AUCTORES
Globalize your Research

Journal of Thoracic Disease and Cardiothoracic Surgery
Behnam Sanei *

Open Access

Case Report

Challenging Biliary Reconstruction: A Case of Hepaticojejunostomy in Neurofibromatosis

Behnam Sanei

Associated Professor of General Surgery, Fellowship of Liver Transplant and Hepatopancreatobiliary Surgery, Isfahan University of Medical Sciences.

***Corresponding Author:** Behnam Sanei, Associated Professor of General Surgery, Fellowship of Liver Transplant and Hepatopancreatobiliary Surgery, Isfahan University of Medical Sciences.

Received date: October 17, 2025; **Accepted date:** October 29, 2025; **Published date:** November 06, 2025

Citation: Behnam Sanei, (2025), Challenging Biliary Reconstruction: A Case of Hepaticojejunostomy in Neurofibromatosis, *J Thoracic Disease and Cardiothoracic Surgery*, 6(6); **DOI:10.31579/2693-2156/153**

Abstract

A Neurofibromatosis type 1 (NF1) is a genetic disorder, caused by peripheral nerve sheath tumors in the skin or nervous system. Its prevalance is 1 in 2600 to 3000 individuls, but only 5-25% of these patients may appear intra-abdominal manifestations, which makes it a rare medical condition. This case discusses an uncommon presentation of visceral NF1 in a 75-year-old patient with cutaneous lesions of NF1, who was admitted with right upper quadrant pain and jaundice to our hospital. Preoperative imaging studies showed a dilated and tortous common bile duct, filled with multiple stones. Because of prior unsuccesful endoscopic retrograde cholangiopancreatography (ERCP) procedures, elective choledechotomy was applied for her. The main challenge of this case was finding an area without presence of neurofibroma lesions in the small intestine for anastomosis to prevent postoperative complications, such as leakage. The other consideration for finding a suitable area was probability of obstruction of the lumen, due to growth of these lesions. This case, highlights the importance of management of patients with gastrointestinal manifestations of neurofibromatosis type 1, requiring abdominal surgical intervention.

Keywords: sinus of valsalva aneurysm; aorto-right atrial fistula; sinusoplasty

Introduction

Neurofibromatosis type 1 (NF1), or von Recklinghausen, is an autosomal dominant disorder known as peripheral nerve sheath tumors in the skin or nervous system [1]. Main clinical manifestations are cutaneous neurofibromas, café-au-lait spots, axillary and inguinal freckling, and hamartomas or Lisch nodules of the iris [2]. Diagnosis is based on these clinical features and symptoms [3].

The prevalence of NF1 is approximately 1 in 2600 to 3000 individuals. However, neoplastic intra-abdominal manifestations, may appear only in 5-25% of NF1 patients. According to the rarity of intra-abdominal lesion of NF1 and non-specific symptoms, compared to cutaneous type, has made the diagnosis and treatment approach complicated [3,4]. In previous studies, involvement of the liver, colon, and small intestine have been reported in patients who were suffered from intra-abdominal lesions [5–7]. Non-specific symptoms are the main characteristic of gastrointestinal NF1, which is recognized by abdominal pain, obstruction, and jaundice [8].

Due to the complexity of the situation that arose during surgery, this case is noteworthy. The major challenge of this procedure was locating a region of the intestine free of neurofibroma in order to guarantee ideal anastomosis, reduce leakage, and prevent other possible problems.

Case Presentation

This case, describes a 75-year-old female, known case of von Recklinghausen (with café-au-lait spots and neurofibromatosis lesions on her body) who presented with right upper quadrant (RUQ) pain and jaundice. The patient did not mention any symptoms of gastrointestinal neurofibroma, such as constipation, obstruction, or abdominal mass. Past medical history of hypertension or diabetes mellitus was not reported. She had undergone appendectomy and hernia repairing surgery in recent years. Her gallbladder had been removed 8 years ago, followed by three endoscopic retrograde cholangiopancreatography (ERCP) procedures due to common bile duct (CBD) stones, with the most recent occurring 5 years ago. Despite these interventions, she continued to experience abdominal pain, indicative of choledocholithiasis.

Pre-operative:

Imaging studies, such as sonography and magnetic resonance cholangiopancreatography (MRCP), were performed as part of the preoperative evaluation for diagnosis and ruling out other medical conditions, such as malignancy. These revealed a significantly dilated and tortuous CBD, with a maximum diameter of 30 mm, filled with stones, and significant ectasia of the intrahepatic biliary ducts (Figure 1).

