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Short Communication: Gastric Outlet Problems in a 51-Year Old Man with Down Syndrome

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Abstract

A 51-year-old man with Down syndrome was admitted with long-term gastric outlet problems. Extensive repeated gastroduodenoscopy showed stasis of debris, a large distention of the stomach and proximal duodenum suggestive for obstruction, which was confirmed by addition CT-scanning. Upper gastrointestinal series showed a double-bubble-sign, highly suggestive for a congenital duodenal obstruction. The patient was diagnosed with a duodenal web, a condition caused by failure of epithelial obliteration during the early embryological period. Although mostly diagnosed in the neonatal period, imperforated duodenal webs could be identified in adult patients, mostly associated with various congenital syndromes including down-syndrome. Until recently surgery was the only treatment option. However, with endoscopic experience and options expanding, endoscopic dilatation is a suitable alternative.

Keywords: Gastric Outlet; Duodenum Artresia; Down Syndrome

Introduction

A 51-year-old man with Down syndrome was submitted for evaluation of a symptomatic anemia and melena. His past medical history included long-term gastric outlet problems and complaints of reflux in which a proton pump inhibitor was prescribed. During the incomplete gastroduodenoscopy a ulcer of the bulbus was diagnosed, most likely causing upper gastric bleeding.

Although proper preparation gastroduodenoscopy failed multiple times due to ominous stasis of debris (Image A) along with large distention of stomach and proximal duodenum suggesting distal duodenal obstruction. In the work-up an abdominal CT-scan was obtained showing no signs of extrinsic compression causing the obstruction (Image B). An upper gastrointestinal series showed slow transit of the Barium meal with a stenosis at the transition of the vertical to the horizontal part of the duodenum. In addition, a possible "double bubble-sign" could be identified (Image C). As final treatment, the patient underwent surgery (duodenoduodenostomy). Pathological examination showed a mid-duodenal obstruction caused by fenestrated epithelial tissue.

Image A: gastroduodenoscopy with stasis of debris.

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Image B: CT Scan showing distention of the stomach and proximal duodenum.



Image C: Gastrointestinal series showing a "double-bouble sign".

Question: what is the diagnosis?

Answer: The patient was diagnosed with a duodenal web, a congenital duodenal obstruction.

A stenosis of the distal duodenum along with a typical air-configuration also-called a "double bubble sign" was shown at the upper gastrointestinal series, both highly suggestive for congenital intrinsic obstruction. Although most congenital intrinsic duodenal obstructions such as duodenal atresia are diagnosed in the early neonatal period a duodenal web can be discovered in (late) adulthood [1,2]. Caused by a failure of epithelial obliteration in the early embryological duodenum an epithelial web is formed, varying from continues epithelial obstruction to fenestrated webs in which intestinal transit is possible [1,3]. Multiple congenital disorders, including Down syndrome, renal and cardiac anomalies and vertebral defects are associated with this condition. Since intestinal transit is permitted by a (partial) imperforated epithelial web most patients suffering various obstructive symptoms, including biliary vomiting, upper abdominal discomfort and failure to thrive [4]. Although surgical treatment, including duodeno-duodenostomy or a bypass surgery, was until recently the preferred treatment option, endoscopic experience and options are expanding. Recent studies have shown successful endoscopic dilatation of 3 patients with a duodenal web [1].

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