "apple peel" appearance, in which extensive mesenteric defect is associated with ischaemia in the distal ileum [6,16].

Unlike duodenal atresia [6], the spectrum of possible anatomic lesions in PJA underscores the need for exploring cost-effective, but sufficiently discriminative radio-diagnostic tools. Furthermore, in view of the characteristic radiological findings of co-existing morbidities and differential considerations, imaging modalities remain essential pre-operative diagnostic tools of jejuno-ileal atresia. In our setting, cost considerations and the paucity of radiologic expertise, favour plain abdominal radiographs and ultra-sonographic studies.

The "triple bubble" sign is usually demonstrable on the erect plain abdominal radiograph after the first 4 hours of life [1]. It has been attributed to the natural contrast provided by swallowed air, with the consequent formation of intra-luminal gas-fluid levels in the dilated stomach, duodenum and proximal jejunum. This is unlike ileal atresia where the bubbles are more in number and the dilated intestinal loops may be difficult to differentiate from the ahaustral colon of the neonate. The diagnostic importance and the ease of identifying the "triple bubble" sign on the erect plain abdominal radiograph in proximal jejunal atresia is therefore a noteworthy clue for practitioners in our environment. Although a prospective radiologic series would be necessary to clarify its usefulness in types III and IV varieties of jejuno-ileal lesions, our experience in this case suggests that the "triple bubble" sign is almost pathognomonic, and may require no further radiologic studies [3,9,17]. The progressive jejunal distention, which could lead to gangrene and perforation [18], constitutes valid reasons for pursuing an early diagnosis and surgical intervention. Significant dehydration, hypovolaemia, acid-base aberrations and shock are possible additional consequences of a delayed diagnosis.

In conclusion, this case emphasizes the importance of the "triple bubble" sign on the plain abdominal radiograph in the diagnosis of PJA. We therefore suggest that the presence of this radiologic sign in the "gasless abdomen of the newborn"[3] should prompt the need for immediate confirmatory contrast studies, early surgical evaluation and intervention.

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