Ultrasonography in infantile hypertrophic pyloric stenosis

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Abstract

Infantile hypertrophic pyloric stenosis (IHPS) is usually seen among the first born male infants. In recent time, ultrasonography has become the investigation of choice and has almost completely replaced the radiographic upper gastrointestinal series for the diagnosis of the disease. Here we report a case of 31 days infant with hypertrophic pyloric stenosis. Our purpose of reporting this case is to show the importance of ultrasonography in diagnosing infantile hypertrophic pyloric stenosis.

Key words: Infantile hypertrophic pyloric stenosis, ultrasonography, upper gastrointestinal series.

Introduction

Hypertrophic pyloric stenosis in infants is characterized by progressive muscular hypertrophy of the pylorus, which eventually leads to near occlusion of the gastric outlet. Typically, infants with IHPS are clinically normal at birth. During the first few weeks of postnatal life, they develop nonbillous forceful vomiting described as 'projectile'. Gastric outlet obstruction leads to emaciation and if left untreated, may result in death. Surgical treatment is curative. The clinical diagnosis is based on palpation of thickened pylorus or 'olive'. In last few decades, imaging modalities, especially ultrasound, has been playing a great role in diagnosing this disease.

Case report

A 31 days old male infant was sent for USG from emergency department. The infant presented with non-bilious projectile vomiting for last 4-5 days immediately after feeding. Initially the vomiting was infrequent but was progressively worsening. The patient's mother reported that the infant had been feeding well until last week. History was negative for diarrhea or bloody stool. The infant was born full term via normal spontaneous vaginal delivery. The review of patient's past medical and family history was noncontributory. On physical examination the patient was ill-looking and irritable. He was afebrile. Chest was clear on auscultation. On palpation, abdomen was mildly distended with normal bowel sound. Anterior fontanel was depressed. Lab tests showed Hb 12.7 mg/dl, TC 8.9, DC $N_{69}L_{30}E_{1}M_{0}B_{0}$, ESR 08, CRP negative.

In ultrasound, in right hypochondriac region, longitudinal scan (figure:1) showed markedly thickened, hypoechoic elongated canal shaped structure with narrowed lumen measuring approx. 5.6 mm in width and 1.7 cm in length. Transverse scan (figure: 2) showed typical hypoechoic doughnut.

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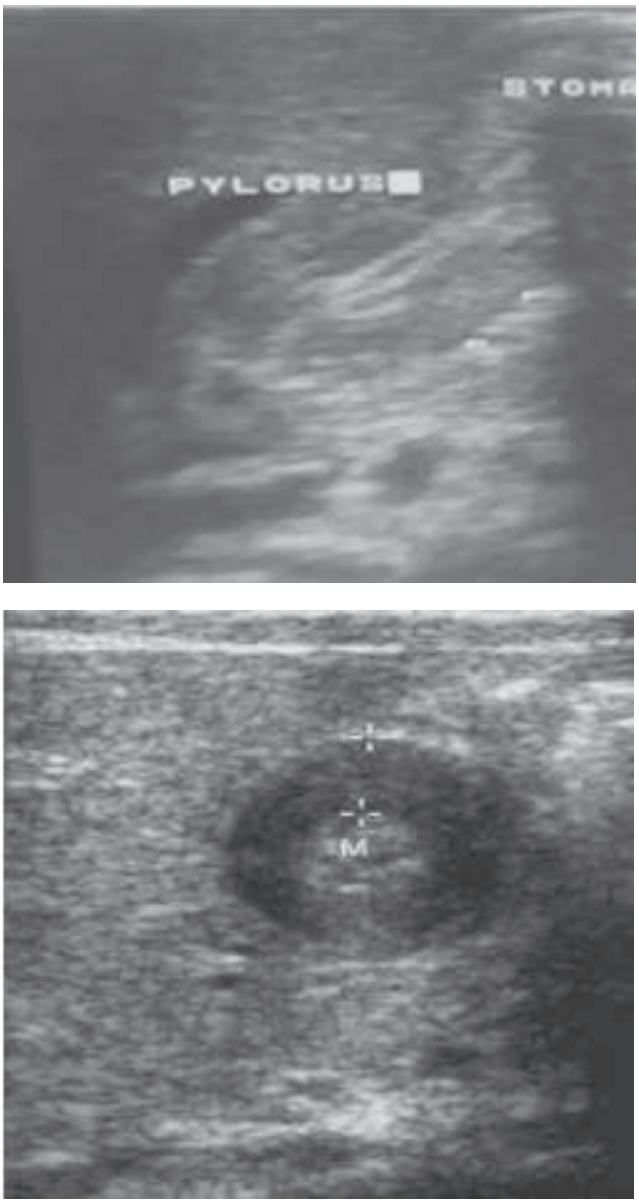

Figures:1 Longitudinal scan shows markedly thickened hypoechoic gastric pyloric muscle. (2). Transverse scan shows typical hypoechoic doughnut.

