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Case Report

VACTERL syndrome with late presentation of annular pancreas with duodenal web: Case report ☆

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ARTICLE INFO

Article history:
Received 15 February 2022
Accepted 22 February 2022

Keywords:
Annular pancreas
Duodenal web
VACTERL Syndrome
Foreign body

ABSTRACT

VACTERL Syndrome affects multiple body systems and can include various anomalies of the Vertebral column, Anus and/or rectum, heart (Cardiac), Tracheo-Esophagus, kidneys (Renal), and Limbs. Patients with VACTERL syndrome are at increased risk of having a congenital duodenal obstruction that may be extrinsic in the form of an annular pancreas or intrinsic in the form of duodenal atresia, stenosis or web. Simultaneous presentation of both the annular pancreas and duodenal web is a rare clinical entity and typically presents in neonates. However, late presentation of annular pancreas combined with a duodenal web is exceedingly uncommon. We present a case of late diagnosis of annular pancreas with duodenal web resulting in an entrapped ingested foreign body.

Introduction

Congenital duodenal obstruction has been reported with an incidence of 13.5 per 100,000 and is most often caused by duodenal atresia, stenosis, or a duodenal web [1]. Although rare, congenital obstruction is the most common cause of proximal bowel obstruction to present in the neonatal period [2]. Delayed presentation of congenital duodenal abnormalities is more likely to occur when the narrowing of the duodenum is incomplete and less severe. Annular pancreas is an extrinsic cause of duodenal narrowing, resulting from a ring of pancreatic tissue encircling the second portion of the duodenum, and can be associated with additional congenital abnormalities of the duodenum [3–4]. The simultaneous presence of both annular pancreas and duodenal web is rare, especially to present with obstruction outside the neonatal period. We present a case of delayed diagnosis of an annular pancreas with a duodenal web in a 4-year-old female with VACTERL syndrome presenting with proximal bowel obstruction with an ingested foreign body; with endoscopic, fluoroscopic, and surgical correlation.

Case

A 4-year-old female with a history of VACTERL syndrome initially presented to an outside hospital with vomiting. An abdominal radiograph demonstrated a coin-shaped foreign body

☆ Competing Interests: The authors have no disclosures and declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2022.02.062

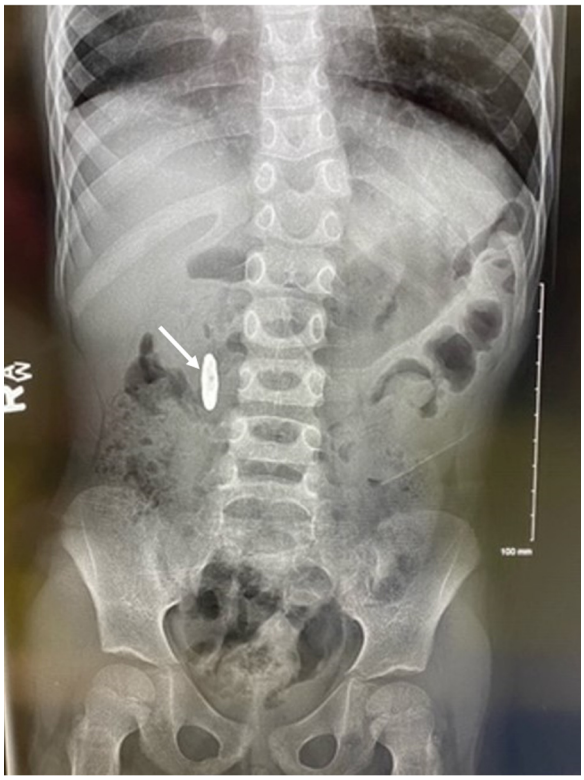

Fig. 1 – AP abdominal radiograph demonstrates a coin-shaped foreign body (arrow) projecting over the mid-abdomen at the expected location of the second portion of the duodenum.

projecting over the mid-abdomen at the expected location of the second portion of the duodenum (Fig. 1). The repeat radiographs the day after demonstrated no significant progression of the foreign body. She subsequently underwent upper endoscopy which retrieved a coin (penny) with some erosive changes obstructing a narrowed opening of a duodenal web (Fig. 2) at the distal, second portion of the duodenum. Furthermore, there was an evidence of significant narrowing at the proximal, second portion of the duodenum as well as difficulty passing the scope past the web like obstruction. The patient recovered and was later referred to our radiology department for an upper gastrointestinal (GI) examination.

