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Twins with congenital hypertrophy of infantile pyloric stenosis

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ABSTRACT

Congenital hypertrophy of infantile pyloric stenosis (CHIPS) is an acquired condition where the thickened pyloric sphincter causes high grade gastric outlet obstruction. The incidence is higher in the West compared to the East. Over a 15 years period, there has only been a single case of pyloric stenosis in a patient of Caucasian descent in our local setting. A rare and interesting set of six weeks old male twins with CHIPS is reported. Both patients responded well to pyloromyotomy.

Keywords: Gastric outlet obstruction, pyloromyotomy, pyloric stenosis

INTRODUCTION

Congenital hypertrophy of infantile pyloric stenosis (CHIPS) is actually an acquired condition where the pyloric sphincter becomes hypertrophied and causes high grade gastric outlet obstruction. CHIPS typically manifest from three to six weeks after birth with recurrent projectile vomiting leading to dehydration, hypoglycaemia and metabolic alkalosis. The incidence is much higher in the West with rates of between 0.2 to 0.4% of all live birth compared to the East. [1] In RIPAS Hospital, there has only been a single case (0.001%) of pyloric stenosis in a Caucasian descent over a period of 15 years (from 1984 to 1999). A rare and interesting set of six weeks old male twins with CHIPS is reported. Both responded well to pyloromyotomy.

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CASE REPORT

A set of six-week-old male Malay identical twins from the Kuala Belait District was referred to the Department of General Surgery in RIPAS Hospital with symptoms of projectile, non-bilious vomiting and dehydration. The gestations and birth were uncomplicated. One of the twins (Twin 1) was found to have a tender right inguinal hernia and was operated on for suspected obstructed right inguinal hernia on the day of admission. The other twin (Twin 2) was managed conservatively. Despite this, both twins failed to settle and continued to have persistent projectile vomiting. At this time, repeat examinations showed a palpable olive-sized mass present in the epigastrium of the abdomen in both twins. Coincidentally, a non-tender reducible right inguinal hernia was also detected in Twin 2. A differential diagnosis of pyloric stenosis in both twins was made and confirmed with ultrasound scan.

Both twins continued to receive intravenous hydration and on the fifth day of admission were operated on. Laparotomy via upper transverse abdominal incisions on both twins revealed pyloric stenosis (Figure 1a), Ramstedt's pyloromyotomy (Figure 1b) was performed for each of the twins. This was carried out via a small superficial incision of the pylorus on the serosa at the anterior surface from the antrum to a point proximal to the duodenum. The myotomy was then deepened bluntly to the submucosa and the overlying muscle fibres were gently spread and mobilised to allow the submucosa and mucosa to bulge out to the level of the serosa. Care was taken to avoid tearing the underlying mucosa, particularly the duodenal end. Twin 2 also had a concomitant right inguinal herniotomy.

Postoperatively, both twins had uneventful recoveries. Follow-up reviews at one week (Figure 2), one month and three months were satisfactory. Both twins had been feeding well and had put on weight as expected. Both were later discharged to the outpatient paediatric clinic for further follow up.

DISCUSSION

Pyloric stenosis is generally rare in the East including Brunei Darussalam. Our cases of twin CHIPS are the first two cases encountered in our local population. Of greater interest is the fact that this had occurred in both twins raising the possibility of a genetic aetiology.

The first accurate clinicopathologic description of CHIPS was accredited to Hirschsprung in the late nineteenth century when surgical treatment with gastrojejenostomy was associated with a high mortality of 60%.

The underlying pathogenesis of CHIPS is not clear. Some have attributed it to being congenital in origin while other studies have suggested a possible link to maternal exposure to the erythromycin and azithromycin group of antibiotics during the prenatal period or disordered innervation of the pylorus of unknown aetiology. [2-8] Infective aetiologies have also been postulated. Viruses and bacteria such as mycobacterium have been isolated from biospies taken at the time of Ramstedt's procedure.

