CASE REPORT

A novel variant of pancreaticobiliary maljunction: Complete opening of the common bile duct into the duct of Santorini in a patient with pancreas divisum

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ABSTRACT

Pancreaticobiliary maljunction (PBM) is a rare congenital anomaly in which the pancreatic and bile ducts unite outside the duodenal wall, forming an abnormally long common channel that permits reciprocal reflux of pancreatic juice and bile. This leads to a high risk of recurrent pancreatitis, cholangitis, biliary stone formation, and biliary tract malignancy. Type D PBM, particularly when associated with complete pancreas divisum and aberrant bile duct drainage, represents an exceptionally rare and complex variant. We report the case of a 20-year-old female with a long-standing history of recurrent cholestasis and acute pancreatitis who presented with fever, jaundice, and abdominal pain accompanied by laboratory findings consistent with cholangitis and pancreatitis. Cross-sectional imaging and magnetic resonance cholangiopancreatography demonstrated type D PBM, complete pancreas divisum, and anomalous drainage of the common bile duct into the duct of Santorini with intraductal sludge. Emergency endoscopic retrograde cholangiopancreatography confirmed the diagnosis, and endoscopic sphincterotomy with biliary stenting led to clinical improvement. Given the persistent risk of malignancy and refractory clinical course, extrahepatic bile duct resection with Roux-en-Y hepaticojejunostomy was performed. Despite initial improvement, intermittent episodes of acute pancreatitis persisted, ultimately necessitating a pylorus-preserving pancreaticoduodenectomy. Histopathological examination revealed chronic inflammatory changes without dysplasia or malignancy. The patient recovered uneventfully and remains asymptomatic. This rare case illustrates the complexity of type D PBM associated with complete pancreas divisum and highlights the importance of early anatomical recognition and a stepwise, individualized treatment strategy. In patients with refractory symptoms and complex ductal anomalies, surgical intervention beyond endoscopic management may be required to prevent recurrent disease and reduce the risk of malignant transformation.

Keywords: Acute pancreatitis; endoscopic retrograde cholangiopancreatography; hepaticojejunostomy; pancreas divisum; pancreaticobiliary maljunction.

INTRODUCTION

Pancreaticobiliary maljunction (PBM) is a rare congenital anomaly in which the pancreatic and bile ducts join outside the duodenal wall, resulting in an abnormally long common channel that allows bidirectional reflux of bile and pancreatic juice. This pathological reflux predisposes patients to chronic inflammation, biliary stone formation, recurrent pancreatitis, and an increased risk of biliary tract malignancy.

PBM is generally classified into four morphological types: A (stenotic), B (non-stenotic), C (dilated channel), and D (complex). In addition, cases are further stratified based on the presence or absence of extrahepatic bile duct dilatation, a key determinant in both clinical prognosis and surgical decision-making. Notably, the reported risk of malignancy is approximately 21.6% in patients with bile duct dilatation and rises to 42.4% in those without dilatation.[1-10]

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The coexistence of PBM and pancreas divisum is a highly potent anatomical combination that significantly increases the risk of pancreatitis through the simultaneous effect of two separate pathophysiological mechanisms: pancreatic drainage disorder and bile reflux. While PBM is known to increase the risk of chronic inflammation and malignancy due to abnormal mixing of bile and pancreatic enzymes, pancreas divisum has been reported to lead to recurrent pancreatitis attacks due to inadequate drainage through the minor papilla. The coexistence of these two anomalies increases the inflammatory load as described in the literature, leading to a more resistant, recurrent, and progressive clinical course.

In this report, we describe a rare case of type D PBM in a young adult with concomitant complete pancreas divisum and an anomalous drainage of the common bile duct into the duct of Santorini. The patient was managed using a staged surgical approach, beginning with extrahepatic bile duct resection and ultimately requiring a pylorus-preserving pancreaticoduodenectomy due to refractory pancreatitis. This case highlights the critical importance of early anatomical recognition and individualized surgical planning in the management of complex PBM variants.

CASE REPORT

A 20-year-old woman, born in 2002, was referred to our institution on May 05, 2022, for endoscopic retrograde cholangiopancreatography (ERCP) with a preliminary diagnosis of bile duct-related cholangitis. She presented with abdominal pain, nausea, vomiting, jaundice, and fever. Physical examination revealed icterus and tenderness in the epigastric and right upper quadrants without rebound tenderness.

