2019-03-15

Aortoduodenal Fistula Forms From Primary Aortic Stump Graft in a Two-Time Multi-Visceral Transplant Patient with Presentation of Gastrointestinal Bleed and Bowel Perforation: A Case Report

Brielle Corrente

Aortoduodenal Fistula Forms From Primary Aortic Stump Graft in a Two-Time Multi-Visceral Transplant Patient with Presentation of Gastrointestinal Bleed and Bowel Perforation: A Case Report

Brielle Marie Corrente PA-S

Duquesne University

Word Count: 1875

Abstract:

Usually not diagnosed until open laparotomy, aortoduodenal fistulas (ADF) are one of the rarest complications of intestinal transplant surgery. With an incidence rate of only 0.04% at autopsy and only 250 documented cases since the early 1800's, aortoduodenal fistulas are the most deadly complications of intestinal transplantation with a mortality rate of 100% without surgical intervention. A 39 year old, two-time multi-visceral transplant African American female patient suffered from a primary aortoduodenal fistula formation in a primary modified multi-visceral transplant aortic stump graft site. With emergency open laparotomy repair, revascularization of the secondary multi-visceral transplant was performed, saving the life of the patient and preserving the current multi-visceral transplant. Due to the rising number of intestinal transplants and multi-visceral transplants performed, clinicians should always have high suspicion of aortoduodenal fistulas in any transplant patient that presents with acute abdominal pain and lower gastrointestinal bleeding. With quick diagnosis, survival rate of aortoduodenal fistulas in multi-visceral transplant patients may improve.

Introduction

Multi-visceral transplant (MVT) is the least common transplant surgery. A rare complication of intestinal transplant (IT) is aortoduodenal fistulas (ADF) with an incidence rate of 0.02% of primary fistulas and 1% in patients with abdominal aorta reconstruction, such as a superior mesenteric artery (SMA) graft from a MVT. Most frequently, ADF is a lethal condition and requires surgery in 100% of cases with the first case was reported by Salmon in 1843.[1] Since then, only 250 cases have been reported.[2] This being the first case ever reported of an aortoduodenal fistula presenting in a two time multi-visceral patient with fistula formation in the primary aortograft stump. Understanding rare complications of MVT, the importance of having a

clinical suspicion of ADF in MVT patients and surgical management is essential in decreasing mortality.

Case Synopsis

A 39-year-old African American female patient presented after transfer to the Small Bowel Transplant Unit with lower GI bleeding and acute abdominal pain. Abdominal CT scan revealed a duodenal perforation requiring emergent surgery (Appendix A, Figure 1&2). The patient's past medical history included a diagnosis of Gardener's syndrome approximately 25 years ago. Gardener's Syndrome is a rare autosomal dominant genetic disorder, which is a subgene type of familial adenomatous polyposis (FAP). Symptoms of this disease include multiple dental abnormalities, adenomatous polyps of the stomach and small intestine, adrenal masses and desmoids tumors. Other types of cancers such as colon, small bowel, stomach, pancreas, thyroid, central nervous system, liver, bile ducts, and/or adrenal gland can also occur. In this patient's case, she suffered from multiple symptoms including polyps of the stomach, small bowel and colon, multiple large desmoids tumors that were resected, and chronic bowel obstruction. After several failed treatments and resection surgeries, the patient underwent a modified multi-visceral (stomach, pancreas, small bowel, colon) transplant surgery ten years prior. Following rejection of this graft and liver failure the patient then underwent full multi-visceral transplant (stomach, liver, pancreas, small bowel, colon) two years later. The patient originally presented to an outside hospital after being found by EMS surrounded in a pool of blood said to be coming from the patient's rectum. Upon arrival to the outside emergency department the patients vital signs were a respiratory rate of 32 breaths per min, a heart rate of 122 beats per min, a blood pressure of 149/78 mmHg, and a body temperature of 36.5 C. Physical examination revealed pale conjunctiva, no heart murmur, clear breath sounds, a

