



## Case report

## Ectopic papilla of Vater in the pylorus presenting with cholangitis: A case report with literature review

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## ABSTRACT

**Introduction:** The papilla of Vater is situated in the second part of the duodenum. The current study aims to report a rare occurrence of an ectopic papilla of Vater in the pyloric region presenting with cholangitis.

**Case report:** A 59-year-old male patient presented with right upper quadrant pain, anorexia, nausea, and jaundice. He was feverish and exhibited tenderness in the right upper quadrant. Endoscopic retrograde cholangiopancreatography revealed an ectopic papilla of Vater on the pyloric canal. A gastroscop was used instead of a duodenoscope for better visibility of the opening, easier cannulation, and a less risky sphincterotomy. He returned one year after his last procedure with no symptoms and no recurrence of acute cholangitis.

**Discussion:** It has been suggested that developmental defects are acquired during embryogenesis. If subdivision happens early in embryogenesis, leaving the pars hepatica above the zone of proliferation that divides the stomach from the duodenum, the pars hepatica will develop into a duct that empties into the pylorus area.

**Conclusion:** It is preferable to use a gastroscop rather than a duodenoscope to visualize and manipulate the common bile duct in the case of an ectopic papilla of Vater in the pylorus.

## 1. Introduction

The papilla of Vater is an opening at the junction of the common bile duct (CBD) and the pancreatic duct. It is situated in the second part of the duodenum and is encircled by the sphincter of Oddi. The abnormal ending of the CBD is known as the ectopic papilla of Vater. It is a rare condition and may be associated with faulty embryogenetic development [1]. Because of the small number of cases, the incidence may be underestimated, and the condition may go unrecognized until symptoms appear [2]. An ectopic origin distal to the second duodenum part, the third and fourth duodenal parts, has been commonly reported, with a frequency rate ranging from 5.6% to 23% [3]. Increases in the prevalence might be associated with the increasing use of endoscopic retrograde cholangiopancreatography (ERCP) [2]. However, ectopic papillae are extremely rare in proximal positions, with only a few cases having been reported in the duodenal bulb, pylorus, and stomach [4].

The current study aims to report a case of an ectopic papilla of Vater

in the pyloric region presenting with cholangitis. The report has been arranged in line with SCARE guidelines and includes a brief literature review [5].

## 2. Case report

## 2.1. Patient's information

A 59-year-old male patient was referred by a general surgeon for doing endoscopic retrograde cholangiopancreatography (ERCP). He had right upper quadrant (RUQ) pain, anorexia, nausea, and jaundice intermittently for two months duration. The pain was severe, a score of 7 out of 10. In the last two weeks before referral, the pain became continuous with a milder pain score and a persistent fever. Two years before the recent presentation, he underwent cholecystectomy (that was difficult and had a prolonged period procedure) due to a diagnosis of calculus cholecystitis. He had unremarkable past medical, family, and

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social history with no chronic drug use.

## 2.2. Clinical examination

The patient looked ill and had jaundice. Vital signs only showed fever (38.4 °C), and he had tenderness in RUQ Region. No abdominal mass, no evidence of organomegaly or lymphadenopathy.

## 2.3. Diagnostic assessment

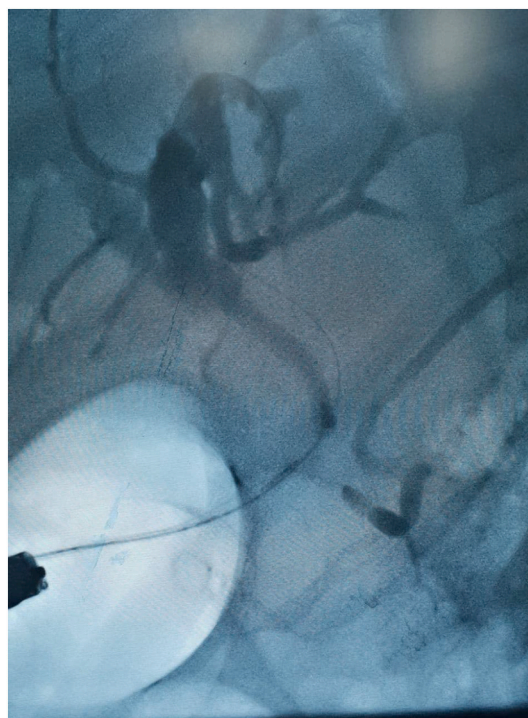
Liver biochemical tests revealed ALT (175 IU/ml), direct type total serum bilirubin (3.5 mg/dl), and alkaline phosphatase (ALP) (96 IU/ml). Pancreatic enzymes and renal function tests were normal. Granulocyte leucocytosis was seen ( $28 \times 10^9/l$ ). Trans-abdominal ultrasonography (TAUS) showed dilated intra- and extra-hepatic bile ducts, with CBD reaching 15 mm and a minor filling defect in the distal CBD. A CT scan at the beginning of the disease revealed a CBD of 17 mm and a mid-CBD filling defect. During a recurrence of symptoms and after a failed trial of the first ERCP due to the absence of a papilla, magnetic resonance cholangiopancreatography (MRCP) and endoscopic ultrasound (EUS) were performed. MRCP revealed a mid-CBD stone, and EUS revealed evidence of thickened CBD wall, indicating cholangitis. Other causes of increased liver enzymes were investigated and found to be negative, including viral hepatitis, although TAUS revealed mild fatty liver.

