



Adenocarcinoma and Intestinal Duplication of the Ileum: Report of a Case Highlighting the Malignant Potential of an Uncommon Condition

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Abstract

Introduction: Gastrointestinal duplications are recognized, however uncommon developmental abnormalities, and may occur anywhere along the whole gastrointestinal tract, most commonly at the ileum. Their presentation in adults is an infrequent clinical condition. A recent review found only 15 cases of ileal duplication in adults published since 1884, with adenocarcinoma in 4 of those 15 cases (27%) highlighting the malignant potential of this uncommon condition.

Case Presentation: A 36-years-old female was admitted to the Emergency Unit. A Computed Abdominal Tomography (CAT) Scan demonstrated small bowel loops surrounding a tubular cystic structure which was compressing the adjacent bowel. Surgical exploration revealed multiple malignant implants covering the visceral and parietal peritoneum and infiltrating completely the omentum. At the ileal mesentery a tubular cystic whitish tumor was found. We unroofed the cyst and resected as much wall as we could, however a complete resection of the tumor was not considered. Thirty-four days after the operation the patient died. The histopathology of the cystic wall was compatible with the architecture of the intestinal wall extensively infiltrated by a mucinous adenocarcinoma moderately differentiated; a mucosal lining in parts atrophic and in parts infiltrated or replaced by adenocarcinoma was also observed.

Conclusion: This uncommon malignancy developing in an even more uncommon developmental condition such as ileal duplication highlights the importance of diagnosis and appropriate treatment in the infancy because the threatening malignant potential when presenting late in life.

Keywords: Intestinal duplication; Ileal duplication in adults; Small bowel adenocarcinoma

Introduction

Primary adenocarcinoma of the small intestine is rare, and accounts for 1% to 5% of all gastrointestinal tumors [1,2]. This neoplasm presents a challenge to the clinician due to its rarity combined with non-specific signs and symptoms that delay prompt diagnosis and treatment making its prognosis dismal with 5-year survival rates ranging from 20% to 30% [1-3]. Frequently, these tumors are found at emergency surgery for complications such as intestinal obstruction, perforation or hemorrhage, with a survival of less than a year in all cases [4,5]. Gastrointestinal duplications are recognized but uncommon developmental abnormalities, and may occur anywhere along the whole gastrointestinal tract, more commonly at the ileum. These are usually diagnosed and surgically treated in children, making their presentation in adults an infrequent and curious clinical condition [6,7]. A recent review found only 15 cases of ileal duplication in adults published since 1884 [8]. The same article reports the development of adenocarcinoma in 4 of those 15 cases (27%) and 1 case complicated with squamous cell carcinoma (7%), highlighting the malignant potential of this uncommon condition.

Case Presentation

A 36-years-old female without any previously known diseases, was admitted to the Emergency Unit of our institution with the presumptive diagnosis of acute abdomen and secondary peritonitis. She complained of abdominal pain starting seven days before associated to abdominal distension, nausea and vomiting. On physical examination the blood pressure was 131/76, with a cardiac frequency of 126 beats per minute and 38.9 °C rectal temperature. The skin was pale and diaphoretic.

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Figure 1A: Computed abdominal tomography scan showing an intestinal duplication filled with thick fluid (white arrow). Some ecstasic small bowel loops are seen surrounding the tubular structure (black arrow).

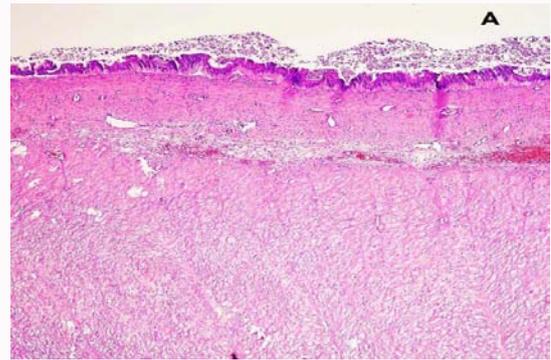


Figure 2A: Intestinal wall where it can be recognized mucosa with atrophic vellosities, areas of infiltrated adenocarcinoma, and well developed muscular wall (HE 10x).



Figure 1B: Ileal loops can be appreciated surrounding and adhering to the malignant cystic duplication. There were many visceral and parietal peritoneal implants.

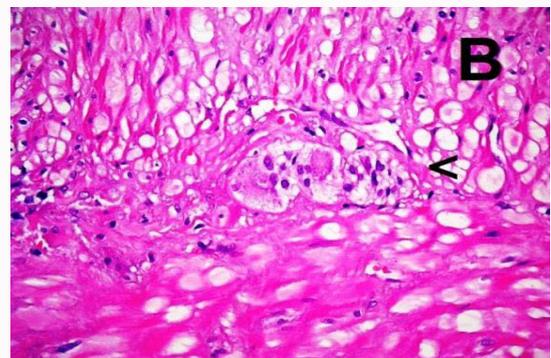


Figure 2B: Muscular wall where some myenteric plexus were recognized (arrowhead) (HE 40x).