previously healed laparotomy operative scar over middle abdomen, and hypoactive bowel sound with tenderness over the epigastric and periumbilical region. Labs revealed leukocytosis (WBC: 25.3), anemia (Hgb: 5.7), and a PT/INR of 36.6/3.6. While in surgery, it was discovered that the patient's original aortic graft/stump had fistulae with the small bowel/duodenal transplant graft. CT findings showed duodenal perforation and free air in the retroperitoneum or within the aortic wall, and contrast enhancement within the duodenum suggestive of hematoma formation. Due to the patient's well-known case and history, the patient was transferred to the Small Bowel Transplant Surgery Department for emergent open laparotomy. Resections of the perforated duodenum and end-to-end duodenal anastomosis were performed. While in surgery, it was discovered that the patient's original aortic graft/stump had fistulae with the small bowel/duodenal transplant graft. At this point, the patient was stabilized, a skin only closure was performed, and the patient was transferred to the transplant intensive care unit in a guarded condition due to the skin only closure raising the risk for infection. After discussion with vascular surgery a second procedure was scheduled the following day to repair the allograft ADF and reconstruct both the aortic and small bowel graft. During the second procedure, the patient was again opened in a full open vertical laparotomy, and the ADF was located. It was found that the first superior mesenteric arterial stump was, in fact, where the fistula has formed, and the new SMA was completely intact with no signs of fistula formation. The ADF was resected and an abdominal aortic repair, bowel resection, and end-to-end duodenal anastomosis were performed. The patient tolerated the procedures well and was transferred to the transplant intensive care unit in stable condition. Two days post op, the blood work revealed normalizing values of a PT/INR of 15.1/1.2, WBC of 10.5, Hbg/Hct of 8.6/26.6 and normal liver profiles (AST-28, ALT-26, ALKP-46). At the patient's most recent Small Bowel Transplant Clinic visit,

the patient was hemodynamically stable with a CBC and CMP within normal reference range limits. Her creatinine was 1.6 secondary to non-bloody diarrhea and Clostridium difficile stool studies were negative. The patients INR was within therapeutic range at 2.0. The patient's most recent computed tomography of her abdomen and pelvis showed normalized vasculature and fully healed aortic graft repair without acute changes or intra-abdominal collections or abnormalities associated with the reconstructed aortagraft. The patient is able to fully tolerate oral intake maintaining a BMI of 40. The patient's symptoms of abdominal pain and lower gastrointestinal bleeding have completely resolved with no signs of reoccurrence.

Literature Review:

Aortoduodenal fistula is rare, but now should be considered as a possible adverse effect of multi-visceral transplant surgery. Once diagnosis has been made via CT and CTA, exploratory laparotomy should be performed to lower the risk of mortality. Ultimately, treatment is also achieved through surgical approach by aortic reconstruction (in situ aortic reconstruction or extra-anatomical bypass) and duodenal repair. A comprehensive search of relevant information was done using the electronic database (PUBMED from 2014-2018). The search strategy was based on the keywords 'aortic graft fistulas', 'multi-visceral transplant complications', 'aortoenteric fistulas', 'aortic stumps', 'Primary aortoduodenal fistula', '("aortic graft AND aortoduodenal fistula"), and '("multi-visceral transplant AND aortoduodenal fistula")'. All articles were full text ~~article~~s with 4 primary resources and one secondary reference. Most literature showed an incidence rate of 0.02% of primary fistulas and 1% in patients with abdominal aorta reconstruction, most commonly diagnosed at autopsy. Literature from 2016-2019 reveals evidence based on patients with a native small bowel aortic fistula, not an allograft

fistula, making the incidence rate of allograft aortoduodenal fistula less than 0.0007 per million. This being the only case study ever reported.

DISCUSSION

According to Children's Hospital Pittsburgh's transplant team's article on the history of intestinal transplant article, over the past decade, transplantation of the intestine alone or as a component of multi-visceral grafts has progressed from "a completely experimental procedure to one that appears destined to replace chronic total parenteral nutritional support as the preferred method of treatment".[3] Prior to 1990, the literature only addressed mortality as the outcome. However, in 1995, rapid improvements in outcomes were noted due to technical and immunosuppression development. Tacrolimus or Prograf was introduced in late 1995, by the University of Pittsburgh Medical Center, as the first-immunosupressive drug since cyclosporine in the 1980s designed for use in transplant rejection prevention.

Currently there are only 30 centers worldwide doing intestinal and multi-visceral transplants. In 2017, there were 107 intestinal transplants performed with less than 1/8 of them being modified or full MVT. Since the beginning of multi-visceral transplantation about 350 transplants have been performed; fifty of those being second time patients, like the patient in this case.

In this case, the patient presented with lower gastrointestinal bleeding and a pulsatile abdominal mass palpated through the lower ¾ section of duodenum during the duodenal perforation, open laparotomy and emergency repair surgery. The patient also had a past surgical history of two multi-visceral transplant grafts which included an SMA graft, helping to prompt the diagnosis of ADF, most likely forming from the first graft connection of the superior mesenteric artery to the abdominal aorta.

