

Case Report

SMALL BOWEL CARCINOID - A RARE CAUSE OF RECURRENT SMALL BOWEL OBSTRUCTION

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ABSTRACT

Carcinoids are less aggressive malignant growths arising in organs derived from the embryonic foregut, midgut or hindgut. They are rare tumors of the gastrointestinal tract but most common primary tumor of small bowel. Barring the typical carcinoid syndrome, the symptoms are often vague & atypical, and vary widely. Among the commonly reported symptoms are anorexia, weight loss, bleeding, abdominal pain and mass. However, majority of carcinoid tumours are diagnosed late because of the non-specificity of complaints. We here report a similar instance of small intestinal carcinoid who presented with features of recurrent intestinal obstruction resulting in delayed diagnosis.

Key Words: *Carcinoid, Small Bowel Obstruction, Terminal Ileum*

INTRODUCTION

Carcinoid term was first applied to hormonally active tumor by Oberndorfer in 1907. It follows a more benign clinical course than most other malignancies. Approximately 85% of carcinoid tumors arise in the gastrointestinal tract, the commonest site being the appendix (50%) followed by the small intestine in 3% of patients (Bader et al. 2001 and Maglinte 2001).

Carcinoid of the small intestine is the most common distal small bowel malignancy. Patients with midgut carcinoids frequently have symptoms for long periods before a specific diagnosis is made. In these patients, early diagnosis can potentially lead to a cure by surgical resection of the primary tumor. The most common signs and symptoms of an intestinal carcinoid are abdominal pain, palpable abdominal mass and intermittent obstruction.

Obstruction usually occurs after invasion of mesentery and the resulting desmoplastic reaction with scarring. Matting of small bowel loops in turn can produce a mass and intermittently obstruct intestine. Symptoms of partial intestinal obstruction can be the result of an intense desmoplastic reaction characteristic of carcinoid tumor.

CASES

A 40 year old male reported to our surgical OPD, with one year history of recurrent abdominal pain, vomiting and constipation. Pain was initially colicky in nature which increased in intensity and frequency since past one month. He was also having recurrent vomiting for three months which used to occur 3-4 hours after meals. He also complained of constipation off and on.

There was no history of passage of blood per rectum (fresh or altered), hematemesis, or anorexia. The patient had not observed any loss of weight.

He had received Anti Tubercular Treatment for 3 months in the past; there was no other significant personal history. There was nothing pertinent in the family history either. Abdominal examination revealed a soft, mildly distended abdomen without any tenderness. No lump or organomegaly was detectable; the bowel sounds were exaggerated. On rectal examination no growth was identified. All the hernial orifices were intact.

Rest of the examination was normal. The patient was admitted as a case of recurrent intestinal obstruction most probably due to intestinal tuberculosis. Another possibility of band obstruction was kept in mind. Patient was thoroughly investigated to search for the cause of recurrent obstruction. His blood

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investigations were within the normal range except for a raised ESR(32 mm in 1st hour.) Montoux test reaction was 8x6 mm.. Nothing significant was found on chest x-ray. Abdominal x-ray (erect and supine) showed dilated small bowel loops with a few air fluid levels. CECT abdomen revealed only dilated small bowel. There was no mass, lymphadenopathy or ascites. Diagnostic laparoscopy was planned with consent of full laparotomy if needed. Laparoscopy, revealed a stricture in the ileum, 3 ft proximal to the ileo-caecal junction. No lymphadenopathy, ascites or tubercles were detected.

The liver was normal. Resection with side-to-side anastomosis was performed after giving a 5cm midline incision. The histopathological examination of the resected mass from ileum revealed a carcinoid tumor. Grossly, a small growth was seen in the ileum just underneath the strictured area. It measured 2x2 cm and the cut surface was solid, homogenous, and yellowish in color.

Microscopically, a submucosal growth consisting of solid nests of tumor cells having a monotonous appearance with small round nuclei and stippled chromatin was seen. These tumor cells were seen extending into the muscle layer.

