Case Report

Annular pancreas mimicking hypertrophic pyloric stenosis in a female infant✩

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ABSTRACT

Annular pancreas is a rare congenital intestinal obstruction to be found in infants. It is characterized by nonbilious vomiting, abdominal distention, and feeding intolerance that share similar symptoms with other intestinal obstructions. We reported a case of 11-month-old female infant with a history of nonbilious vomiting after coughing as much as 6 times per day 1 day before admission. The patient had an incorrect provisional diagnosis of hypertrophic pyloric stenosis based on ultrasound. Later on, an upper gastrointestinal series showed dilatation of the second part of the duodenum which was successfully treated by a duodenostomy during laparotomy procedure. It is suggested that treating an intestinal obstruction needs a further investigation when the symptoms were continuously repeated.

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Introduction

Annular pancreas is a rare congenital anomaly that occurs when the ventral pancreatic bud fails to rotate in the normal fashion, leaving a segment of the pancreas that encircles the second portion of the duodenum, caused by failed migration of the Anlagen, and causes varying degrees of duodenal obstruction [1]. Annular pancreas was first recognized by Tiedemann in 1818 and from the early autopsy and surgical series the estimated incidence to be approximately 3 cases in 20,000 births and only 737 cases have been reported in the English literature [2]. Patients with an annular pancreas may also have other congenital anomalies, trisomy 21 (Down syndrome) was the most commonly associated followed with cardiac anomalies; such as atrioventricular canal, tetralogy of Fallot, ventricular septal defect, atrial septal defect, and cleft mitral valve; intestinal malrotation, tracheoesophageal fistula, gastrointestinal anomalies, genitourinary anomalies, cerebral palsy, spinal cord defect and up duodenal atresia and stenosis [2],[3].

The clinical manifestations of the annular pancreas depend on the degree of obstruction or the duodenal stenosis but most patients are asymptomatic. Li et al. [4] reported the demographic of clinical presentation from 11 children with the annular pancreas that all of them are present with bilious

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vomiting, with 1 patient followed with anal atresia [4]. However, clinical severities can vary over a wide series. The annular pancreas has similar symptoms with another “high” intestinal obstructions, neonates, and infants typically present with usually a nonbilious vomiting, abdominal distention, and feeding intolerance [5],[6].

Radiological findings using ultrasonography or CT/MRI could be the first choice for imaging evaluation in pediatric patients with “high” intestinal obstruction. These findings can all demonstrate similar findings with varying degrees of duodenal obstruction and enlargement of the pancreatic head with soft tissue that is identical in appearance to the pancreas surrounding the second portion of the duodenum [2],[5],[7].

In this report, we present an 11-month-old infant with an annular pancreas who presented with nonbilious vomiting after coughing who was diagnosed with upper gastrointestinal series after a misleading provisional diagnosis of hypertrophic pyloric stenosis by the abdominal ultrasound. The annular pancreas performed the surgical procedures later.

Case report

An 11-month-old female infant was referred to our university hospital with a history of nonbilious vomiting after coughing as much 6 times per day 1 day before admission. Symptoms followed by coughing and fever. History of choke, breathless, and diarrhea was denied. The patient has a history of inpatient last December and underwent an abdominal ultrasound that recognized hypertrophic pyloric stenosis during the examination. The patient also has a history of chronic suppurative otitis media benign type on the right ear, type I and III laryngomalacia, and disturbance of airway clearance. The patient’s birth was via spontaneous vaginal delivery with normal weight at normal weeks of delivery. Family history is unknown. The patient was born with Down syndrome.

Based on the physical examination, the patient was 58 cm in length and 3700 g in weight, with a heart rate 138 bpm, respiratory rate 34 × /min, 38.9°C in temperature, and 94% of oxygen saturation (patient were using a nasal cannula with oxygen 2 L/min). The patient has pale conjunctiva, no icteric sclera were found. The patient’s abdomen presented a bowel contour and a distended stomach with positive bowel sounds. Due to the symptoms, an upper gastrointestinal series was ordered to complete the investigations of the previous abdominal ultrasonography.

The laboratory findings from blood routines before surgery show that patient has anemia (Hb 7.0 g/dL and Hct 21.9%) and electrolyte imbalance (Na⁺ 130 mEq/L, K⁺ 4.2 mEq/L, and Cl⁻ 106.0 mEq/L). The previous abdominal ultrasonography revealed a temporary thickening of the pyloric at the relaxation stage and was wrongly diagnosed as hypertrophic pyloric stenosis (Fig. 1). The upper gastrointestinal series was described as a dilatation of the second part of the duodenum (Fig. 2). This finding raised our suspicion of intestinal obstruction around the duodenum. A CT or MRI was not performed on the patient.

After a wrong diagnosis made by abdominal ultrasonography, the following upper gastrointestinal series result in the suggestive annular pancreas. The laparotomy procedure showed the pancreas encircles the second part of the duodenum that confirms the annular pancreas diagnosis (Fig. 3). A duodenostomy was done to release the abdominal compression. One day after the operation, no vomiting was reported. The postoperative course in this patient was sufficient; there was an increase in the patient’s nutritional status. The patient was discharged after 10 days of hospitalization. Any data that have been released and all procedures were performed under the patient’s permission.

