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Carcinoid causing Bowel Obstruction; a Case Report with Literatures Review

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Abstract

In 1907, Siegfried Oberndorfer (1876–1944), a German pathologist at the University of Munich, coined the term karzinoide, or "carcinoma-like," to describe the unique feature of behaving like a benign tumor despite resembling a carcinoma microscopically. Carcinoid tumours are characteristically low grade malignant tumors with neuroendocrine differentiation that have been described in several locations, including the gastrointestinal, respiratory, hepatobiliary, and genitourinary systems. Carcinoid tumors most commonly occur in the gastrointestinal tract (74%) and bronchial system (25%). less than 1% of cases these tumors have been reported in the genitourinary system.

In this report we describe a 55-year-old house wife female who presented with vague gastrointestinal complaints for long period then she developed intestinal obstruction due to small bowel carcinoid. The clinical findings are used to illustrate pathophysiology, classification, management of carcinoid tumours through review of literatures.

Keywords: carcinoids tumours, neuroendocrine tumours, small bowel obstruction.

Introduction

In 1907, Siegfried Oberndorfer (1876–1944), a German pathologist at the University of Munich, coined the term karzinoide, or "carcinoma-like," to describe the unique feature of behaving like a benign tumor despite resembling a carcinoma microscopically [(1)]. Carcinoids of the small bowel arise from the enterochromaffin cells, or Kulchitsky cells, found in the crypts of Lieberkuhn. These cells are also known as argentaffin cells because of their staining by silver compounds. Carcinoids may be classified by embryologic site of origin (foregut, midgut and hindgut) and secretory product (multihormonal and 5-HT, 5-HT and substance P, multihormonal) [(2)]. More recently, the World Health Organization (WHO), in an effort to clarify the classification of carcinoid tumors and to standardize a system that would enable clinicians to compare patients and predict outcomes accurately, proposed a new classification of gastroenteropancreatic NETs; based on their malignant potential [(3)].

Within the gastrointestinal tract, nearly 45% of carcinoids arise in the small intestine, making this the most common location for carcinoid tumors. Likewise, carcinoid tumors account for the highest percentage of small bowel tumors, representing approximately one third of all small intestinal neoplasm's [(4)].

These tumors commonly present in the sixth or seventh decade of life with symptoms of abdominal pain or small bowel obstruction (SBO). Small bowel carcinoids are frequently multiple, exhibiting multicentricity in up to 30% of patients, and often display metastases to the lymph nodes (39%) or the liver (31%). Development of typical carcinoid syndrome is rare, manifesting in approximately 5%-7% of patients; however, younger patients are more

likely to develop carcinoid syndrome and display a worse prognosis [5].

Malignant carcinoid syndrome, the predominant clinical feature of carcinoid tumors, results from excessive secretion of hormone products into the systemic circulation. These hormones (peptides and amines) cause the extreme symptoms of the disease, and a reduction in their circulating blood concentrations through targeted treatment is a therapeutic goal [6].

Carcinoid syndrome does not usually develop until a tumor has metastasized - usually to the liver - and the hormonal products released by the tumor reaches the circulation in substantial concentrations. The likelihood of occurrence and the associated severity of carcinoid syndrome depend on several factors: tumor size, whether its location is in an area draining into the systemic circulation, and the degree of metastasis, Carcinoid syndrome exhibits slow growth with early ill-defined symptoms and is frequently misdiagnosed as "irritable bowel syndrome" or "spastic colon". Surgical removal of the tumors is the primary therapeutic option; chemotherapy is less effective. Octreotide is the primary medical therapy for the management of certain symptoms associated with carcinoid syndrome [7].

After the onset of clinical symptoms, median survival times of 3.5 to 8.5 years have been reported; 5-year survival has ranged from 30% to 67%. The prognosis for poor patient outcome generally correlates inversely with increasing levels of urinary 5-HIAA excretion. Other biochemical indicators of a bad prognosis are high levels in the plasma of neuropeptide K and chromogranin A [8] .

Case report

The patient was a 65-year-old house wife, female who presented with vague gastrointestinal complaints, namely, softer stools, abdominal cramps, and poor appetite for more than 6 months. She developed lower colicky abdominal pain increased in severity and associated with bile stained vomiting of 3 days, the condition not responded to the usual medication and supportive measures so that the patient referred to Al-Karama Teaching Hospital for further evaluation at 25th of June, 2008. On examination an elderly female looks ill, dehydrated, vital signs show tachypnea, tachycardia and hypotension with abdominal distention and sluggish bowel sounds.