However, serosa was uninvolved. The postoperative stay was uneventful and the patient was discharged on 5th postoperative day. Patient is under regular follow up with no evidence of recurrence.



Figure 1: Gross Pathology

Figure 1 shows the gross pathology ie 2 cm tumor arising in the wall of the bowel as a submucosal mass resulting in scarring and kinking of the surface.

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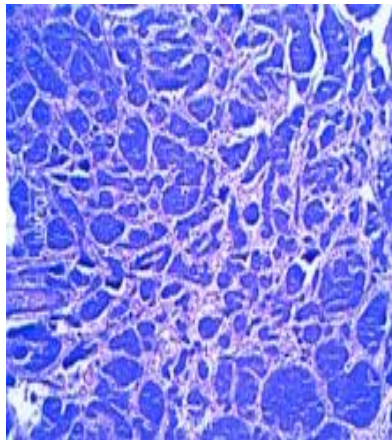


Figure 2a

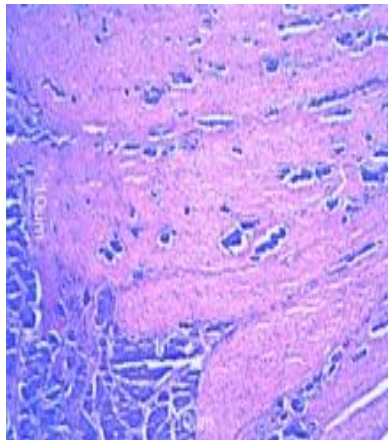


Figure 2b

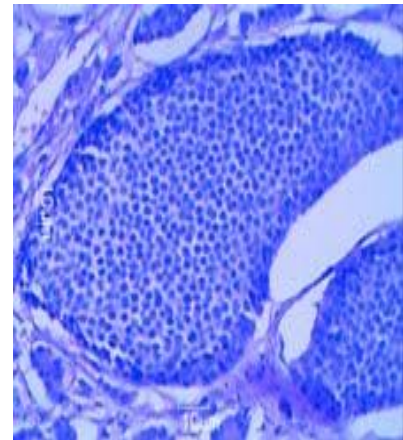


Figure 2c

Figure 2a: Scanner view showing solid nests of tumor cells having a monotonous appearance (H&E, 5X)

Figure 2b: Low power view showing tumor cells infiltrating into the muscle layer (H&E, 10X)

Figure 2c: High power view showing nests of tumor cells with peripheral palisading having regular round nuclei and stippled chromatin (H&E, 40 X)

Microscopy showed a tumor made up of small round regular cells containing a round nucleus and clear cytoplasm (Figure 2 a, b, c).

DISCUSSION

Small bowel is a less favored site for growths - only 5% of all gastrointestinal neoplasms and 1-2% of all malignant tumors of the gastrointestinal tract occur in the small bowel. The mean age at onset is about 59 years for benign tumors and about 57 years for malignant lesions; the occurrence of malignant carcinoids peaks at 62 years (Singhal *et al.*, 2013). The age of our patient was only 40 years.

Oeberndorfer was the first to use the term Karzinoide (carcinoid= carcinoma-like) to denote a less-aggressive behaviour in these tumours (Oberndorfer, 1907). Overall carcinoids are less aggressive, locally as well as distantly; the malignant potential however varies depending on the location and size of the tumour, the depth of invasion & the growth pattern. While only about 3% of appendiceal carcinoids metastasize, about 35% of ileal carcinoids are associated with metastasis, mainly in liver. The likelihood of metastasis is related to tumour size: The incidence of nodal and liver metastases is approximately 20–30% in patients with tumors smaller than 1 cm but increases to almost 60–80% for nodal metastases and 20% for liver metastases when tumors are 1–2 cm (Strodel *et al.*, 1983) In patients with primary tumors greater than 2 cm, the incidence of nodal metastases is 80% and of liver metastases is 40–50% (Strodel *et al.*, 1983). The size of the tumor in our patient was 1.5cm but there was no evidence of any metastasis to liver or any other site.