One month prior, she had been hospitalized for 14 days with acute pancreatitis and cholangitis and was discharged following partial clinical improvement. Her symptoms worsened in the 2 days preceding admission. Laboratory evaluation showed leukocytosis (12,000/μL), elevated pancreatic enzymes (amylase: 341 U/L; lipase: 981 U/L), increased liver function markers (Aspartate transaminase: 126 U/L, alanine transaminase: 162 U/L, alkaline phosphatase: 288 U/L, gamma-glutamyl transferase: 234 U/L, total bilirubin: 3.2 mg/dL, direct bilirubin: 3.0 mg/dL), and markedly elevated inflammatory markers (C-reactive protein [CRP]: 281 mg/L; procalcitonin: 2.47 ng/mL). Empirical intravenous piperacillin–tazobactam therapy was initiated.

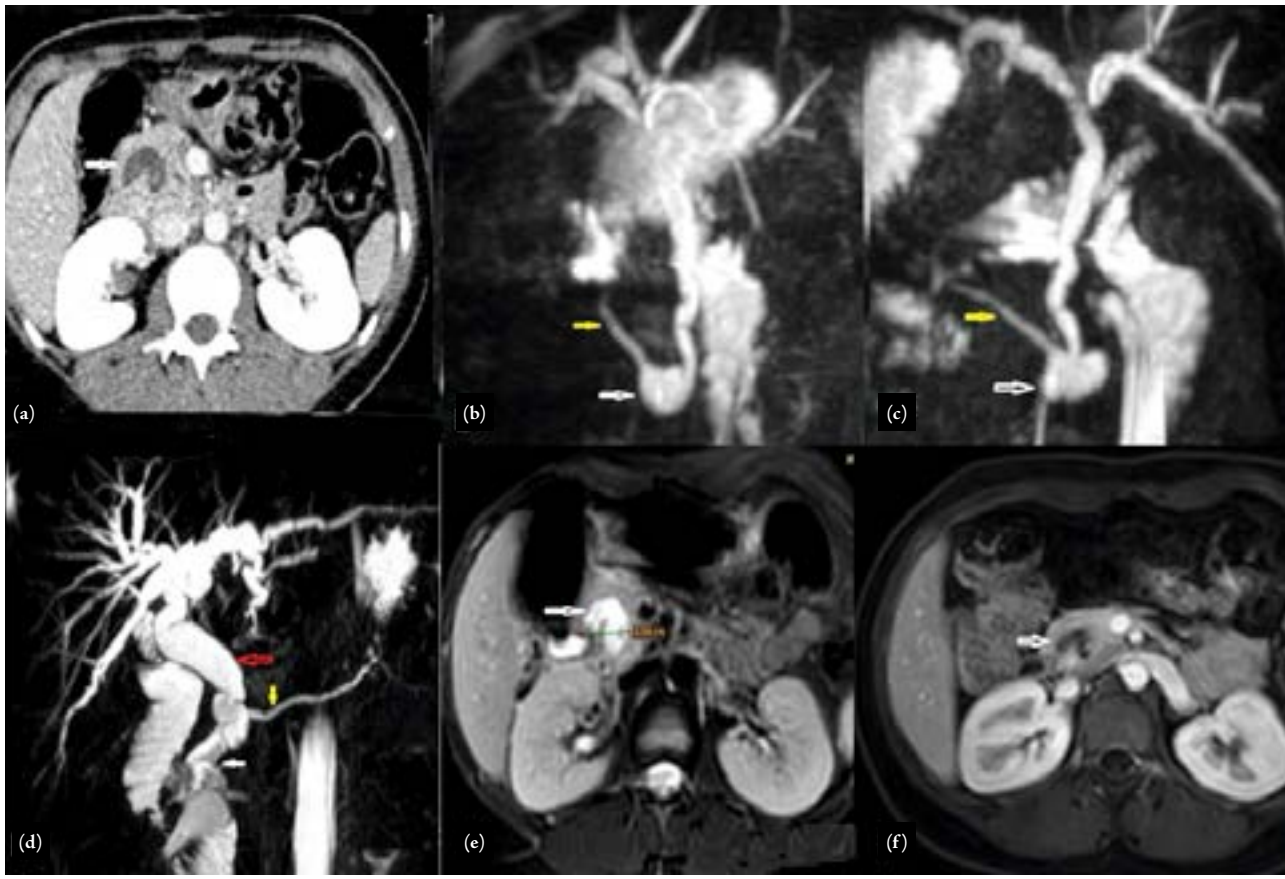