Blood investigations show elevated WBC (14000), blood urea (50 mg/ dl) and normal blood sugar and haematocrit. Plain x-ray of abdomen shows dilated small bowel loops with air fluid levels (Figures 1).

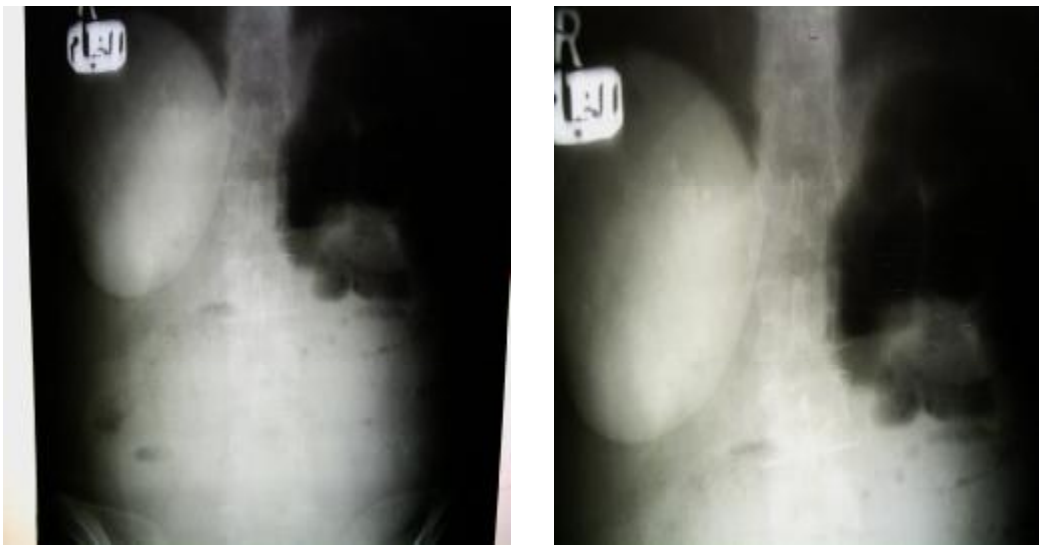

Figure 1. Plain x- ray of abdomen showing dilated segment of bowel with air fluid levels.

The diagnosis of intestinal obstruction confirmed and after adequate resuscitative measures the abdomen explored under general anesthesia through a midline incision where the cause of obstruction identified as a constricting mass in the terminal ileum and right hemicolectomy performed. The patient had uneventful smooth postoperative period while awaiting for the result of histopathology, no further treatment was required a part from follow up using regular ultrasound scanning, CT scan of abdomen and investigation including 5 Hydroxi–Indol Acetic Acid in urine which was negative both in the postoperative period and for 1 year following operation.

Macroscopic findings

Grossly, Segment of bowel, with mesentery and omentum, with the cecum 25 cm in length, with a constricting like mass (Figure 2).

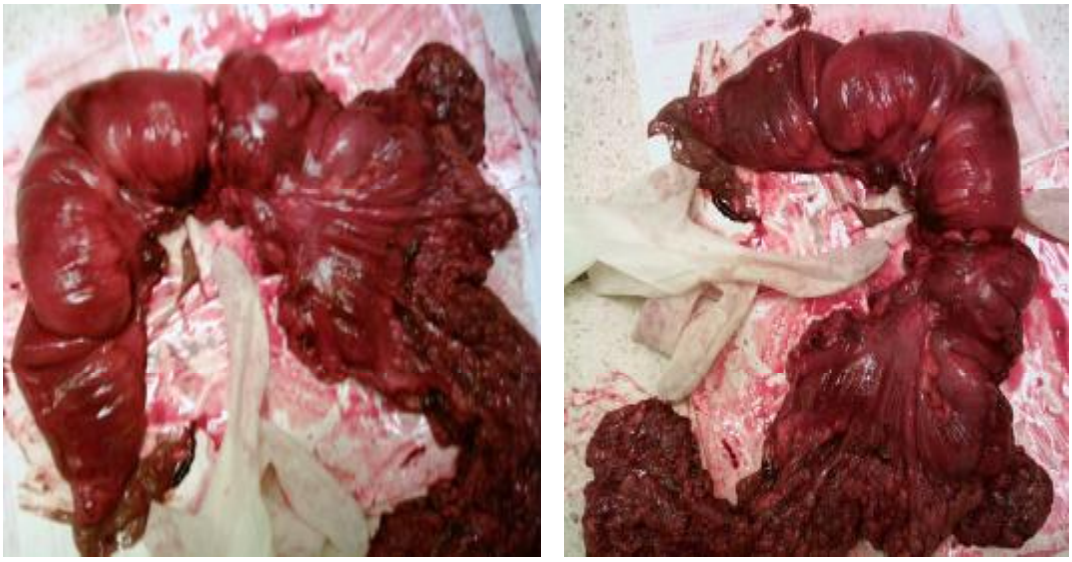

Figure 2. Segment of bowel including the cecum, with mesentery and omentum

Microscopic findings

The histopathological examination revealed solid mass of monotonous appearing cells with small nuclei, inconspicuous cytoplasm with tumor emboli in lymphatic vessels. Picture goes with carcinoid tumour of small intestine; resection margins are free from tumour (Figure 3).