Fig. 1 – An abdominal ultrasonography showed a thickening part of the pyloric spasm which found out in the relaxation stage; misled the diagnosis as hypertrophic pyloric stenosis.

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Fig. 2 – Upper gastrointestinal series define a dilatation of second pars duodenum which mimicking hypertrophic pyloric stenosis.

**Discussion**

The human pancreas is divided into the head, neck, and tail of the pancreas, which is normally formed with a combination of anterior and posterior buds in the fourth-eighth weeks of embryonic life [8]. In the annular pancreas, the anterior pancreas buds adhere abnormally to the duodenum and leads to an unusual rotation of this part, leaving a segment of the pancreas enircles the second portion of the duodenum by the pancreatic tissue in the form of a ring including the main pancreatic duct [7]. The prevalence of annular pancreas is difficult to determine specifically. In adults, 5-15 cases per 100,000 in autopsy series to 1 in 250 cases on an endoscopic retrograde cholangiopancreatography study were reported [8],[9]. Meanwhile, this congenital anomaly has a prevalence of 1 case per 12,000-15,000 births or to be approximately 3 cases in 20,000 births and only 737 cases have been reported in the English literature [2],[6]. As we know from the case that the patient was born with a Down syndrome which associates congenital abnormality, annular pancreas in this case [2],[10].

The investigation started from the patients’ chief complaints to the radiological finding. As we reported from our patient that 1 day before admission patient presented with non-bilious vomiting after coughing as much as 6 times per day. The patient's symptoms can vary widely including vomiting or history of vomiting with or without bilious, bloating, and feeding intolerance. All these symptoms are related to duodenal obstruction in children. Vice versa in adults, only 24% presented with gastrointestinal symptoms, including nausea, vomiting, and abdominal bloating. While abdominal pain was the major symptom to be found following by pancreatitis and jaundice [2],[11].

Based on the nutritional status of the patient, both graphics of weight-for-age and length-for-age are below the fifth percentile. We presumed that the patient was vomiting repetitively and combined with poor feeding. The annular pancreas is known to cause complete or incomplete duodenal ob-
struction where this may lead to both previous symptoms. And so that this will manifest as malnutrition and failure to thrive if there is a delay in hospital admission [10]. During the physical examination the patient reported a fever that represents the activation of the compensatory mechanism for dehydration. The patient was not either irritable to eliminate the probability of pancreatitis. The patient’s pale conjunctiva and hemoglobin were described as anemia that represents malnutrition.

The patient underwent abdominal ultrasonography followed with an upper gastrointestinal series. A temporary thickening of the pyloric was found at the gastrointestinal relaxation stage from the abdominal ultrasonography examination which later known become a wrong diagnosis hypertrophic pyloric stenosis; at this stage, the pyloric spasm should have not noticing as an abnormality. From the last admission to our hospital, upper gastrointestinal series was performed and described a dilatation of the second part of the duodenum. From these discoveries, we were suggestive of the diagnosis of the annular pancreas that it is connected to the patient’s previous history of vomiting and another congenital anomaly. Moreover, the surgery was done after both states of dehydration and malnutrition had been resolved and the annular pancreas diagnosis was made during laparotomy. Supporting radiographic examinations, for example, abdominal radiograph and contrast enema may affirm the nearness of hindrance yet not the genuine reason as 40% of the annular pancreas is quite affirmed during laparotomy [12]. A duodenostomy was acted in the patient and a visual discovery shows the pancreas encircles the duodenum which made a dilatation of the second part of the duodenum. Fu et al. stated that every system is legitimate for a specific sort of patient [13]. In more youthful patients, duodenostomy or duodenojejunostomy is favored because the need for vagotomy might be limited [14]. Postoperative course in this patient was sufficient, no vomiting was reported after 1 day of operation, and the patient was discharged following 10 days of hospitalization.

In summary, the annular pancreas is a rare congenital abnormality found either in children or adults. A variety of intestinal obstruction symptoms should be monitored closely as it can lead to an emergency stage if there is a delay in medical treatment. An approach of imaging modality is important to diagnose the annular pancreas. Therefore, prompt diagnosis and early surgical management could improve patient outcomes.

Conclusion

Annular pancreas is a rare congenital anomaly to be found both in children and adults, which needs a close monitor for the wide range of presenting symptoms. Careful interpretation of imaging is fundamental. A high file of suspicion by radiologists is required. Always considering multimodality imaging if there is still a discrepancy between symptoms and one modality imaging result, especially ultrasound examination in pediatric bowel obstruction.

Patient consent statement

Dear Editorial Board of Radiology Case Reports,

Along with this letter, we would like to confirm that our patient has agreed that her daughter’s medical history can be published as a case report paper. In order to protect the patient’s privacy, we did not include the physical appearance of the patient. Furthermore, we focused on imaging and proof of surgical outcomes of the patient, so we did not violate any patient’s privacy.

We hope that this letter is enough to comply the regulation of patient’s privacy in this journal.

Thank you very much for your time and consideration.

Yours Sincerely,

Damayanti Sekarsari, MD

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