Figure 1. (a) Contrast-enhanced computed tomography showing a dilated common duct (white arrow) located in the head of the pancreas, **(b-d)** Magnetic resonance cholangiopancreatography images demonstrating the dilated common duct in the pancreatic head (white arrows), the pancreatic duct extending through the corpus and tail of the pancreas (yellow arrows) and common bile duct (red arrow), **(e and f)** Magnetic resonance imaging revealing the dilated common duct in the pancreatic head (white arrows), with visible intraductal sludge.

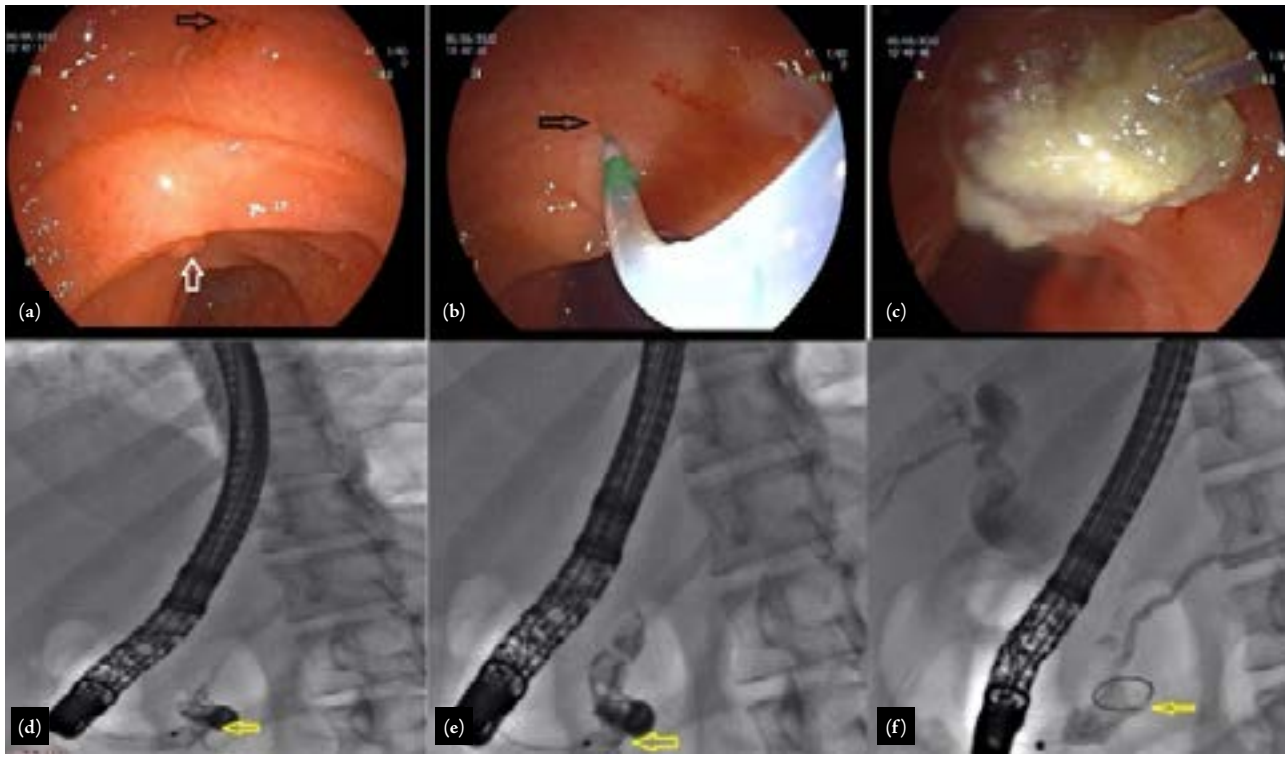

Figure 2. (a and b) Endoscopic views of the major papilla (white arrow) and minor papilla (black arrow), **(c)** Proteinaceous sludge evacuated from the common channel using a balloon catheter, **(d-f)** Fluoroscopic images showing the dilated common channel containing sludge, the dilated and tortuous distal common bile duct opening into the duct of Santorini, and the pancreatic duct extending into the corpus and tail regions of the pancreas.