## 2.4. Therapeutic intervention

The first ERCP failed because cannulation was not feasible; the second ERCP revealed that the opening of the papilla of Vater was on the distal margin of the pyloric canal, as shown in Fig. 1. It was decided to use a gastroscope instead of a duodenoscope for better visibility of the papillary aperture, easier cannulation, and a less risky sphincterotomy. Following cannulation, cholangiography and pancreatography revealed a distal main pancreatic duct (MPD) in the region of the distal section of the CBD during its entry into the papilla, as shown in Fig. 2, as well as a dilated intrahepatic biliary tree and CBD. Finally, the CBD stone was evacuated, along with the placement of a biliary stent and adequate bile drainage.



**Fig. 1.** Forward view by gastroscope showing a guidewire inside papillary orifice at distal rim of pyloric canal.



**Fig. 2.** Cholangiographic image demonstrating the use of a gastroscope in the lower left corner of the image, reaching the papillary orifice with a guidewire inside CBD and the major pancreatic duct direction toward the papillary aperture. Dilated intrahepatic biliary tree (IHBT) and common hepatic duct are also demonstrated.

## 2.5. Follow up

After ERCP, the patient experienced mild epigastric pain, which was confirmed by pancreatic enzymes to be post-ERCP pancreatitis (PEP). On the third day, the patient was sent home pain-free, with a gradual decrease in TSB and liver biochemical testing. He returned after two months and had the biliary stent removed through ERCP. He returned one year after his last ERCP with no symptoms and no recurrence of acute cholangitis.

## 3. Discussion

The papilla of Vater is the major outflow for bile and pancreatic secretions that flow down along the CBD and the main pancreatic duct [6]. It is usually found in the posteromedial wall of the second part of the duodenum [7]. The opening of the CBD can sometimes be found in unusual places throughout the duodenum, primarily in the third or fourth segments of the duodenum or very rarely in the duodenal bulb [6]. More proximal locations, such as the stomach and pylorus, have been reported very rarely [8]. Lindner et al. studied 1000 intraoperative cholangiograms and discovered a 13.1% rate of distal opening compared to the typical anatomic position; however, no proximal opening was detected [9]. A retrospective analysis of 6133 patients who had ERCP in Taiwan between 1988 and 2010 found an incidence of 0.13%, which corresponds to Taiwan's general population prevalence rate [2]. In 67.8% of people with congenital biliary dilatation, the papilla of Vater was inappropriately positioned, strongly suggesting that the disorder might be caused by an ectopic papilla of Vater [10]. The relationship of embryonal abnormalities with sex has not been examined in the published literature, but case reports show a clear male predominance. In one series, 15 of the 18 patients were male, whereas, in another, seven of the eight patients were male [4,11].

Although the cause of this anomalous opening is unknown, it has

been suggested that developmental defects acquired during embryogenesis, which are yet unclear, might be a causative factor. The liver is formed by the hepatic diverticulum, which has two parts: pars hepatica, which gives rise to the intrahepatic and common hepatic ducts, and pars cystica, which gives rise to the gall bladder and cystic duct [12]. If subdivision happens early in embryogenesis, leaving the pars hepatica above the zone of proliferation that divides the stomach from the duodenum, the pars hepatica will develop into a duct that empties into the pylorus area [7].

The age of presentation ranges from 36 to 78 years old, with a median age of 55 years old [8]. Since the diagnosis was established when the patients were in their fifties, it's safe to assume that some of them remained asymptomatic for a long time [8]. The common presenting symptom in the ectopic ampulla of Vater is biliary colic pain, which is present in 95–100% of patients at the time of presentation [8]. The most common presenting symptoms reported by Taş et al. were biliary colic (100%), cholangitis (60%), acute renal failure (10%), prior bleeding (20%), and liver abscess (20%) [13]. Gurung et al. reported a case that presented with gastric outlet obstruction from pyloric stenosis, secondary to reflux of biliary and pancreatic drainage [14]. The current case was a 59-year-old male who presented with intermittent severe right upper quadrant pain, associated with anorexia, nausea, and jaundice for two months duration.