Multiple fluoroscopic images from the upper GI demonstrated 2 major findings. There was persistent narrowing at the second portion of the duodenum (Fig. 3, arrow); and correlation with the prior contrast-enhanced CT confirmed that the narrowing was secondary to a rim of pancreatic tissue or annular pancreas extrinsically compressing the duodenum (Fig. 4). Our patient also showed the pathognomonic radiographic appearance of the duodenal web, extending toward the third part of the duodenum, known as the windsock sign with a sac-like dilation of bowel, resembling the appearance of a windsock (Fig. 3, dash arrow). A small amount of the barium contrast is seen past the obstruction which supports the diagnosis of a partial obstruction (Fig. 3, arrowhead).

Intraoperatively, both the annular pancreas and duodenal web were confirmed (Fig. 5) and were contributing to the duodenal stenosis. The duodenal web was excised, duodenoduodenostomy was performed, and a nasogastric tube was placed in the stomach for proximal decompression. Post-operatively this tube was removed once the output quantity decreased

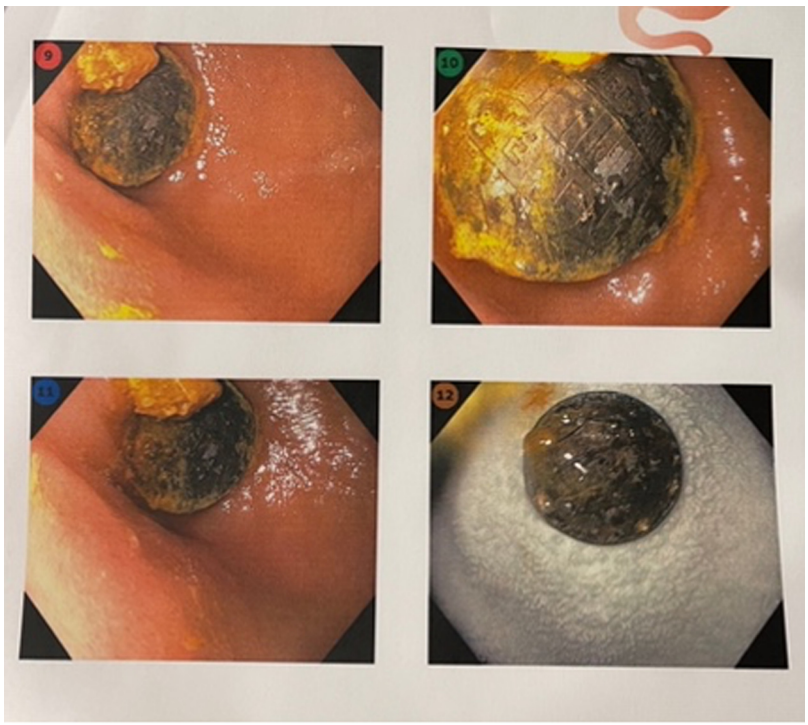

Fig. 2 – Endoscopic images a coin (penny) entrapped within the second portion of the duodenum.

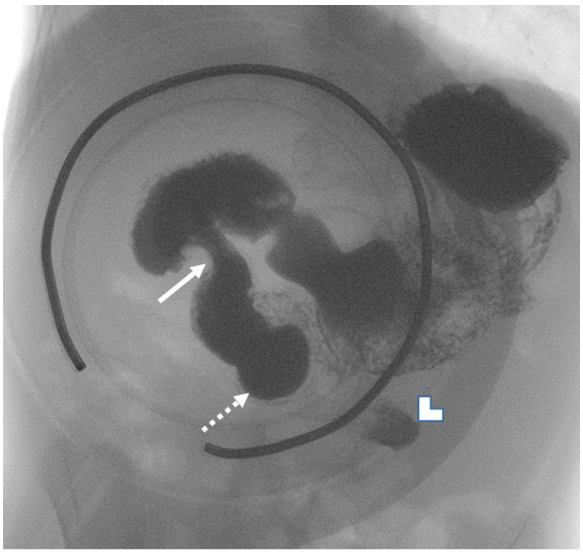

Fig. 3 – AP fluoroscopic image from the upper GI demonstrates a persistent narrowing at the second portion of the duodenum (arrow) representing the extrinsic stenosis from an annular pancreas. In addition, there is sac-like dilation of the bowel resembling a "windsock" (dash arrow) representing the intrinsic narrowing from a duodenal web. A small amount of the barium contrast is seen past the duodenal web without complete obstruction (Fig. 3, arrowhead).

and the output no longer was bilious. Her diet was then slowly advanced. She was tolerating a regular diet at the time of discharge. Informed consent to publish this case was provided by the patient's parents.

