

Case Report

Adult idiopathic hypertrophic pyloric stenosis: an infrequent cause of gastric outlet obstruction

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ABSTRACT

Hypertrophic pyloric stenosis is a congenital disease, presenting within two weeks of birth. However adult idiopathic hypertrophic pyloric stenosis (AIHPS) presents in middle age, predominantly in males and usually without any antecedent cause. Secondary variant may be due to intra gastric causes or extra gastric post-operative adhesions. Patient presents with symptoms of gastric outlet obstruction. Diagnosis depends on clinical, radiological and endoscopic findings. Treatment is subtotal gastrectomy. Pyloroplasty and endoscopic dilatation may be tried in debilitated patients. We present a case of AIHPS presenting as gastric outlet obstruction in a 16 year old female, that was surgically managed with an antrectomy.

Keywords: AIHPS, Antrectomy, Cervix sign, Endoscopic balloon dilatation, Pyloromyotomy, Pyloroplasty

INTRODUCTION

Adult hypertrophic pyloric stenosis is a rare entity, with less than 400 cases reported to date.¹ It may present insidiously with vomiting, abdominal distension with no childhood history of the same. It seldom presents as an abdominal mass. It may be idiopathic or secondary to diseases of upper gastrointestinal tract. It is largely benign and is characterised by hypertrophy of circular muscles of pyloric canal, seen as narrowing on radiology.² Diagnosis is made using clinical presentation, examination findings, radiological images and endoscopic images showing 'cervix sign'.³ Treatment includes endoscopic dilatation, pyloromyotomy, pyloroplasty, antrectomy, partial or total gastrectomy.⁴

CASE REPORT

A 16 year old female, presented with complaints of vomiting since 3 months. Vomitus was insidious in onset. Gradually, she vomited after every meal, vomitus contained food particles and was neither blood nor bile

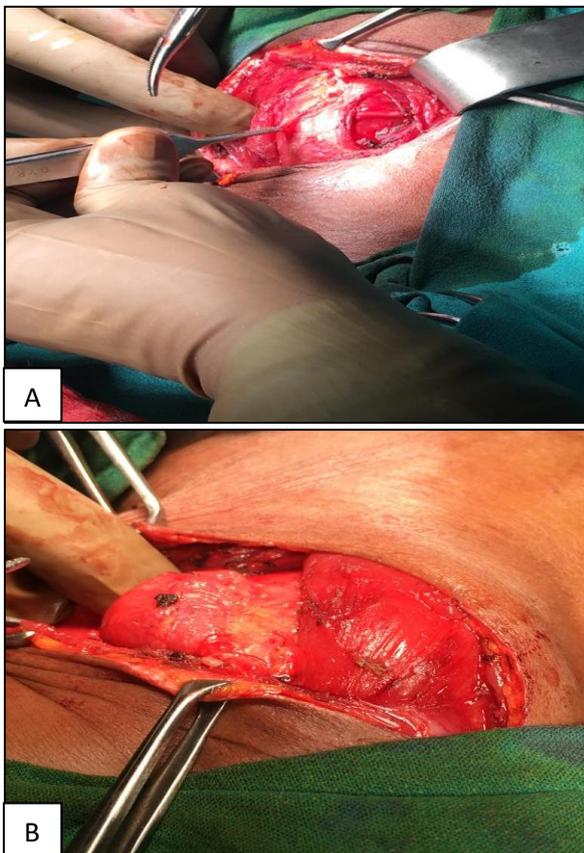
stained. Patient could only tolerate liquid diet and had lost 5 kgs, equivalent to 13% of her body weight in 3 months. She had no history of similar complaints in her childhood. There was no contributory medical or surgical history. On Clinical examination, patient was thin built and poorly nourished. On examining her abdomen, a palpable lump was found in her epigastrium, firm in consistency and with a smooth surface.

Patient underwent a gastroduodenoscopy. Stomach contained solid and liquid food residues, with narrowing of pyloric opening. The endoscope could not be negotiated through the duodenum due to pyloric stenosis.

