The Triple Bubble Sign: A Neglected Radiologic Feature of Proximal Jejunal Atresia

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Citation

A Amole, R Johnson, O Adesiyun. *The Triple Bubble Sign: A Neglected Radiologic Feature of Proximal Jejunal Atresia*. The Internet Journal of Radiology. 2002 Volume 3 Number 1.

Abstract

Proximal jejunal atresia (PJA) remains a common cause of intestinal obstruction in the newborn. Despite the need for an early surgical intervention (to preempt associated morbidity and indeed occasional mortality), a timely identification is frequently precluded by the absence of specific clinical and investigative clues. Against the background of the limitations of making a timely diagnosis of PJA in a tropical setting where opportunities for high-tech imaging tools are few, we report here the diagnostic utility of the "triple bubble" sign on the plain radiograph of a Nigerian infant. As was the case in this baby, who had non-specific symptom-complex of upper alimentary tract obstruction, the radiologic finding of the "triple bubble" sign led to an early confirmation with the more invasive barium study, and ultimately a prompt surgical extirpation. In the tropical health facility with limited radiologic service and expertise, we proffer that the presence of this sign constitutes an invaluable and near-pathognomonic clue of an anatomic/surgical, rather than a "medical" cause like neonatal sepsis. The clinical clues of PJA, possible genesis of the "triple bubble" radiologic sign, and the difference(s) from the findings in other congenital causes of anatomic obstruction of the alimentary tract are highlighted briefly under the discussion.

INTRODUCTION

Jejunal atresia is an entity within a spectrum of congenital intestinal anomalies, which includes ileal and colonic atresia among others. With a prevalence rate of about 1 per 3000 live births₁, 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 absent₂. Hence, in pursuing the desirable early surgical intervention, a careful radiologic evaluation of such infants constitutes an indispensable diagnostic tool. Whereas a number of radiologic features had earlier been described in congenital intestinal atresia3,4,5, radiographic evaluation of plain abdominal radiographs, ultrasonography and contrast studies constitute the most practicable 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 atresia₆, the potential diagnostic value of the radiographic "triple bubble" in isolated proximal jejunal atresia (PJA) had eluded appropriate emphasis to date. A recent and extensive search surprisingly showed only two relevant citations³, on the potential diagnostic value of the this latter sign. Indeed, neither of these two reports emanated from tropical Africa,

where opportunities for modern high-tech imaging modalities remain few. The present report, borne out of our recent experience in managing a Nigerian infant with isolated PJA, highlights the diagnostic importance of the "triple bubble" sign on the plain abdominal radiograph.

CASE REPORT

Baby A.A, was a full-term male singleton. He was the product of a normal vaginal delivery after an uncomplicated pregnancy. Specifically, prenatal clues of possible polyhydramnios were denied, and this was consistent with the normal sonographic findings at the gestational age of 28 weeks. Both parents were Nigerians, and were unrelated. Presentation at our Health Facility was on the second day of life, with the major parental concerns comprising progressive abdominal distention from birth, recurrent vomiting, and failure to pass meconium. The vomiting was said to be projectile in nature, and vomitus was essentially bile stained recently offered feed/gastric contents. He was said to be making urine, and the stream had been noticeably good.

At presentation, the baby was apparently uncomfortable and was crying inconsolably. He was however neither jaundiced nor pale, but the hydration status was adjudged sub-optimal. The admission temperature was 37.00C. Although the abdomen was grossly distended, there was no visible peristalsis, and it was soft and tympanitic. Abdominal mass(es) were absent, as were physical signs of ascites. Widespread crackles were heard over both hemithoraces, especially posteriorly. A working diagnosis of intestinal obstruction was made, and this was localized to the upper segment, and the chest findings were attributed to an associated pneumonitis, presumably from pre-consultation aspiration of gastric contents

A sepsis work-up revealed the absence of haematologic and microbiologic clues of neonatal septicaemia. The subsequent abdominal ultrasound (using a Siemens® Sonoline Sx machine with a 5MHz mechanical sector transducer), however showed a grossly distended abdomen with features of exaggerated peristalsis. The bowel loops were dilated and filled with fluid and gas. No intra-peritoneal (ascitic) fluid was demonstrated. The liver, spleen and both kidneys were essentially normal. The biochemical profile showed a mild/moderate hypo-natraemia, hypochloraemia, and alkalosis. The serum creatinine was within the local reference values, and this was adjudged consistent with the normal urinary output and stream. The clinical features of pneumonia were corroborated by the presence on the chest radiograph, of widespread inflammatory opacities in both lungs. The cardiac silhouette was however of normal size and shape. The bony thorax and soft tissue were also essentially normal. A plain abdominal radiograph (Figure I the individual "bubbles" are highlighted in Figure II) showed a grossly distended abdomen, the lower half of which was devoid of gas.

Figure 1

Figure I. The "triple bubble" sign on the erect plain abdominal radiograph.



Figure 2

Figure II. Illustration of the erect abdominal radiograph highlighting the three bubbles of gas.