CT findings showed duodenal perforation and free air in the retro-peritoneum or within the aortic wall, and contrast enhancement within the duodenum suggested hematoma formation (Appendix A Figure 2). The perforation most likely occurred secondary to the fistula formation and the increase threshold of blood flow through the bowel. CT/CTA imaging did not show the fistula due to the free fluid and hematoma formation of the duodenum obstructing the view of abdominal aortic fistula with the duodenum, leaving the only diagnostic option to be open laparotomy.

Both the celiac axis and superior mesenteric artery are re-vascularized in all of the multiple organ visceral graft combinations. A Carrel patch with the origins of these vessels can be anastomosed directly to the native aorta above or below the level of the renal arteries or through an interposition graft of donor thoracic or abdominal aorta.[4] Alternatively, the braches of bifurcation of the aortoiliac arterial graft can be directly sutured to the native aorta and individually connected to the celiac axis and superior mesenteric artery. In our case, both transplants were done via Carrel patch above the level of renal arteries each time moving the anastomosis distally in the recipient aorta, closing off the first stump and creating a new Carrel patch. The original stump, or Carrel patch repair site is, in fact, what became connected to the duodenum creating a direct arterial blood flow into the small bowel producing increased pressure in the bowel and GI bleeding. The increased pressure is likely what caused the duodenal perforation and the increased blood flow adding to the lower gastrointestinal bleed and changed lab values specifically lowered hemoglobin and hematocrit, increase PT/INR and increased leukocytosis**.**

The recommended surgical approach for ADF consists of aortic reconstruction (in situ aortic reconstruction or extra-anatomical bypass) and duodenal repair.[3] In this case, following

standards of care, aortic reconstruction was performed following bowel perforation surgery since the ADF was found during surgery. According to Tzu-Chieh Lin's case report on ADF associated with abdominal aortic aneurysm with presentation of gastrointestinal bleeding, Aortoenteric fistula (AEF) is a rare, but life-threatening condition with an annual incidence of 0.007 per million.[5] Jolanta Šumskienė's also concluded that this condition is extremely rare with an incidence rate at autopsy of 0.04% to 0.07%.[1] This evidence is based on patients with a native small bowel aortic fistula, not an allograft fistula, like in this case; making the incidence rate less than 0.0007 per million. Due to the high mortality associated with ADFs it is important for all transplant patients to be aware of the signs of aortoenteric fistulas.

CONCLUSION

Intestinal transplant is one of the least common forms of organ transplant either performed in isolation or in part of a MVT. [4] Complications of isolated IT and IT as part of MVT include complications shared with other types of organ transplants and mechanical bowel complications such as infection, obstruction, stricture, perforation and enterocutaneous fistula or vascular complications of both the venous and arterial anastomoses including stricture and pseudoaneurysm. Diagnosis of ADF should be considered in any IT/MVT patient with acute abdominal pain associated with a midline mass or lower gastrointestinal bleeding of unexplained etiology. While CT and CTA imaging may be used for confirmation, exploratory laparotomy should be performed to lower the risk of mortality. Further research into the treatment and diagnostic tools used in cases of aortoduodenal fistulas in previously transplanted patients would be beneficial for the continuing research on intestinal transplant due to the increasing numbers of transplants performed every year. Next steps in research should outline the risk factors, such as

multiple aortic manipulation surgeries, and the need for immediate diagnosis and repair by open laparotomy technique along with duodenum and aortic repair.

Appendix A.

Figure 1.