CECT abdomen was performed. It was suggestive of an over-distended stomach with fluid and hyperdense food contents within with circumferential wall thickening of pylorus causing luminal compromise. Few subcentimeter pre/para aortic and mesenteric lymph nodes noted. Liver was normal.

Patient was hydrated, Ryle's tube was inserted and stomach wash was given. Routine blood investigations and ABG were within normal limits. Patient was posted for exploratory laparotomy. Intraoperatively pylorus and distal antrum were thickened. Few enlarged mesenteric lymph nodes were seen. Liver surface appeared normal. An antrectomy with Roux en Y anterior isoperistaltic Gastrojejunostomy was performed and mesenteric lymph node was sent for biopsy. Patient was started on oral feeds on post-operative day 3, and had an uneventful recovery, getting discharged on day 7 of surgery. Patient was on full diet by day 10.

Histopathology report was suggestive of marked hypertrophy of muscularis propria with smooth muscle cells arranged in fascicles along with inflammatory cells in between fibres. Transition between thickened and normal area was gradual. No evidence of atypia or malignancy seen. Mesenteric lymph node was suggestive of reactive lymphadenitis.



Figures 1 (A and B): Intraoperative hypertrophied antrum and pylorus.

DISCUSSION

Adult idiopathic hypertrophic pyloric stenosis was first described by Jean Curveilhier in 1835.⁵ Adult hypertrophic pyloric stenosis is classified into three variants. Infantile type is present since infancy and gradually presents with symptoms. Second type is the one

that occurs secondary to any gastric pathology like malignancy, hiatus hernia, duodenal/ gastric ulcer, inflammatory disease, bezoars and even extrinsic post-operative adhesions. The third or idiopathic variant occurs in the absence of any pre-existing pathology.⁶ Documented cases were predominantly in middle aged men, due to no apparent cause.⁷

The etiology of adult idiopathic hypertrophic pyloric stenosis is unknown. But many theories have been postulated that suggest an interplay between genetic and environmental factors. It has been hypothesised that a milder form of the infantile variant persists and presents itself in adulthood. This is due to marked histological and anatomical overlap between the two variants.⁸ Other possible etiologies include protracted pylorospasm, vagal hyperactivity, and neuromuscular incoordination due to changes in Auerbach's plexus.⁹ Some authors have suggested familial tendency to develop IHPS, although most cases occur de novo.¹⁰

Clinical presentation is similar to gastric outlet obstruction. Patient presents with vomiting, post prandial abdominal distension, pain in abdomen, loss of weight, rarely a palpable mass in abdomen. Later patient may develop electrolyte disturbances, namely hypochloremic hypokalemic metabolic alkalosis. Differential diagnosis include malignancy and diabetic gastropathy.¹¹

Radiology examination may be normal.¹² Certain signs are seen on radiology. Normal pyloric canal is less than 1cm, it increases to upto 4cms in AIHPS. 'Kirklin Sign' is convex indentation at the base of duodenal bulb seen in AIHPS. 'String sign' is concentric narrowing of pylorus and 'Twining's sign' is a barium filling defect seen on either or both sides of pylorus.¹³ Endoscopy demonstrates narrowing of pylorus, also known as 'cervix sign'. Endoscopy helps differentiate AIHPS from other causes and also helps rule out malignancy by taking a biopsy.

Treatment is surgical. Subtotal gastric resection has been widely done in the past. Treatment options include gastrojejunostomy, gastrectomy, pyloroplasty, pyloromyotomy and endoscopic balloon dilatation. Pyloroplasty is undesirable as there is possibility of mucosal laceration and secondary diverticula. Pyloroplasty is difficult in grossly thickened pylorus and is reserved for debilitated patients, and can be done laproscopically.¹⁴ Endoscopic balloon dilatation has a very high recurrence rate and can be done only if the patient is debilitated.¹⁵

CONCLUSION

Adult idiopathic hypertrophic pyloric stenosis is a rare cause of gastric outlet obstruction. Diagnosis depends on history, clinical, radiological and endoscopic findings. Treatment is surgical, with a distal, subtotal gastrectomy giving a favourable outcome.

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