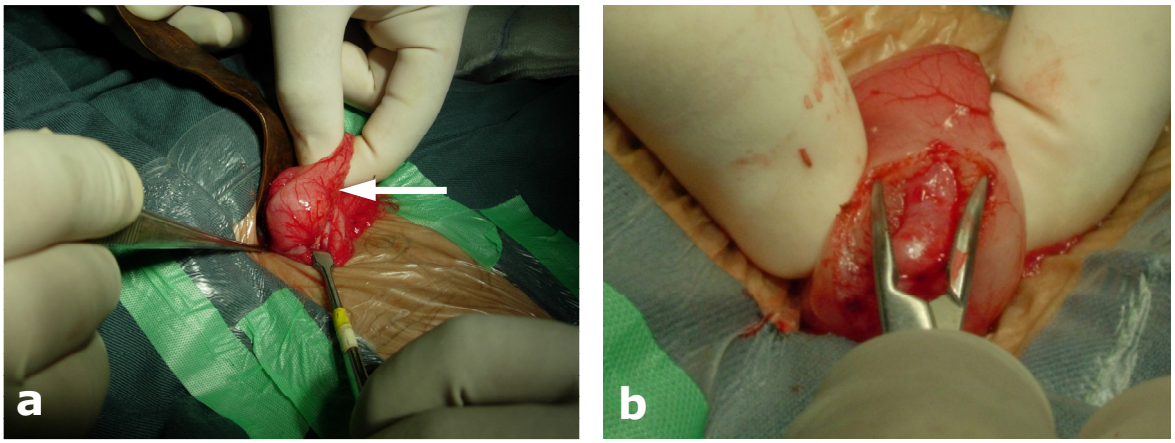

Figure 1: a) Laparotomy showed bulky pyloric tumour of pyloric stenosis (white arrow), b) dissection of Standard Ramstedt's pyloromyotomy.

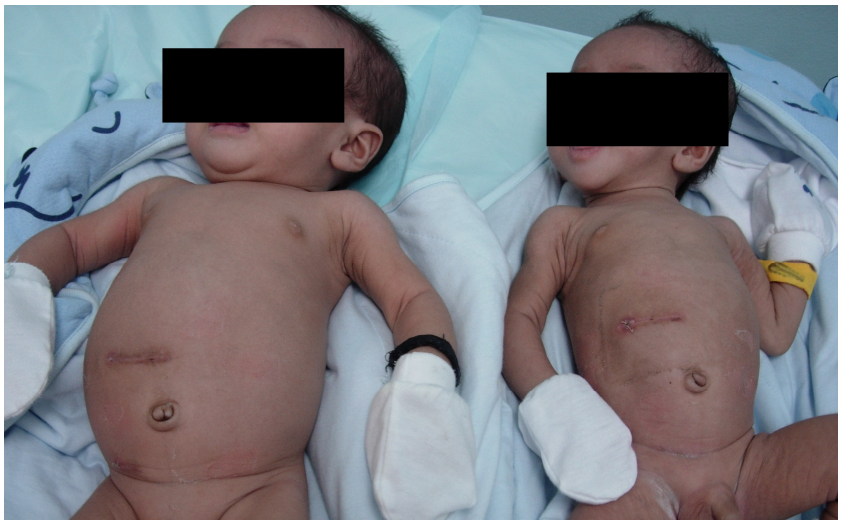

Fig 2: One week post operations with the smaller Twin 1 on the left and the bigger Twin 2 on the right.

The clinical manifestations of CHIPS are fairly typical. The attending paediatrician or surgeon has to raise the suspicion of this condition in an infant who has projectile non-bilious vomiting. The typical clinical presentation is a term male infant between three and six weeks of age who has progressive, non bilious, projectile vomiting resulting in significant dehydration with hypochloremia, hypokalaemic metabolic alkalosis associated with paradoxical aciduria in severe cases. The hallmark of the diagnosis is the finding of a small, mobile, ovoid mass in the epigastrium. A positive examination of this mass is very accurate reaching a sensitivity of 97%. [9] In negative examination, real-time ultrasound measurement of the pylorus channel of 17 mm diameter, 17 mm length and 4 mm wall thickness is suspicious of pyloric stenosis. Barium study of the upper gastrointestinal tract can help further improve the accuracy in diagnosis. [10-12]