Gallstones, choledocholithiasis, obstructive jaundice, cholangitis, pancreatitis, and peptic ulcers have all been reported as consequences of ectopic papillae in previous publications [6]. The following are some of the possible causes of biliary disease: First, the distal CBD's hook-shaped configuration, due to an acute angulation of the CBD, can result in bile stasis and cause defective bile drainage, which might lead to choledocholithiasis. Second, a sphincter of Oddi that is underdeveloped or missing allows intestinal germs and duodenal contents to reflux into the biliary system, resulting in cholangitis in the setting of biliary blockage [6,7]. Almost always, recurrent duodenal ulcers and duodenal stenosis are present [15]. According to two small studies, the prevalence of bile duct stones, cholangitis, and acute pancreatitis is 56%, 39%, and 18.2%, respectively, owing to the distal common bile duct's hook-shaped form [3,4]. The current case presented with cholangitis, and he had a past surgical history of cholecystectomy.

Although CBD congenital abnormalities are more prevalent in children, they can also be found in adults. An anomalous opening of the CBD in adulthood can be discovered incidentally based on symptoms mentioned previously [3]. Currently, percutaneous transhepatic cholangiography, upper gastrointestinal endoscopy, ERCP, MRCP, and barium studies can all be used to diagnose an ectopic papilla of Vater [16]. The absence of visibility of the main papilla in the second duodenal segment may be the key to excluding a fistula caused by an ulcer. Furthermore, because the dilated CBD might terminate abruptly, it can be difficult to distinguish abnormal CBD discharge from periampullary malignancy [8].

The management of this anomaly is controversial, with symptomatic patients being treated surgically or endoscopically. When there is an ectopic papilla of the Vater with choledocholithiasis, endoscopic stone removal might be challenging. In some people, the intramural component of the duct is not fully formed. As a result, endoscopic sphincterotomy carries a significant risk of perforation or hemorrhage. Endoscopic sphincterotomy should therefore be avoided in these individuals [4,17]. ERCP-related procedures, such as balloon catheterization or stenting, should be the primary therapy [17]. In the vast majority of the cases, the ectopic opening is obscure, making cannulation difficult during ERCP [6]. The rate of cannulation failure in patients with ectopic papilla of Vater has not been studied [3]. Due to the high prevalence of duodenal stenosis in patients with ectopic papilla of Vater, gastroscopy-based ERCP is more commonly used [18]. In the current case, a gastroscopy was used instead of a duodenoscopy for better visualization, easier cannulation, and a less risky sphincterotomy. Following cannulation, cholangiography and pancreatography revealed

a distal MPD in the region of the CBD's distal segment during its entrance into the papilla, as well as a dilated intrahepatic biliary tree and CBD. Finally, the CBD stone was removed, accompanied by the placement of a biliary stent and proper bile drainage. The overall complication rate of ERCP was 18.7%, including PEP (3.7%), biliary tract infection (2.8%), and hyperamylasemia (12.1%). All of these complications are minor and may be treated with conventional therapies [18].

In conclusion, an ectopic papilla of Vater in the pylorus is a very rare condition. It is preferable to use a gastroscopy rather than a duodenoscopy to visualize and manipulate CBD in the case of an ectopic papilla of Vater in the pylorus. If the papilla cannot be seen, it is best to consider ectopic papilla before discontinuing the procedure.

## Consent

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

## Ethical approval

Approval is not necessary for case report (till 3 cases in single report) in our locality.

The family gave consent for the publication of the report.

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## Guarantor

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## Research registration number

Not applicable.

## CRediT authorship contribution statement

Abdulwahid M. Salh: major contribution of the idea, literature review, final approval of the manuscript.

Dana T. Gharib: physician performing the manage, final approval of the manuscript.

Fahmi H. Kakamad: Writing the manuscript, literature review, final approval of the manuscript.

Suhaib H. kakamad, Shevan M. Mustafa, Shvan H. Mohammed, Taha Alkarboly: literature review, final approval of the manuscript.

## Declaration of competing interest

None to be declared.