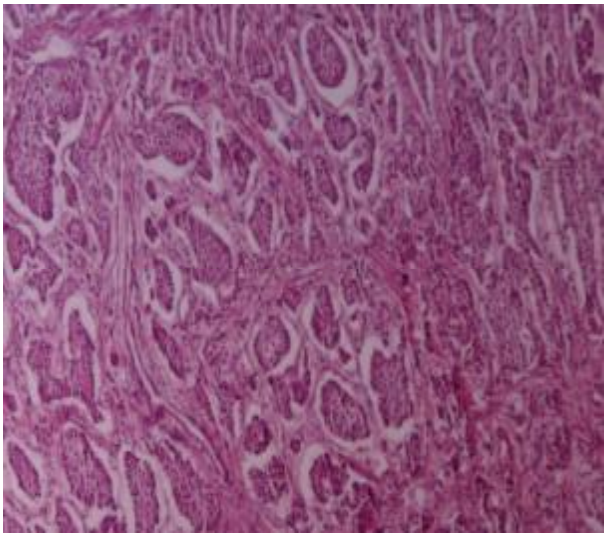

Figure 3. Solid mass of monotonus appearing cells with small nuclei.

Discussion

Carcinoid tumors are characteristically low grade malignant tumors with neuroendocrine differentiation that have been described in several locations, including the gastrointestinal, respiratory, hepatobiliary, and genitourinary systems [9]. The incidence of carcinoid tumours diagnosed during life is rising with gastrointestinal carcinoids making up the majority; earlier estimates were of fewer than 2 per 100 000 per year [10]. More recent studies have found rates approaching 3 per 100 000, the changes in incidence may result more from changes in detection than in the underlying burden of disease [11].

Many clinicians prefer to avoid surgery in patients with carcinoid neoplasia, because of its slow growth and relatively favorable prognosis, nevertheless, the commonest cause of death in patients with carcinoid is advanced metastatic disease, and both clinical and epidemiological data indicate that the more effectively the disease is ablated, the more long-lasting the benefit [12].

Although primary size is correlated with the presence of nodal with or without liver metastases, carcinoid tumours < 1 cm in diameter may be metastatic at presentation, particularly those arising within the small intestine, resection of all sizes of carcinoid with local and regional nodes is preferred to prevent nodal dissemination causing mesenteric ischemia with or without infarction, histopathological assessment helps to determine the need for hemicolectomy [12]. Liver resection has been followed by prolonged 5 year survival in several series and is recommended in appropriate patients to attempt cure or to debulk metastatic disease [13,14].

Chemoembolization may play a role in relieving symptoms and providing sustained tumor control, the aim of hepatic artery chemoembolization is to control hormone-related symptoms, to inhibit growth, and to improve the chances of survival [15]. The technique usually consists of the injection of a mixture of cytotoxic drugs, iodized oil, and Gelfoam (gelatin sponge, Upjohn, Kalamazoo, MI) into the branches of the hepatic artery supplying the tumor, this technique can result in a decrease in 5-HIAA, an improvement of symptoms, and a decrease in size of tumors [15].

Emergency surgery is likely to be indicated in the presence of an acute abdomen, whether carcinoid has previously been identified or not, or is suspected or not – more commonly not. Such surgery should be directed to remove the immediate threat to life, its extent being limited by the condition of the patient. With incipient or established multiorgan failure such circumstances are best managed with limited corrective surgery, reserving until recovery further surgery for tumour clearance or debulking if appropriate. Thus a limited emergency small bowel resection for an obstructing carcinoid tumour might be followed at a later date by elective surgery to remove further small bowel, particularly if by then a second tumour has been identified, and to undertake mesenteric lymphadenectomy [16, 17].

Conclusion

Although carcinoids are rare tumour; they should not be forgotten among causes of intestinal obstruction or patient with vague abdominal symptoms.

Recommendation

A population-based perspective of natural history and of the impact of treatment is essential to formulating an overall approach to tumour management. This is especially true for rarer tumours such as carcinoid.

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