Once the diagnosis has been confirmed, surgical correction should be planned immediately. However, it is often advisable to resuscitate adequately (usually for 24 to 48 hours) prior to surgery to correct the metabolic and hydration disturbances. The treatment of choice is pyloromyotomy introduced by Ramstedt in 1911. [13] Nowadays, laparoscopic pyloromyotomy is widely practiced in most paediatric surgical units. [14, 15] Ramstedt's pyloromyotomy is usually straightforward and recovery is uncomplicated. A feeding regimen is usually started four to eight hours after operation with a small volume of sugar water, advancing volume and osmolarity every two to three hours until the infant is taking formula or breast milk *ad libitum*. Persistent vomiting of more than two weeks may occur but re-exploration needs to be considered for the possibility of incomplete pyloromyotomy. A contrast study is usually required in this case to exclude duodenal perforation (three to 30%) and mucosal leak as this can produce fluid collection resulting in gastric outlet obstruction. Wound infection and dehiscence are relatively uncommon nowadays. There is no long term adverse effect of pyloromyotomy in terms of gastric emptying and abdominal symptoms in patient with pyloric stenosis as

compared to the general population. [16]

In conclusion, this report highlighted two rare case of CHIPS occurring in a pair of Malay twin boys in a setting where this condition is exceptionally rare.

REFERENCES

1: Apselund G, Langer JC. Current management of hypertrophic pyloric stenosis. Semin Pediatr Surg. 2007; 6:27-33.

2: Mitchell LE, Risch N. The genetics of infantile hypertrophic pyloric stenosis. A reanalysis. Am J Dis Child 1993; 147:1203-11.

3: Katz S, Basel D, Branski D. Prenatal gastric dilatation and infantile hypertrophic pyloric stenosis. J Paed Surge 1988; 23:1021-2.

4: Mahon BE, Rosenman MB, Kleiman MB. Maternal and infant use of erythromycin and other macrolides antibiotics as risk factors for infantile hypertrophic pyloric stenosis. J Paed 2001; 139:380-4.

5: Sørensen HT, Skriver MV, Pedersen L, Larsen H, Ebbesen F, Schønheyder HC. Risk of infantile hypertrophic pyloric stenosis after maternal use of macrolides. Scand J Infect Dis 2003; 35:104-6.

6: Kobayashi H, O'Brien DS, Puri P. Immunochemical characterization of neural cell adhesion molecule (NCAM), nitric oxide synthase, and neurofilament protein expression in pyloric muscle of patients with pyloric stenosis. J Paed Gastroenterol Nutr 1995; 20:319-25.

7: Langer JC, Berezin I, Daniel EE. Hypertrophic pyloric stenosis:ultrastructural abnormalities of enteric nerves and the interstitial cells of Cajal. J Paed Surg 1995; 30:1535-43.

8: Kobayashi H, MiyaharaK, Yamataka A, et al. pyloric stenosis: new histopathologic perspective using confocal laser scanning. J Paed Surg 2001; 36:1277-9.

9: Godbole P, Sprigg A, Dickson JA, et al. Ultrasound compared with clinical examination I infatile hypertrophic pylori stenosis. Arch Dis Child 1996; 75:335-7.

10: Stunden RJ, LeQuesne GW, Little KE. The improved ultrasound diagnosis of hypertrophic pyloric stenosis. Pediatr Radiol. 1986; 16:200-5.

11: Lamki N, Ahey PA, Round ME, et al. Hypertrophic pyloric stenosis in the neonate: diagnostic criteria revisited. Can Assoc Radiolo J 1993; 44:21-4.

12: Westra SJ, de Groot CJ, Smith NJ, et al. Hypertrophic pyloric stenosis: Use of the pyloric volume measurement in early US diagnosis. Radiology 1989; 172:615-9.

13: Murtagh K, Perry P, Corlett M, Fraser I. Infantile hypertrophic pyloric stenosis. Dig Dis. 1992; 10:190-8.

14: Greason KL, Thompson WR, Downey EC,et al. Laparoscopic pylromyotomy for infantile hypertrophic pyloric stenosis: report of 11 cases. J Paed Surg 1995; 30:1571-4.

15: Fujimoto T, Lane GJ, Segawa O, et al. Laparoscopic extramucosal pyloromyotomy versus open pyloromyotomy for infantile hypertrophic pyloric stenosis: which is better? J Paed surg 1999; 34:370-2.

16: Ludtke FE, Bertus M, Voth E, et al. Gastric emptying 16 to 26 years after treatment of infantile hypertrophic pyloric stenosis. J Paed surg 1994; 29:523-6.