Computed tomography, magnetic resonance imaging, and magnetic resonance cholangiopancreatography demonstrated type D PBM with complete pancreas divisum and anomalous drainage of a dilated common bile duct into the duct of Santorini. Intraluminal filling defects compatible with proteinaceous sludge were also identified (Fig. 1). An emergency ERCP was therefore performed.

The patient's medical history was notable for recurrent cholestatic episodes beginning at 2 years of age. After exclusion of Wilson's disease, cystic fibrosis, and metabolic and autoimmune liver disorders, benign recurrent intrahepatic cholestasis was diagnosed. A liver biopsy at the age of 5 revealed canalicular cholestasis, bile duct proliferation, and portal fibrosis. She experienced her first episode of acute pancreatitis at the age of 12 years, followed by a second episode 1 year later. At that time, a presumed cystic pancreatic lesion prompted exploratory surgery including cholecystectomy and pancreatic head biopsy, both yielding benign findings.

Despite these interventions, she continued to experience recurrent episodes of acute pancreatitis and cholangitis, occurring 3–4 times/year. ERCP in 2016 identified pancreas divisum without intervention. Genetic analysis later revealed a heterozygous mutation in the SPINK1 gene, while CFTR and PRSS1 mutations were excluded. In 2019, a minor papilla sphincterotomy and placement of a 5-Fr pancreatic stent in the duct of Santorini resulted in temporary symptom resolution.

During the emergency ERCP in May 2022, contrast injection via the minor papilla revealed a 2.5-cm dilated common bile duct containing proteinaceous sludge, which was cleared using balloon and basket catheters. A 10-Fr, 8-cm plastic stent was subsequently placed in the bile duct (Figure 2), resulting in rapid clinical improvement. CRP levels decreased significantly, and the patient completed a 2-week antibiotic regimen.

Given the markedly increased risk of malignancy associated with PBM and the history of recurrent inflammation, extrahepatic bile duct resection with Roux-en-Y hepaticojejunostomy was performed in May 2022. Histopathological analysis demonstrated epithelial hyperplasia, fibrosis, and wall thickening, without dysplasia or malignancy.

Despite surgery, the patient experienced occasional mild episodes of pancreatitis. Repeat ERCP in August 2022 revealed further sludge formation, and an 8.5-Fr pancreatic stent was placed following balloon dilation of the minor papilla. The stent was ultimately removed after another mild attack in May 2023.

Persistence of symptoms despite optimal endoscopic and biliary surgical management led to the decision to perform a pylorus-preserving pancreaticoduodenectomy in November

2024. Histopathology revealed chronic inflammatory changes without evidence of malignancy. The post-operative course was uneventful, and the patient remains symptom-free to date.

DISCUSSION

PBM is a rare congenital anomaly in which the pancreatic and biliary ducts unite anatomically outside the duodenal wall, resulting in an abnormally long common channel. This structural configuration allows reciprocal reflux of bile and pancreatic juice, predisposing the biliary epithelium and pancreatic ductal system to chronic inflammation, biliary stone formation, recurrent pancreatitis, and malignant transformation.[1,3,7,9]

Clinically, PBM is classified based on the presence or absence of extrahepatic bile duct dilatation, a distinction that has important implications for both cancer risk and surgical management. A large nationwide study from Japan demonstrated that the incidence of biliary tract cancer was significantly elevated in adults with PBM, reported as 21.6% in those with bile duct dilatation and up to 42.4% in those without it. In addition, PBM accompanied by bile duct dilatation has been strongly associated with both gallbladder cancer (62.3%) and bile duct cancer (32.1%), whereas PBM without dilatation is predominantly associated with gallbladder cancer (88.1%) and only rarely with bile duct malignancy (7.3%).[4,6,10]