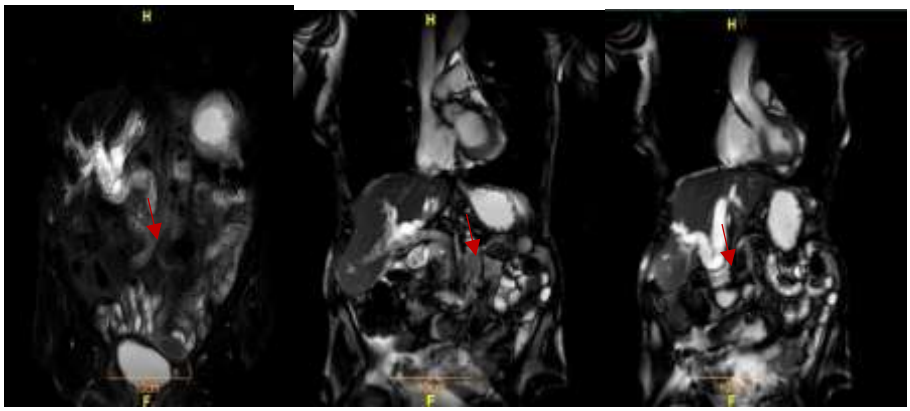

Figure 1: MRCP demonstrating dilation of CBD and presence of stones.

Initial laboratory tests revealed white blood cell count of 3820/µL, hemoglobin of 12.4 g/dL, platelet count of 137000/µL, AST level of 84, ALT level of 95, ALP 839, PT of 13.2 seconds, and PTT of 39.6 seconds. Tumor markers were negative.

Given these findings and the previous unsuccessful ERCP procedures, the decision was made to perform an elective choledochotomy. Due to presence of cholangitis, hydration and antibiotic therapy were commenced. Pre-operative evaluation was done and her ejection fraction (EF) was calculated 50-55% by echocardiography as ordered.

During operation:

An appropriate surface was achieved by doing a midline incision. Adhesion bands were observed during surgical exploration due to previous surgeries, necessitating meticulous enterolysis for improved access to liver hilum. Choledochotomy was applied as soon as the surgical site was cleared.

The CBD was explored and exposed carefully with great care that was taken to minimize injury to crucial tissues. Biliary sludge and many stones of various sizes (maximum size was 30 mm) were removed, and the CBD was irrigated. After confirming the absence of stones, Roux-en-Y choledochojejunostomy was performed for the patient.

The main challenge during surgery was identifying a suitable segment of the small intestine devoid of lesions for performing anastomosis on jejunal wall, due to the complication introduced by widespread neurofibromas (Figure 2).

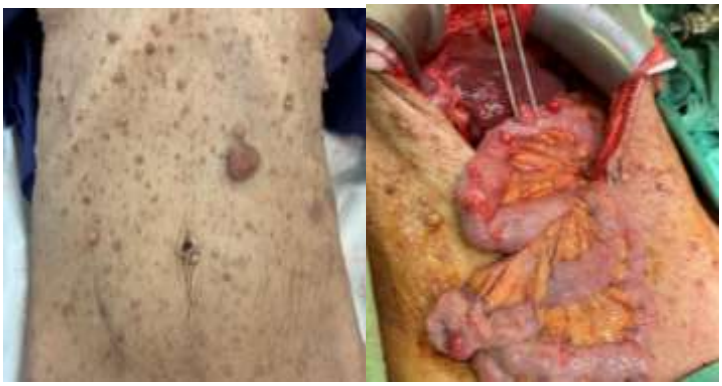

Figure 2: a: Widespread cutaneous neurofibroma lesions on the trunk b: Visceral neurofibromas in many segments of small intestine and liver, detected during surgery.

In order to reduce the chance of difficulties and guarantee a clear area for the anastomosis, the surgeon explored around mentioned lesions. After dividing the jejunum with a linear cartridge stapler and bringing the Roux limb near the hepatic duct, a side-to-side anastomosis was created between the common hepatic duct and jejunal limb to make future ERCP(s) possible, if required.

In the next step, enteroenterostomy was applied. The presence of neurofibroma restricted this surgical step, as the surgeon had to find a clear site for optimal anastomosis to ensure that the least complications such as leakage or obstruction would occur postoperatively.

Post-operative management:

The patient's recovery was stable and uneventful without any special medical problem. She was also monitored for possible complications, such as bile leakage, but none were reported. Per oral (PO) regimen was started the following day, and on the 5th day after operation, the patient discharged in good general health condition and no complications.

Discussion

Intra-abdominal neurofibromas are extremely rare, which only 5-25% of patients with NF1 may be suffered [4]. Ait Ali et al, reported a patient with intestinal obstruction and solitary neurofibroma of small bowel, which did not experience any clinical symptoms of gastrointestinal neurofibroma similarly to our case [5].

Al-Harake et al, described a case of a patient with peptic ulcer disease and hiatal hernia, who presented with epigastric abdominal pain. The patient was diagnosed with sever gastritis. Distention of stomach, duodenum, jejunum and ileum, caused by intestinal intussusception due to an ileal tumor was obvious in CT scan image. Final investigations revealed an

isolated intestinal neurofibroma with no associated syndromes in ileum [9].

In another study, Fujisawa et al, reported a 26-year-old man, who underwent a cholecystectomy prodecure. During the operation and imaging studies, he was known as a case of intra-abdominal plexiform neurofibromatosis, including periportal, mesentric, and gastrointestinal tract involvement [10].

