Case Report:

Infantile hypertrophic pyloric stenosis(IHPS) in identical male twins

[1]Rahul Gupta, [2]Tariq Ahmed Mala* , [3] Shahid Amin Malla

[1] Postgraduate , Department of Surgery , GMC Jammu (J&K) India
[2] Postgraduate, Department of surgery, ASCOMS and Hospital Jammu (J&K) India-180017
[3] M.B.B.S Student , S.S. Medical College Rewa (M.P) India
***Corresponding author**: Email: drtariq_6481mala@rediffmail.com

Abstract:

We reported one pair of identical male twins with IHPS who were born with caesarian section. During the third week of life, they suffered from episodic non bilious vomiting. Examination revealed low activity levels, dry lips, sunken eyes, and depressed anterior fontanelle with moderate degree of dehydration. Olive was palpable in both twins and ultrasonography showed a hypoechoic target lesion in the the right upper quadrant of abdomen in both twins. IHPS were confirmed. Ramstedt extramucosal pyloromyotomy was done on same day, hospital stay remained uneventful and both continued to gain weight.

Key words: Infantile hypertrophic pyloric stenosis (IHPS), identical twins, monozygotic twins

Introduction:

Infantile hypertrophic pyloric stenosis (IHPS) is a condition affecting neonates three to eight weeks after birth [1].The incidence of IHPS is approximately 2 to 5 per 1000 live births in most white population [1]. IHPS is less common in India and among black and Asian populations, with a frequency that is one-third to one-fifth that in the white population [2]. IHPS is a functional gastric outlet obstruction characterized by hypertrophy and hyperplasia of the pylorus muscles. The standard procedure to correct IHPS is a Ramstedt extramucosal pyloromyotomy. It's a highly effective simple, elegant, and inexpensive operation described as 'one of the most easy and gratifying procedures performed by pediatric surgeons. Despite knowing how to treat IHPS, the pathogenesis and genetic patterns of IHPS are not fully understood. Herein, we present one pair of male twins with IHPS and review the associated articles about the pathogenesis and genetic patterns of IHPS.

Case report:

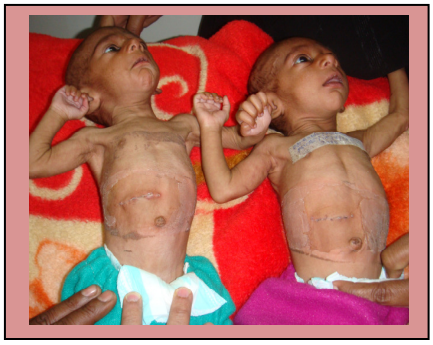
The parents of the identical male twins are of Muslim faith. The mother (G2P2) did not have a medical history, there was no history of intake of erythromycin, no history of consanguinity but there was history of intake of ovulation induction treatment prior to conception. There was no family history of IHPS. Pregnancy was full term and planned caesarian section was performed. The male twins were second in birth order, with an elder female sibling. Both twins passed meconium on the first day of life. During the third week of life, they suffered from episodic postprandial non bilious vomiting with a time lag of 3 to 4 days between them.

On admission physical examination revealed low activity levels, dry lips, sunken eyes, and depressed anterior fontanelle with moderate degree of dehydration. Olive was palpable in both twins. No other congenital abnormality was present. The weight of twin A (Babblu) and twin B (Guddu) were 2kg and 1.8 kg respectively. Ultrasonography showed a hypoechoic target lesion in the the right upper

quadrant of abdomen in both twins. IHPS were confirmed in both twins concurrently. The laboratory values and sonography findings have been summarized in the table1. After intraveneous hydration and correction of electrolyte imbalance, they underwent Ramstedt extramucosal pyloromyotomy on same day. Both were fed with breast milk the day after surgery. Hospital stay was uneventful and both continued to gain weight.

Table 1:- Laboratory values and sonography findings in both twins.

Investigations	Twin A	Twin B
D.O.B	9/12/2011	9/12/2011
Birth Weight (Kg)	2.5	2.4
Weight on admission (Kg)	2	1.8
Age at onset	Late 3rd week	Early 3rd week
Hb(gm%)	12	11.4
TLC(mm^3)	9100	8800
AST(U/L)	72	106
ALT(U/L)	36	62
Na^+(meq)	135	132
K^+(meq)	4.2	3.77
cl^-(meq)	101	101
Urea(mg%)	104	54
Creatinine(mg%)	2	1
Total bilirubin(mg%) Indirect bilirubin	0.8	1.9 1.1
Pyloric canal length(mm)	17.4	16
Pyloric thickness(mm)	4.3	4.5
Target diameter(mm)	13.2	13



Photographs: Preoperative & Postoperative

Discussion:

Idiopathic hypertrophic pyloric stenosis (IHPS) is the second most common cause for abdominal operation during infancy, according to the western literature [3]. Patients with IHPS present with non bilious vomiting and with gradual progression of the disease, vomiting becomes projectile and visible gastric peristalsis and a palpable mass in the epigastric area may be observed[4]. All these findings were consistent in our twins. Laboratory studies reveal metabolic alkalosis, hyponatremia and sometimes hypochloremia [4]. Twin A had deranged renal functions, while twin B was hyponatremic. Hyperbilirubinemia was present in twin A and it has been documented in patients with IHPS, but no confirmed pathogenesis has been proven [5]. In our case, elevated levels of serum aspartate aminotransferase were noted in both twins.

Though true pathogenesis of IHPS is still not clearly known, several features of the genetic transmission of IHPS have been noted, including male predominance [6], high incidence in the offspring of affected parents[6], high incidence in the relatives of affected patients[6] and high incidence of IHPS in twins compared with the general population[2,6]. The frequency of twin births in most white populations is approximately 1% and the expected frequency of twins among all infants is approximately 2% [7]. The frequency of twin infants has been reported in 7 series of consecutive cases of pyloric stenosis studied by Mac Mahon, include 12,149 cases among whom 257 (2.1%) were twins. Thus, it appears that twins are no more susceptible to pyloric stenosis than are single-born infants. The 257 affected twins just referred to came from a total of 228 pairs, of which

both members were affected in 29 (8.3%). The risk to the twin sibling of an affected infant, while almost 30 times higher than that in the general population (3 of 1000) and appears to be not much higher than that to a non twin sibling (5.8%). There is higher concordance rate in monozygotic twins than dizygotic twins [6, 7], probandwise concordance in monozygotic twins is 0.25–0.44, and that in dizygotic twins is 0.05–0.10 [6]. Prevalence in male twins is much higher than in female twins[8]. Within the monozygotic group, approximately 50% of twin pairs are not affected [9].

Thus, environmental effects and other factors like gene expression should be considered for understanding the disease. As for our twins, it was sad that no further study could be performed for our patients.

Conclusion:

Both genetic and environmental factors may play a role, there is high concordance rate in twins as reported in the literature, which justifies to consider examining the asymptomatic co-twin when one of the twins presents with IHPS.

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