At the molecular level, carcinogenesis in PBM has been linked to the accumulation of genetic alterations such as K-ras and p53 mutations in both malignant and non-malignant biliary epithelium. These changes support the classical "hyperplasia–dysplasia–carcinoma" sequence, initiated by prolonged epithelial exposure to refluxed pancreatic enzymes and bile acids, perpetuating chronic injury and abnormal cellular proliferation.[7,9]

In our patient, the heterozygous SPINK1 mutation likely acted as a disease-modifying factor that amplified the clinical impact of her underlying anatomical abnormalities. SPINK1 normally prevents premature intrapancreatic trypsin activation, and variants that reduce its function are known to increase susceptibility to pancreatic inflammation. Although heterozygous mutations alone rarely cause disease, they significantly enhance vulnerability when additional stressors such as pancreas divisum or PBM are present—an association also supported by large cohort studies showing earlier onset and more recurrent attacks in SPINK1 carriers. Experimental models further demonstrate that heterozygous loss of SPINK1 increases the severity of acute injury and promotes progression toward chronic pancreatitis, consistent with the persistent relapsing course observed in our case. Therefore, the coexistence of PBM, pancreas divisum, and a heterozygous SPINK1 mutation in the present case may have created a multifactorial predisposition to recurrent pancreatitis, with genetic susceptibility interacting synergistically with ductal anatomical abnormalities.[11,12]

Given this substantial malignant potential, current consensus, particularly from Japanese and East Asian expert panels, recommends early prophylactic surgical intervention following diagnosis of PBM, even in asymptomatic individuals. The standard operative strategy in patients with bile duct dilatation consists of cholecystectomy, complete resection of the extrahepatic bile duct, and Roux-en-Y hepaticojejunostomy. Importantly, excision should extend to the intrapancreatic portion of the bile duct to prevent remnant disease and stone formation. In morphologically complex PBM subtypes (particularly type B and D), intraoperative cholangiography and endoscopic guidance may be necessary to achieve complete resection.[4,9]

The present patient exhibited a type D PBM, an infrequently reported variant, concomitant with complete pancreas divisum. Uniquely, the common bile duct drained entirely into the duct of Santorini, an anatomical configuration that, to our knowledge, lacks prior documentation in the literature. Although no malignancy was detected histopathologically, the presence of epithelial hyperplasia, fibrosis, and chronic inflammatory change closely aligns with the early stages of PBM-related carcinogenesis, thereby supporting the appropriateness of the initial prophylactic surgical approach.[2,5,7,9,10]

Despite definitive biliary surgery, recurrent episodes of acute pancreatitis persisted. This clinical course has been described in PBM cases associated with pancreas divisum and is believed to stem from inadequate pancreatic duct drainage through the minor papilla, in combination with continued biliopancreatic reflux. In addition, the formation of proteinaceous plugs, resulting from premature trypsinogen activation and insoluble lithostathine aggregation, may contribute to ductal obstruction, elevated intraductal pressure, and recurrent inflammation.[1,2,5,9]

Given the refractoriness of symptoms to repeated endoscopic interventions, including minor papilla sphincterotomy and temporary stenting, a pylorus-preserving pancreaticoduodenectomy was ultimately performed.

The decision to perform pylorus-preserving pancreaticoduodenectomy (PPPD) in this patient is supported by current evidence indicating that PBM and pancreas divisum create a persistent inflammatory environment that often becomes refractory to endoscopic management. Surgical treatment is recommended when recurrent pancreatitis continues despite optimal endoscopic and biliary interventions, particularly in the presence of structural anomalies that perpetuate ductal obstruction and enzyme reflux.[11,12] Moreover, PBM is associated with increased long-term malignancy risk, making definitive resection essential once inflammation becomes chronic and difficult to control.[12] In our case, repeat episodes of pancreatitis and cholangitis despite sphincterotomy, stenting, and extrahepatic bile duct resection fit well within accepted indications for surgical escalation, reinforcing PPPD as an evidence-based and clinically justified intervention.

Although conventionally reserved for malignancy, this procedure is increasingly recognized as a valid therapeutic option in selected young patients suffering from disabling, refractory pancreatitis secondary to complex congenital ductal anomalies.[6,10] Final histopathological analysis demonstrated features of chronic pancreatitis without dysplasia or neoplasia, and the patient has remained asymptomatic following surgery.