Abdominal transverse CT image: duodenal rupture

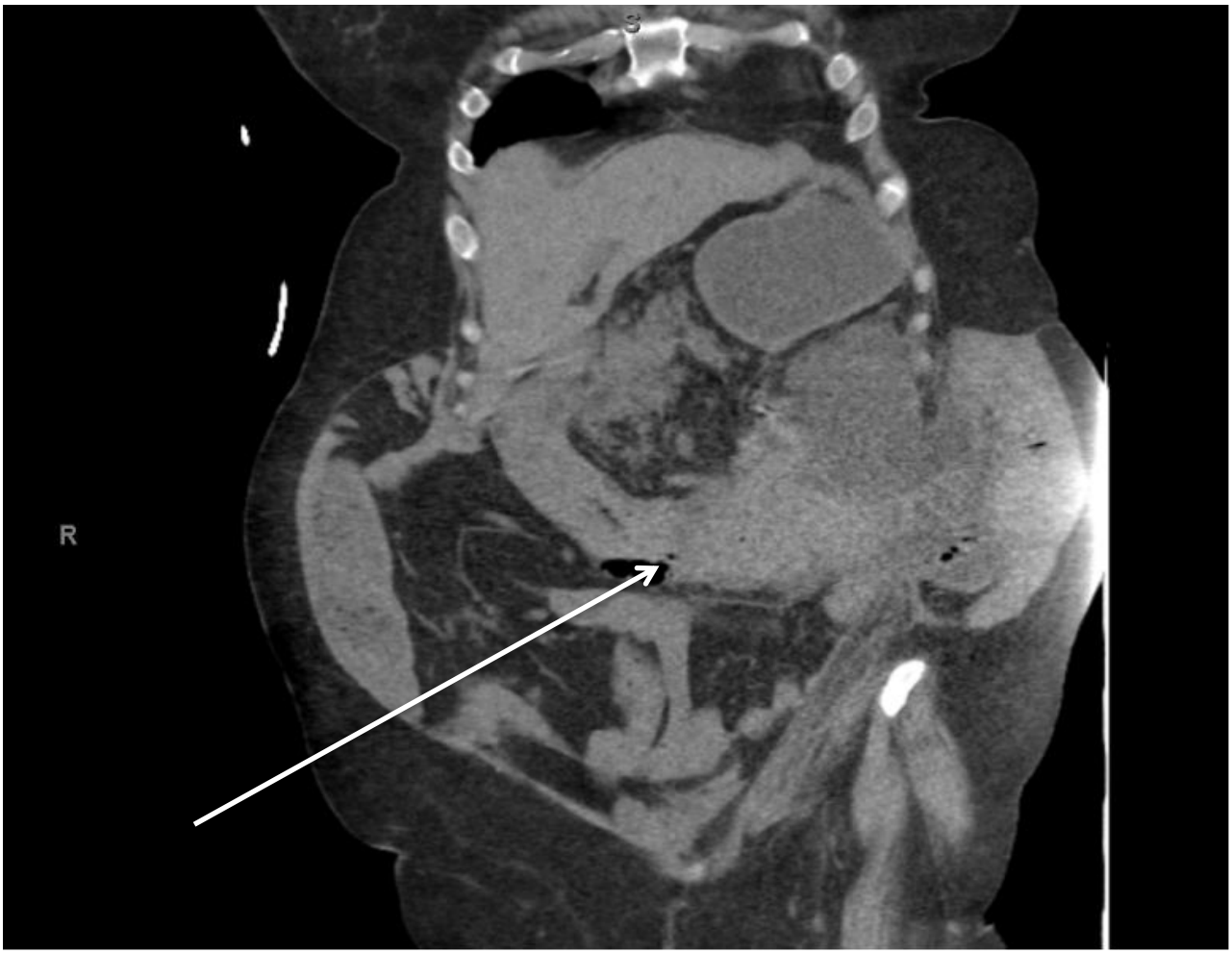
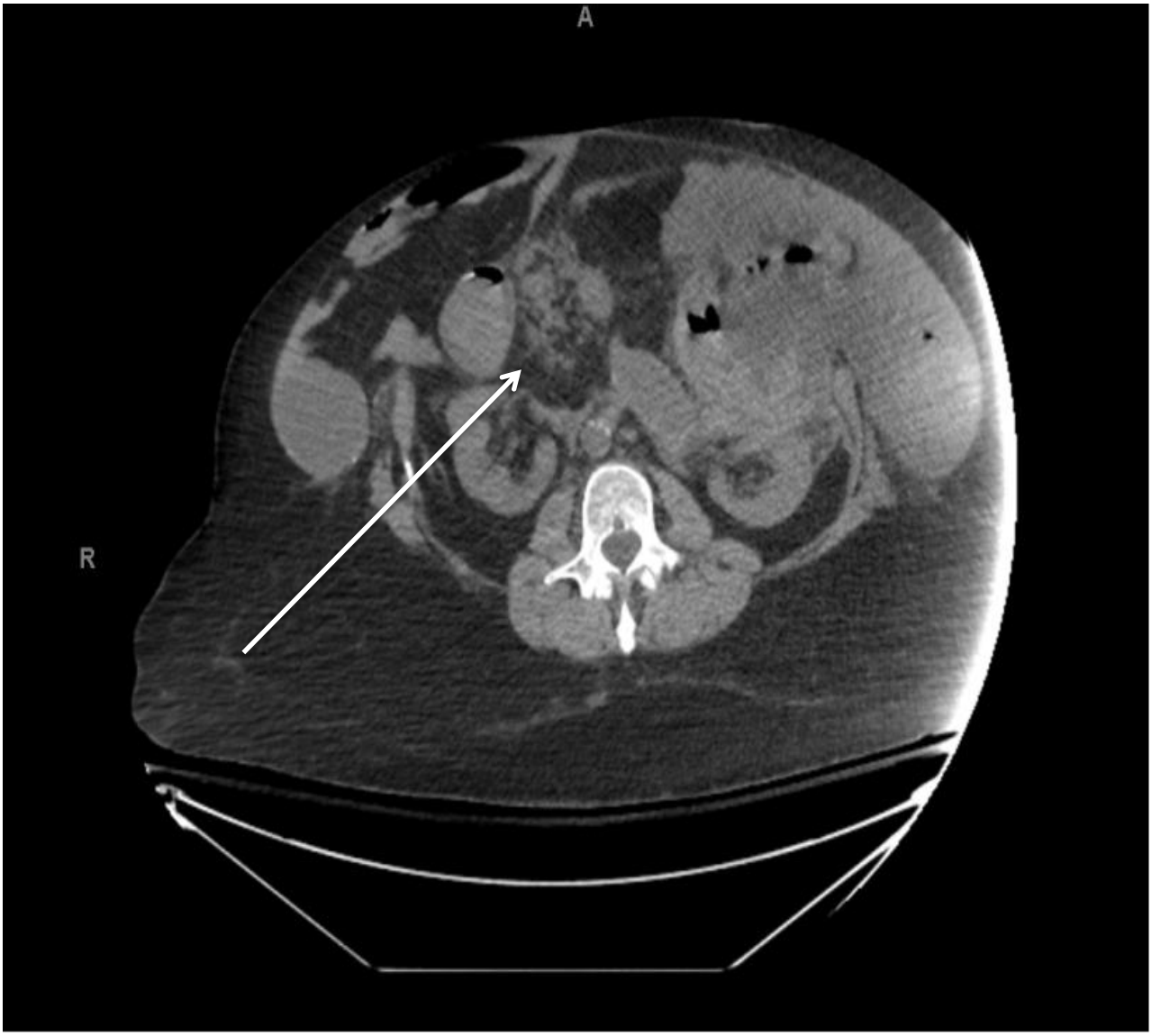