The typical carcinoid syndrome - comprising of episodic attacks of cutaneous flushing, bronchospasm, diarrhea and various cardiovascular manifestations - was first described by Thorson *et al.*, in 1954 (Thorson *et al.*, 1954). This syndrome usually follows massive metastatic infiltration of liver and since that is uncommon with the carcinoids in the midgut-derived parts of intestine, the syndrome occurs in only 10% of small intestinal carcinoids (Horton *et al.*, 2004). There was no history suggestive of carcinoid syndrome in our patient. Diagnosis is not very difficult at advanced stage which has a very poor prognosis. The 5-year survival rate from the time of diagnosis of metastatic disease is 67% (Singhal *et al.*, 2013). No therapy to date has been shown in any randomized clinical trial to prolong survival for patients

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with metastatic carcinoid tumors, and therapy remains palliative. Therefore it is imperative to diagnose a carcinoid before metastasis when it is fully amenable to surgical resection.

Benign lesions are identified more often in autopsy series while malignant neoplasms account for 75% of the symptomatic lesions. A correct preoperative diagnosis is made in only 20% to 50% of symptomatic patients. Patients with midgut carcinoids frequently have symptoms for long periods before a specific diagnosis is made. The average delay between onset of symptoms & diagnosis exceeds 8-9 years (Robertson et al., 2006). Only approximately 20% of patients are diagnosed with potentially respectable disease (Sugimoto et al., 1995). We have diagnosed this case of small bowel carcinoid almost one year after patients became symptomatic. Since the tumour is respectable at early stages, it is vital to arrive at a diagnosis early in the course of disease. The most common symptoms and signs of an intestinal carcinoid are intermittent abdominal pain, intermittent obstruction, and a palpable abdominal mass, each of which occurs in nearly 50% of patients (Singhal et al., 2013). Our patient also presented with recurrent symptoms of abdominal pain and vomiting. As the tuberculosis is very common in our country, our first diagnosis was abdominal tuberculosis, infact ATT was already started by some practitioner empirically. Because this process is extraluminal, results of the radiological examination may be normal approximately half the time while USG is nonproductive (Horton et al., 2004).

The benign tumours may produce obstruction by causing intussusceptions. The malignant intestinal carcinoids almost always lead to intermittent abdominal pain. Intermittent obstruction, seen in 15% to 35% of patients, occurs from intraluminal effects of the tumour, intussusception, or invasion of the mesentery, and the resulting desmoplastic reaction with scarring and matting of small bowel loops from tumor infiltration (Kulke and Mayer, 1999). Intestinal ischaemia, now considered to be more important in the production of abdominal pain than previously, is the result of functional and structural changes in and around the mesenteric blood vessels, caused by vasoactive substances secreted by the carcinoid tumour (Sworn et al., 1989 and Ormandi et al., 1978). In their early stages the tumors are small and confined to the bowel wall, when CT generally does not help (Picus et al., 1984). At this stage, small bowel series yields an accurate diagnosis in 50% to 70% of patients. Lesions smaller than 1 cm will not be detected by small bowel follow through, when CT enteroclysis is a better option since it has a diagnostic accuracy of about 90%. A combination of the two procedures is even more fruitful. In patients in whom there is a high clinical suspicion of carcinoid tumor but inconclusive barium studies or CT, angiography can be performed and may show the submucosal mass because of its high vascularity (Seigel et al., 1980). Lesions bigger than 1 cm are detected by CT; MRI is not very helpful¹ and USG is worthless. Carcinoid tumors of small bowel are rare tumors having an indolent course. They present with non specific symptoms resulting in delayed diagnosis. In these patients early diagnosis can potentially lead to a cure by surgical resection of the primary tumor. Therefore, carcinoid should be considered in the differential diagnosis of patients presenting with non specific symptoms and features of recurrent small bowel obstruction. Moreover, diagnostic laparoscopy is a very useful tool in patients with recurrent symptoms with inconclusive radiological investigations.

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