Discussion

Infantile hypertrophic pyloric stenosis was virtually unknown prior to 1627, when a clinical description with survival was reported by Fabricious Hildanus.[1] At the German Pediatric Congress in Wiesbaden in 1887, Harald Hirschsprung described two infant girls with pathologically proved IHPS, and his seminal article, published in 1888, triggered a scientific interest in this condition.[2]

The incidence of hypertrophic pyloric stenosis is approximately 1 case per 250 live births in white population.[3] 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.[4] Peak incidence is between 2 and 8 weeks of age, although cases in infant as young as 7 days are encountered.[5] There is greater incidence in males (4:1 ratio), in Caucasians, and in those with a family history.[6] The etiology remains largely unknown although abnormal innervations to the circular muscles have been implicated. Upper gastrointestinal series and ultrasonography are the imaging modalities used to diagnose hypertrophic pyloric stenosis.

Upper GI series is performed with the infant in the right anterior oblique position, to facilitate gastric emptying. The examination can be successfully accomplished with the child drinking from a bottle; these infants are usually very hungry and will drink with little effort. Insertion of a nasogastric tube is not necessary; however emptying of an overdistended stomach may help to prevent vomiting if needed. Fluoroscopic observations include vigorous active peristalsis resembling a caterpillar and coming to an abrupt stop at the pyloric antrum, outlining the external thickened muscle as an extrinsic impression, termed the **shoulder sign**. Luminal barium may be transiently trapped between the peristaltic wave and the muscle, and this is termed the **tit sign**. Eventual success of gastric peristaltic activity will propel contrast material through the pyloric mucosal interstices, with the appearance as either the **string sign** or the **double-track sign**. This sign demonstrates the intervening

redundant mucosa outlined as a filling defect by the contrast material; it was reported as specific for IHPS by Haran et al in 1966.[7]

During the past decade, ultrasonography has almost completely replaced the radiographic upper gastrointestinal series for the diagnosis of infantile HPS[5]. Unlike the upper GI series that demonstrates only the indirect effects of pyloric muscle hypertrophy on the gastric lumen, ultrasound allows direct visualization of the gastric muscle thickening that is the hallmark of the disease. Although a few pitfalls in the sonographic diagnosis of HPS exist, the technique is relatively easily mastered and results in greatly improved accuracy of diagnosis and patient outcome. Indeed, the accuracy approaches 100%, and ultrasound is now the procedure of choice for the detection of pyloric stenosis.[8,9]

After the initial documentation of the sonographic detection of the hypertrophied pyloric muscle in pyloric stenosis by Teele and Smith, a plethora of articles ensued describing the characteristic findings of this condition.[10-14] Increased pyloric muscle thickness and canal length, increased transverse diameter of the pylorus, estimation of degree of gastric outlet obstruction, and calculation of pyloric muscle volume have all been used to diagnose pyloric stenosis, but of these criteria, thickening of the pyloric muscle and elongation of the pyloric canal have emerged as the most useful. The thickness at which muscle is considered hypertrophied is 3 mm or greater[8]. Pyloric canal length of 1.5 cm is considered diagnostic of pyloric stenosis when seen in conjunction with thickened pyloric muscle. In practice, however, normal canal length is much shorter than this and is often impossible to measure. Measurement of canal length is more difficult than is measurement of muscle thickness and, therefore, is a less reliable criterion. In borderline cases in which muscle thickness and canal length are less than classic, calculation of the pyloric volume may be helpful.[5]

In the classic case of IHPS, the thickened muscle mass is seen as a hypoechoic layer just superficial to the more echogenic layer of pyloric canal. In cross section, it resembles a sonolucent 'doughnut' medial to the gallbladder and anterior to the right kidney. In longitudinal section, ultrasonography also permits evaluation of functional alternations at the pylorus.[5]

De Backer et al proposed endoscopy as a successful tool in the diagnosis of IHPS.[15] Demonstration of the cauliflower or nipple like projection of the mucosa is characteristic in patients with IHPS. However, the invasiveness and expense of this procedure do not seem to justify its use when other diagnostic methods are available.

In conclusion, ultrasound has emerged as the modality of choice for diagnosing IHPS as it is easily available, inexpensive, and noninvasive and has better sensitivity. Hence, ultrasound must be done in all infants with IHPS.

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