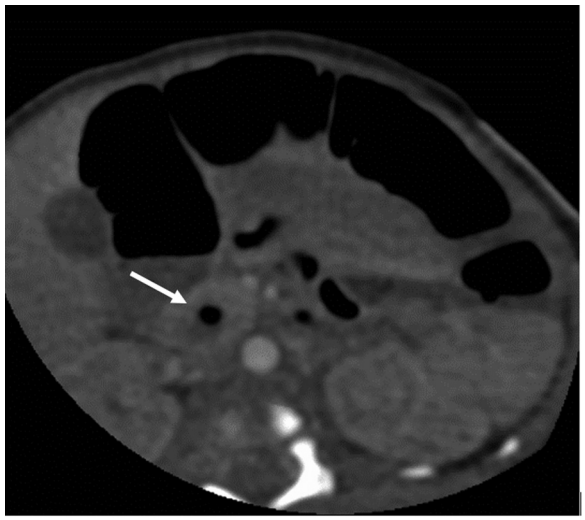

Fig. 4 – Axial CT image of the upper abdomen demonstrates a rim of soft tissue (arrow) surrounding and compressing the second portion of the duodenum, consistent with annular pancreas.

Discussion

Patients with a combination of multi-system congenital anomalies affecting the vertebral column (V), anus and/or rectum (A), heart (C), trachea and/or esophagus (TE), kidneys (R), and limbs (L) may be diagnosed as having a VACTERL syndrome; 3 of which are required for the diagnosis. With a reported incidence of 1 in 10-40,000 live births, VACTERL association most commonly includes a variety of vertebral and/or rib defects; anorectal malformations; renal anomalies, and

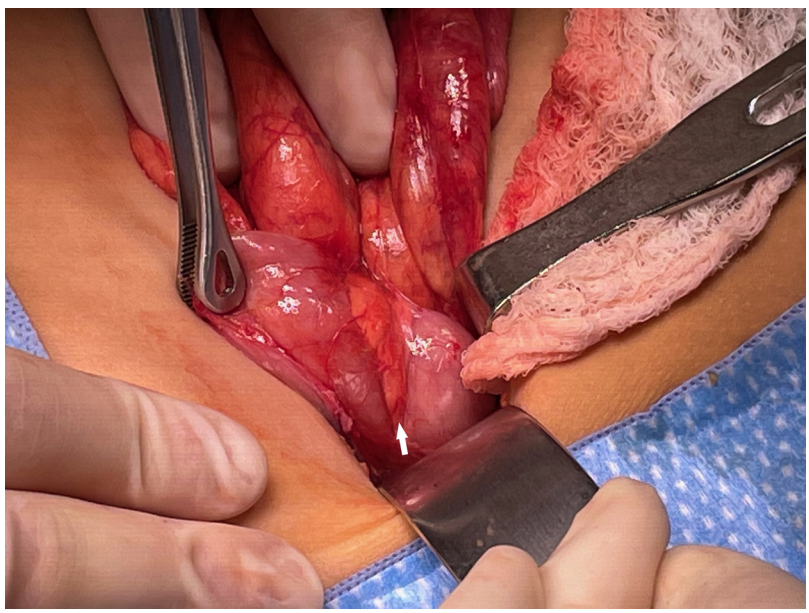

Fig. 5 – Intraoperative photo illustrating dilation of the proximal duodenum to the level of the annular pancreas (arrow).