In contrast to the skin lesions seen in NF1, the diagnosis of intestinal neurofibroma is difficult, because patients may experience severe complications, including intestinal obstruction, ischemic bowel, or perforation. Additionally, patients with gastrointestinal NF1 may suffer from medical conditions requiring intra-abdominal surgical intervention, such as Choledochotomy. The presence of neurofibromas in the abdomen, especially in the small intestine, makes the surgical procedure complicated and obscured for performing delicate intervention [5].

The treatment of choice for common bile duct (CBD) stones is endoscopic retrograde cholangiopancreatography (ERCP), which allows physicians to explore and clear the CBD from stones. In patients with multiple large stones, ERCP is not feasible and open surgical techniques are more effective, as same as our case [11-13].

This case emphasizes the challenges, encountered during the surgery of a patient with intra-abdominal neurofibroma, which made the operation process difficult, especially due to combination with adhesions from prior surgeries. Another phenomenon that surgeon had to consider was obstruction of the lumen, as consequence of growth or thickening of neurofibromatosis lesions in the anastomsosis site. The patient's preferred method of treatment was choledochotomy. This surgical intervention required careful planning with delicate anastomosis, which is important to minimize complications, such as bile leakage postoperatively [14].

Conclusion

The appearance of neurofibroma, posed difficulties by obstructing the surgical site and made it complex for surgeon to find appropriate space for anastomosis, in counter of small bowel region without presence of neurofibroma. An additional factor that surgeon had to take notice was the possibility for growth of these lesions at the anastomosis site, which consequently may cause intestinal obstruction.

This case describes the challenges of treating a patient with intra-abdominal neurofibromas. Patients with generalized skin lesions of NF1, may require surgical intervention in the abdomen and gastrointestinal tract. During surgery visceral lesions can be detected, which complicates the surgery procedure and probable anastomosis. In this case, despite the challenges faced during choledochotomy procedure, the patient's surgery was quite successful without no major complication, which emphasizes the importance of identifying a lesion-free segment of the intestine for anastomosis in cases of intra-abdominal NF1.

References:

1. Le C, Bedocs PM. Neurofibromatosis.
2. D P, Jn Y, H T, R RO, N G, Rm B. Dermatologic Manifestations of Neurofibromatosis Type 1 and Emerging Treatments. Cancers
3. Adil A, Koritala T, Munakomi S, Singh AK. Neurofibromatosis Type 1.
4. Dare AJ, Gupta AA, Thipphavong S, Miettinen M, Gladdy RA. (2020). Abdominal neoplastic manifestations of neurofibromatosis type 1. Neuro-Oncol Adv.;(2): 24-33.
5. Ait Ali H, Zeriouh B, Egyir Ebo U, Serji B, Elharroudi T. (2021). Solitary and complicated neurofibroma of small Bowel: A case report. Int J Surg Case Rep; 78:126-129.
6. Yokogawa Y, Suzuki T, Suzuki H, Nemoto R, Shimizu H, Ueda T, et al. (2023). Neurofibromatosis Type 1 with a Giant Diffuse Plexiform Neurofibroma Invading the Liver. Intern Med Tokyo Jpn.;62(20):2971-2975.
7. Juakiem W, Cebe K, Cochet A. (2016). Neurofibromatosis; A Rare Colon Polyp in a Forgotten Disease: 1358. Off J Am Coll Gastroenterol ACG;111: 612.
8. Agaimy A, Vassos N, Croner RS. (2012). Gastrointestinal manifestations of neurofibromatosis type 1 (Recklinghausen's disease): clinicopathological spectrum with pathogenetic considerations. Int J Clin Exp Pathol.;5(9):852-862.
9. Al-Harake A, Chour M, Al Beteddini OS. (2013). Solitary intestinal neurofibroma with no associated systemic syndromes causing intussusception: Case report and literature review. Int J Surg Case Rep.;4(7):629-632.
10. Fujisawa T, Takata M, Ouchi S, Ueyama S, Nakajima T, Mitsutsuji M, et al. (2011). Intra-abdominal plexiform neurofibromatosis including periportal, mesentery, and gastrointestinal tract involvement in neurofibromatosis type 1: case report and review of the literature. Clin J Gastroenterol.;4(5):292-297.
11. McNicoll CF, Pastorino A, Farooq U, Froehlich MJ, St Hill CR. holedocholithiasis.
12. Chen H, Jorissen R, Walcott J, Nikfarjam M. (2020). Incidence and predictors of common bile duct stones in patients with acute cholecystitis: a systematic literature review and meta-analysis. ANZ J Surg.;90(9):1598-1603.
13. Almadi MA, Barkun JS, Barkun AN. (2012). Management of suspected stones in the common bile duct. CMAJ Can Med Assoc J;184(8):884-892.
14. Seeras K, Qasawa RN, Kashyap S, Kalani AD. Bile Duct Repair.

DOI:10.31579/2693-2156/153