## References

- [1] U. Saritas, A. Senol, Y. Ustundag, The clinical presentations of ectopic biliary drainage into duodenal bulb and stomach with a thorough review of the current literature, *BMC Gastroenterol.* 10 (1) (2010) 1–8.
- [2] Y.C. Peng, W.K. Chow, Ectopic papilla of Vater in duodenum bulb: a hospital-based study, *Medicine* 98 (8) (2019).
- [3] E. Parlak, S. Dişibeyaz, C. Cengiz, B. Çiçek, Y. Özın, S. Kacar, et al., Ectopic opening of the common bile duct and duodenal stenosis: an overlooked association, *BMC Gastroenterol.* 10 (1) (2010) 1–7.

- [4] S.S. Lee, M.H. Kim, S.K. Lee, K.P. Kim, H.J. Kim, J.S. Bae, et al., Ectopic opening of the common bile duct in the duodenal bulb: clinical implications, *Gastrointest. Endosc.* 57 (6) (2003) 679–682.
- [5] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, et al., The SCARE 2020 guideline: updating consensus Surgical Case Report (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.
- [6] J.M. Lee, H.J. Kim, C.Y. Ha, H.J. Min, H. Kim, T.H. Kim, et al., Ectopic opening of the common bile duct into the duodenal bulb accompanied with cholangitis and gallbladder cancer: a report of two cases, *Clin.Endosc.* 48 (3) (2015) 260.
- [7] O. Sezgin, E. Altıntaş, E. Üçbilek, Ectopic opening of the common bile duct into various sites of the upper digestive tract: a case series, *Gastrointest. Endosc.* 72 (1) (2010) 198–203.
- [8] S. Disibeyaz, E. Parlak, B. Cicek, C. Cengiz, S.O. Kuran, D. Oguz, et al., Anomalous opening of the common bile duct into the duodenal bulb: endoscopic treatment, *BMC Gastroenterol.* 7 (1) (2007) 1.
- [9] H.H. Lindner, V.A. Pena, R.A. Ruggeri, A clinical and anatomical study of anomalous terminations of the common bile duct into the duodenum, *Ann. Surg.* 184 (5) (1976) 626.
- [10] L. Li, A. Yamataka, Y.X. Wang, D.Y. Wang, K. Wang, Z.X. Li, et al., Anomalous pancreatic duct anatomy, ectopic distal location of the papilla of Vater and congenital biliary dilatation: a new developmental triad? *Pediatr. Surg. Int.* 19 (3) (2003) 180–185.
- [11] H.J. Lee, H.K. Ha, M.H. Kim, Y.K. Jeong, P.N. Kim, M.G. Lee, et al., ERCP and CT findings of ectopic drainage of the common bile duct into the duodenal bulb, *Am. J. Roentgenol.* 169 (2) (1997) 517–520.
- [12] C.E. Tan, G.J. Moscoso, The developing human biliary system at the porta hepatis level between 29 days and 8 weeks of gestation: a way to understanding biliary atresia. Part 1, *Pathol. Int.* 44 (8) (1994) 587–599.
- [13] A. Taş, B. Kara, S. Ölmez, M.S. Yalçın, N.A. Öztürk, B. Saritas, Retrospective analysis of cases with an ectopic opening of the common bile duct into duodenal bulb, *Adv.Clin.Exp.Med.* 27 (10) (2018) 1361–1364.
- [14] D. Gurung, H. Chung, M. Nassar, M. Alshamam, S. Eskaros, N. Niknam, S3207 ectopic location of ampulla of Vater in the duodenal bulb causing pyloric stenosis and gastric outlet obstruction, *Am. J. Gastroenterol.* 116 (2021), S1319.
- [15] M.H. Song, D.W. Jun, S.H. Kim, H.H. Lee, Y.J. Jo, Y.S. Park, Recurrent duodenal ulcer and cholangitis associated with ectopic opening of bile duct in the duodenal bulb, *Gastrointest. Endosc.* 65 (2) (2007) 324–325.
- [16] S. Nasser-Moghaddam, H. Nokhbeh-Zaeem, Z. Soroush, S.B. Sheybani, M. Mazloum, Ectopic location of the ampulla of Vater within the pyloric channel, *Middle East J.Dig.Dis.* 3 (1) (2011) 56.
- [17] E. Özasan, Ü. Saritaş, G. Tatar, H. Şimşek, Ectopic drainage of the common bile duct into the duodenal bulb: report of two cases, *Endoscopy* 35 (06) (2003) 545.
- [18] J. Hong, W. Pan, W. Zuo, A. Wang, L. Zhu, X. Zhou, et al., Efficacy and safety of therapeutic ERCP in patients with ectopic papilla of Vater, *Medicine* 99 (1) (2020).