Three distinct air fluid levels, consistent with the appearance were however evident in the upper half. A de novo, or associated meconium ileus, with or without perforation and meconium peritonitis were largely excluded by the absence of a right lower quadrant "ground glass" appearance, flank calcification and sub-diaphragmatic peritoneal gas¹,⁶. Based on the presence of the "triple bubble" sign, and the subsequent findings from the limited barium meal studies (Figure III),

Figure 3

Figure III. The barium meal radiograph of the same patient confirming a proximal jejunal obstruction.



A pre-operative diagnosis of an isolated proximal jejunal atresia was made. This was corroborated by the intraoperative findings, which were consistent with those of a Type II atresia of the proximal jejunum, in which the blind ends of the jejunum were separated by a fibrous cord⁶. The atretic segment was resected and an end-to-end anastomosis carried out. The intra- and postoperative managements were uncomplicated, and the baby was discharged on the 10th postoperative day. Follow-up evaluations (three so far) at the Paediatric Surgical Clinic showed that the infant has been thriving to date.

DISCUSSION

Jejuno-ileal atresia is the most common cause of congenital intestinal atresia, and the most frequent cause of neonatal intestinal obstruction¹,₈. In affected patients, there is no known gender skewing in the incidence₉, but when compared with dizygotic twins and singletons, monozygotic twins are reportedly at a higher risk⁶. The conspicuous absence of prenatal clues of polyhydramnios in the current case was consistent with the reported presence in only 25% of cases⁶. The association of PJA with cystic fibrosis (an unlikely underlying disease in an indigenous black African infant such as the current case), renal dysplasia, atresia of the appendix and biliary tract have been reported earlier_{10,11,12,13}. Gladly, none of these was present in the current case.

The pathogenesis of jejuno-ileal atresia has been highlighted earlier^{2,6}. The lesion has been ascribed to a late intrauterine mesenteric vascular accident, presumably from intrauterine accidents like volvulus, malrotation, internal hernia, intussusceptions or strangulation in a tight abdominal wall defect^{2,6}. Pre-natal parvovirus B19 infection and genetic predisposition have been suggested in more recent reports¹³,_{14,15}. With the recent mapping of the human genome, it is only a matter of time before the controversy on the genetic basis of this anomaly is finally laid to rest.

Jejuno-ileal atresia has been classified into four major anatomic types₁₆. The nature of the lesion(s) varies from a luminal diaphragm associated with the type I variety (which is responsible for 20% of cases), to the rare type IV which is characterized by the presence of multiple atretic segments of the bowel, and accounts for only 5% of jejuno-ileal lesions. The type II variety, which was identified at surgery in the present case, is reportedly associated with a solid cord connecting the proximal and the distal bowel, and together with type IIIa, they account for $\sim 70\%$ of cases⁶,¹⁶. Whereas 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. Unlike duodenal atresia⁶, the wide spectrum of possible anatomic lesions in PJA (the clinical correlates of which are hardly specific) 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 (e.g. the "ground glass" glass appearance of meconium ileus, sub-diaphragmatic gas of a complicating perforation, and the early peritoneal calcification of meconium peritonitis⁶,₁₇) imaging modalities remain indispensable pre-operative diagnostic tools of jejuno-ileal atresia. In our setting, cost considerations and the paucity of radiologic expertise, would favour exploring (suggestive clues from) the plain abdominal radiograph, and ultrasonographic studies.

The "triple bubble" sign (which proved invaluable in the pre-operative diagnosis of PJA in this infant) is usually demonstrable on the erect plain abdominal radiographs after the first 4 hours of life¹. 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 in ileal atresia where the dilated intestinal loops may be difficult to differentiate from the ahaustral colon of a neonate. In view of its diagnostic import, the availability of the required facilities, and the simplicity of its identification, the demonstration of the "triple bubble" sign on the erect plain abdominal radiograph in proximal jejunal atresia is clearly a noteworthy diagnostic clue for Practitioners in third world settings. Although a prospective radiologic series would be necessary to clarify the usefulness in the more severe anatomic varieties of types III & IV jejuno-ileal lesions, our experience with this case suggests that the "triple bubble" sign (which interestingly follows on the numerical and anatomic heels of the "double bubble" sign of duodenal atresia) is near-pathognomonic, and may require no further radiologic studies^{3,9,17}. That the progressive jejunal distention could compromise the vascular supply of the intestinal wall, leading to catastrophic consequences like gangrene and perforation₁₈, constitute valid reasons for pursuing an early diagnosis and surgical intervention. Significant dehydration, hypovolaemia, acid-base aberrations, and shock are possible additional consequences of a belated diagnosis.

In conclusion, the present case suggests a potential diagnostic utility for the "triple bubble" sign on the plain abdominal radiograph of an infant with symptom-complex of congenital upper gastrointestinal obstruction. We submit that in a resource-poor tropical setting, the presence of this radiologic sign "in the gasless abdomen of the newborn" ³ should prompt the need for urgent confirmatory contrast studies, surgical evaluation and intervention as appropriate.

ACKNOWLEDGEMENTS

We acknowledge and salute the usual team spirit that subsisted between the Radiologists, Radiographers, Paediatricians and the Paediatric Surgeons before, and after the operative intervention in Baby A. A.

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