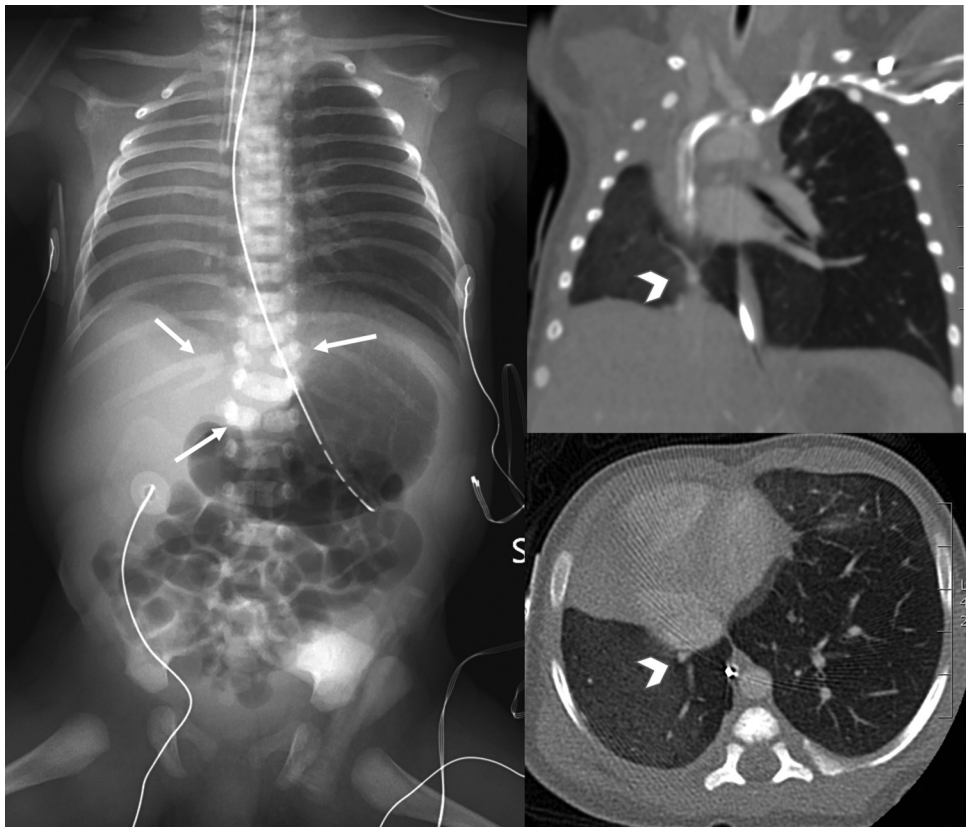

Fig. 6 – Hypogenetic lung, partial anomalous pulmonary venous return (arrowhead), thoracic and lumbar butterfly vertebrae, fused right T11-12 ribs (arrows), and an anorectal malformation (not shown) consistent with VACTERL syndrome.

esophageal atresia, with or without tracheo-esophageal fistula [5–6]. While less common, duodenal stenosis and/or atresia can also be associated with a VACTERL syndrome. In one series, approximately 5% of neonates presenting with duodenal atresia had a VACTERL association [7].

The Hedgehog gene (Hh) has been identified as a crucial ligand involved in the signaling pathways of organogenesis, including the development of the gastrointestinal tract [8]. Abnormalities in the Hh signaling pathway have been implicated in the myriad of disorders associated with VACTERL syndrome and congenital duodenal obstruction [9]. As a single entity, duodenal obstruction (atresia and web) has a high association with an abnormal karyotype, most commonly Trisomy 21 (30%), although not present in our patient [1].

Complete duodenal obstruction resulting from duodenal atresia can be detected by antenatal ultrasound when a double-bubble sign is present [1,7,10]. However, when bowel narrowing is present without complete obstruction, any underlying stenosis or web of the duodenum may be clinically, and radiographically occult. The severity of obstruction from a duodenal web varies depending on the size of the hole or aperture of the distal web. In our case, both the annular pancreas, and the duodenal web were clinically occult until an ingested coin occluded the duodenal web resulting in proximal duodenal obstruction.

At birth our patient presented with a hypogenetic lung, partial anomalous pulmonary venous return, thoracic and lumbar butterfly vertebrae, fused right T11-12 ribs, and an anorectal malformation consistent with VACTERL syndrome (Fig. 6). However, she did not have a tracheo-esophageal fistula or esophageal atresia or bowel obstruction as a neonate. She also had a normal neonatal bowel gas pattern, with no double-bubble sign to suggest duodenal stenosis. Were it not for the swallowed coin, at 4 years of age, resulting in obstruction of her then occult duodenal web, the remaining abnormalities of this syndrome may have remained unknown. Radiologists should consider the diagnosis of congenital duodenal obstruction when encountering patients with a retained foreign body in the duodenum, especially in patients with VACTERL syndrome.

Disclaimer

The views expressed are those of the authors and do not reflect the official views or policy of the Defense Health Administration, the Department of Defense or its components.

Patient consent

The authors confirm that informed consent was obtained from the parents of the pediatric patient about which this case report is written.

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