The abdomen was distended, dull at percussion, and bowel sounds were absent at auscultation. The palpation was painful with rebound sensitivity present mainly over the left lower abdominal quadrant. Suspecting a gynecologic condition we performed a gynecological ultrasonography which was normal; however during the exam the gynecologist noticed an abnormal cystic structure adjacent to the small bowel. Subsequently a Computed Abdominal Tomography (CAT) Scan was performed; it demonstrated some small bowel loops surrounding a tubular cystic structure which was compressing the adjacent bowel, and peritoneal free fluid (Figure 1A). The white blood cell count was raised (25,700 x mm³) and the C-reactive protein was 356.06 mg/l. With the diagnosis of a complicated intestinal cystic tumor the patient underwent emergency surgery. We opened the abdominal wall with a midline incision over and below the umbilicus. The peritoneal cavity was filled with 5 liters of serous fluid that was aspirated; we collected 10 milliliters for cytological study. The exploration revealed multiple malignant implants covering the visceral and parietal peritoneum and infiltrating completely the omentum. At the ileal mesentery we found a tubular cystic whitish tumor measuring 12 cm of diameter and 15 cm of length (Figure 1B). Some loops of ileum were surrounding and adherent to this tumor. We bluntly dissected the ileal loops of the tumor and opened the cyst, it was filled with a dark serous fluid and we sent a sample to cytology. We unroofed the cyst and resected as much wall as we could, however a complete resection of the tumor was not considered due to the extensive peritoneal dissemination observed. The procedure was concluded and the postoperative recuperation was uneventful,

the patient was sent home 7 days after the surgery in relatively good condition and tolerating oral alimentation. Thirty-four days after the operation the patient died. The cytological samples taken from the peritoneal and cystic fluids were positive for malignant cells. The histopathology of the cystic wall was compatible with the architecture of the intestinal wall extensively infiltrated by a mucinous adenocarcinoma moderately differentiated; a mucosal lining in parts atrophic and in parts infiltrated or replaced by adenocarcinoma was observed (Figure 2A). A well structured muscular layer was recognized, and the myenteric nervous plexus was also identified (Figure 2B) which together with the anatomical localization of the tumor confirmed the diagnosis of malignant degeneration from ileal duplication.

Discussion

The few anecdotic reports on malignant degeneration of ileal duplications in adults highlight the important pathological potential of these rare congenital malformations when they are not diagnosed and treated during the infancy, and posteriorly presenting in adult life. According to the available literature, in the adult population, these conditions presents as an acute abdomen requiring emergency surgery, just as occurred with our patient [8-12]. Complications described in adults included intestinal obstruction, perforation, infarction, hemorrhage from ectopic gastric mucosa, and pancreatitis from heterotopic pancreas [7,8,12]. To these habitually described complications we must add malignant degeneration, which as we have seen, constitutes a rare but serious and mortal complication. The cause of intestinal duplications is unknown, however a number

Table 1: General characteristics of adenocarcinoma arising in ileal duplication.

Author	Gender	Age	Emergency surgery	Preoperative diagnosis	Tumor diameter	Segment of ileum	Survival
Babu MS et al. [8]	M	59	Yes	No	16 cm	Distal	1 year
Devos et al. [10]	F	34	Yes	No	NR	NR	NR
Ribaux C et al. [11]	M	54	Yes	No	NR	NR	NR
Tew K et al. [12]	F	57	Yes	Yes	15 cm	Middle	NR
Beltran MA (Present report)	F	36	Yes	Yes	12 cm	Middle	1 month

F: Female; M: Male; NR: Not reported

of theories, mainly vascular, have been proposed. Fifty percent of all gastrointestinal duplications occur in the small bowel, with the majority of them (>70%) in the ileum where they can assume tubular or spherical cystic forms [7]. Small bowel duplications are located on the mesenteric border and share a common blood supply with the adjacent bowel. In 1998 Long et al. [13] proposed the current classification of intestinal duplications, according to this classification; Type 1 duplications are adjacent and lateral to the mesentery of the bowel and their blood supply is parallel and independent from the blood supply of the adjacent bowel, Type 2 duplications are located between the two peritoneal layers of the mesentery and their irrigation is dependent from the adjacent bowel blood supply. The cystic malignant duplication of our patient; was a Type 1 duplication.

Preoperative diagnosis of malignant ileal duplication with ultrasonography and CAT scan was made only in one case before [12], and was suspected in our case as well, on which we also used ultrasonography and CAT scan to make the presumptive diagnosis. Ultrasonographical characteristics of malignant ileal duplications were described as a two-layered wall cyst adjacent to bowel; the inner echogenic layer represents mucosa and the outer hypoechoic layer represents muscle, a hyperechoic mass arising from the inner layer (mucosa) and projecting into the lumen can be seen and is interpreted as a tumor. CAT scan findings are useful to confirm the presence of the cystic double-layered structure and the presence of tumor in the lumen [12,14]. However, even though these characteristics were described in this patient and the radiologist and us, suspected the preoperative diagnosis of a malignant intestinal duplication, we performed an open exploration through an extensive midline laparotomy. Laparoscopic exploration is a validated and useful approach in patients with suspected malignant intestinal obstruction; nonetheless open exploration is also a safe and validated approach in these patients.

According to previous reports the treatment of malignant degeneration of ileal duplications must be resection of the compromised segment and primary anastomosis; apparently with this technique they have achieved excellent outcomes [8-12]. However most of these publications give just little detail regarding the outcome of their patients. What is clear from this small experience is that all patients presented with some complication of their tumors and were operated on emergency grounds. Only in 2 cases, including the present case, the diagnosis was preoperatively suspected based on radiological findings. Three authors reported the diameter of the duplication; all were larger than 10 cm of diameter. The segment of ileum compromised was not reported in 2 cases, in the other 3 there was 2 duplications located adjacent to the middle ileum and 1 at the distal ileum. The survival of the patient was reported only in 1 article and was of over a year, our patient barely survived 1 month, she

had very disseminated disease and, consequently her outcome was expected (Table 1). The aim of the surgical treatment of small bowel cancer is complete resection with curative intention, which can be frequently achieved (75% cases) and influences the prognosis [5]. This fact is probably similar for patients with malignant degeneration of ileal duplications. However, to the traditionally delayed diagnosis of small bowel cancer, we must add the infrequent presentation of ileal duplications in adults, making this rare condition a very dangerous scenario.

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