Emerging data underscore PBM as a dominant etiological factor in early-onset biliary tract malignancies, reinforcing the concept that congenital anatomical derangements play a greater role than environmental exposures in these patients.[6,10] This further emphasizes the necessity of early recognition, careful surveillance, and timely prophylactic intervention in individuals diagnosed with PBM, particularly in children and young adults presenting with recurrent cholangitis or pancreatitis.

CONCLUSION

This case presents a rare and previously undescribed anatomical variant of PBM, characterized by complete drainage of the common bile duct into the duct of Santorini in the presence of pancreas divisum. The coexistence of bile duct dilatation, recurrent pancreatitis, and the well-established malignant potential of PBM justified definitive surgical management. Initial prophylactic extrahepatic bile duct resection was undertaken to mitigate cancer risk, followed by pylorus-preserving pancreaticoduodenectomy due to persistent post-operative pancreatitis.

This report highlights the critical importance of early recognition of PBM in young patients with recurrent pancreatobiliary symptoms. Detailed anatomical assessment and a multidisciplinary, individualized therapeutic approach are essential to optimize outcomes and prevent long-term complications, including biliary tract malignancy.

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OLGU SUNUMU - *ÖZ*

Pankreatikobiliyer malformasyonun yeni bir varyantı: Pankreas divizumu olan bir hastada ortak safra kanalının santorini kanalına tam açılması

Pankreatikobiliyer maljunksiyon (PBM), pankreatik ve safra kanallarının duodenum duvarının dışında birleştiği, anormal derecede uzun bir ortak kanalın oluştuğu nadir bir konjenital anomalidir. Bu durum, pankreatik sıvı ile safranın karşılıklı reflüsüne izin vererek tekrarlayan pankreatit, kolanjit, safra taşı oluşumu ve biliyer trakt maligniteleri için yüksek risk oluşturur. Tip D PBM, özellikle tam pankreas divizumu ve anormal safra kanalı drenajıyla birlikte olduğunda son derece nadir ve kompleks bir varyanttır. Uzun süredir tekrarlayan kolestaz ve akut pankreatit öyküsü bulunan 20 yaşında bir kadın hasta; ateş, sarılık ve karın ağrısı şikâyetleri ile birlikte kolanjit ve pankreatit ile uyumlu laboratuvar bulguları eşliğinde başvurdu. Kesitsel görüntüleme ve manyetik rezonans kolanjiyopankreatografi (MRKP), tip D pankreatikobiliyer maljunksiyon, komplet pankreas divizumu ve ortak safra kanalının Santorini kanalına anormal drenajını, ayrıca intraduktal sludge varlığını gösterdi. Acil yapılan endoskopik retrograd kolanjiyopankreatografi (ERCP) ile tanı doğrulandı ve endoskopik sfinkterotomi ile biliyer stent yerleştirilmesi sonrası klinik düzelme sağlandı. Persistan malignite riski ve dirençli klinik seyir nedeniyle ekstrahepatik safra kanalı rezeksiyonu ve Roux-en-Y hepatikojejunostomi uygulandı. Başlangıçta düzelme sağlanmasına rağmen, aralıklı akut pankreatit atakları devam etti ve nihayetinde pilor koruyucu pankreatikoduodenektomi (Whipple prosedürü) yapılmasını gerektirdi. Histopatolojik incelemede displazi veya malignite saptanmazken kronik inflamatuvar değişiklikler izlendi. Hastanın postoperatif iyileşme süreci sorunsuz seyretti ve halen asemptomatiktir. Bu nadir olgu, tam pankreas divizumu ile birlikte görülen tip D pankreatikobiliyer maljunksiyonun karmaşıklığını ortaya koymakta ve erken anatomik tanı ile basamaklı, bireyselleştirilmiş bir tedavi stratejisinin önemini vurgulamaktadır. Dirençli semptomları ve kompleks duktal anomalileri olan hastalarda, tekrarlayan hastalıkların önlenmesi ve malign dönüşüm riskinin azaltılması için endoskopik tedavinin ötesinde cerrahi girişim gerekebilir.

Anahtar sözcükler: Akut pankreatit; ERCP; Hepatikojejunostomi; Pankreatikobiliyer maljunksiyon; Pankreas divizumu.

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