Figure 2.

Abdominal sagittal view CT image: duodenal perforation and free fluid



References

1. Jolanta Šumskienė, Edita Šveikauskaitė, Jūratė Kondrackienė, Limas Kupčinskas *Acta Med Litu*. 2016; 23(3): 165–168. doi: 10.6001/ actamedica.v23i3.3380 PMCID: PMC5287988
2. Hakim B, Myers DT, Williams TR, Nagai S, Bonnett J. Intestinal transplants: review of normal imaging appearance and complications. *TBJR*. 2018;91(1090):20180173. doi:10.1259/bjr.20180173.
3. CHP. History of Intestinal Organ Transplant | Children's Pittsburgh. Children's Hospital of Pittsburgh: History of Intestinal Transplantation. http://www.chp.edu/our-services/transplant/intestine/advances/history. Published 2019. Accessed April 10, 2019.
4. Starzl TE, Todo S, Tzakis A, et al. The many faces of multivisceral transplantation. Obstet Gynecol Sci. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2655210/. Published May 1991 available in PMC 2009 Mar 13Accessed April 10, 2019.
5. Cvjetko I. Re: Aortoduodenal fistula three years after aortobifemoral bypass: case report and literature review. *Acta Clinica Croatica*. 2017:349-349. doi:10.20471/acc.2017.56.02.21.
6. Lin T-C, Tsai C-L, Chang Y-T, Hu S-Y. Primary aortoduodenal fistula associated with abdominal aortic aneurysm with presentation of gastrointestinal bleeding: a case report. *BMC Cardiovascular Disorders*. 2018;18(1). doi:10.1186/s12872-018-0852-y.
7. Beuran M, Negoi I, Negoi RI, Hostiuc S, Paun S. Primary aortoduodenal fistula; first you should suspect it. *Braz J Cariovasc Surg*. 2016;31:261-3.